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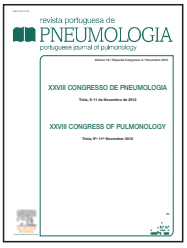

POSTERS

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CORRELATION BETWEEN TUMOR MESUREMENT ON CT SCAN AND RESECTED SPECIMEN SIZE IN LUNG CANCER

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Introduction: The optimal treatment of lung cancer (LC) relies on accurate disease staging, which is based on tumor size, regional nodal involvement, and the presence of metastasis. The 7th edition of the American Joint Committee on Cancer (AJCC) TNM classification of lung cancer difference tumors that are ≤2 cm, 2-3 cm, 3-5 cm and ≥5 cm in maximum dimension These seemingly small differences in maximum dimension play important roles in prognosis as patients are clinically staged based on maximal diameter on CT.
Aim: To compare the radiologic preoperative size of the primary tumor in lung carcinoma with their pathologic size following excision.
Materials and methods: Eighty-eight LC patients who undergone surgeries in our institution from January 2008-December 2011, CT-staged T1 or T2 were included. Images were reviewed by two independent observers. Tumor maximal diameter on axial-plane was obtained using PACS (picture archiving computed system) caliper segmentation algorithm and adjusted based on a radiologist's input; largest single diameter from pathology gross report was utilized. Descriptive statistics were used for demographic classification. A paired t-test was used to examine the measurement difference between CT and Pathology. Agreement was evaluated between CT and Pathology using Bland-Altman methods for measurements and using Cohen-Kappa for T-staging classification.
Results: Of the 88 patients 46 had adenocarcinoma, 31 had squamous cell carcinoma. The mean CT measurement was 30.27 mm, pathology was 30.63 mm. The mean difference between CT and Pathology measurements was −0.35 mm (95% Confidence Interval −2.15; 1.45, P<0.001). The lower and upper 95% limits of agreement were −17.33 mm and 16.62 mm. Clinical T-staging based on CT was T1A=21, T1B=34, T2A=20, T2B=13 and on pathology was T1A=30, T1B=22, T2A=27, T2B=9. Stage agreement was seen in T1A=17/30 (57%), T1B=16/22 (73%), T2A=14/27 (52%) and T2B=8/9 (88%) with a moderate agreement (Cohen-Kappa=0.491).
Conclusion: There was moderate agreement between CT and pathology measurements. Clinical and pathological T-stage revealed a moderate agreement. These results may have implications in decisions based on clinical staging features.

Keywords: *Cancer. Staging. Radiology.*

RARE PRESENTATION OF A PULMONARY B CELL LYMPHOMA

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The large B-cell Lymphomas represent about 25-30% of all non-Hodgkin Lymphomas and pulmonary anaplastic variant is very rare. They are an heterogeneous group of diseases with distinct clinical and pathological manifestations. The authors describe a case of a 58 year-old Caucasian men, smoker of 40 pack-year, admitted to the hospital with general symptoms and chronic cough for the last 5 months. The physical examination revealed diminished breath sounds and dullness on the lower third of the left hemithorax. He presented with anemia, elevation of acute phase parameters and an CA 125. The Chest X-ray showed hypotransparency of the lower third of the left pulmonary field. The TC scan revealed atelectasis of the left lower lobe (LLL), pleural effusion on the same side, mediastinal lymphadenopathy with obliteration of the segments of LIL and reduction of caliber of the left main bronchus (LMB). Thoracocentesis revealed the pleural liquid to be an exudate. Bronchoscopy revealed infiltration of the distal third of the trachea and LMB, luminal reduction of LMB with indication for endoscopic therapy (laser fotocoagulation, mechanical dilation and stent placement). During Hospital stay the patient was diagnosed an esophageal ulcer with covered esophageal stent placement. Bronchial and esophageal biopsies with immunohistochemical staining revealed a large B-cell non-Hodgkin Lymphoma, anaplastic variant, without medullary

infiltration. The patient underwent chemotherapy with R CHOP with good response.

Keywords: *Pulmonary B cell lymphoma. Anaplastic variant.*

RARE PRESENTATION OF A LUNG CANCER OR HAPPY COINCIDENCE?-CASE REPORT

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The authors describe a rare form of lung cancer of a patient, male, 62 years old, vigilant, smoking (20 UMA) with background irrelevant. In August 2011 was diagnosed right total spontaneous pneumothorax and respiratory failure type I. He underwent thoracic drainage with immediate lung expansion and there have been spontaneous exteriorization of chest tube in the second day of hospitalization. The control chest radiograph showed lung expansion and hypotransparency ipsilateral middle and lower third that associated with clinical criteria that however came upon that was consistent with pneumonia. He appealed again after two months with a clinical condition of progressive worsening dyspnea and cough beginning four days before. On chest radiograph was visualized a right total pneumothorax. Chest tube was placed which was removed after five days in view of lung expansion has been obtained. In this hospitalization performed a Computed Axial Tomography (CT) Thoracic that showed "on the right upper lobe in subpleural location, a densification pseudonodular with 16×13 mm". A transthoracic lung biopsy guided by CT was performed to study the lesion, which histological results showed that it was an adenocarcinoma of the lung. Clinical staging was consistent with surgical treatment. Was performed right upper lobectomy and adjuvant chemotherapy and is currently asymptomatic and in surveillance. Spontaneous pneumothorax associated with lung cancer is rarely seen in clinical practice. The hypothesis of tumor necrosis, or other unknown mechanism might be associated with the occurrence of pneumothorax may be plausible, particularly when there is concomitant pulmonary emphysema.

Keywords: *Pneumothorax. Pulmonary cancer. Adenocarcinoma.*

PRIMARY LYMPHOEPITHELIOMA LIKE CARCINOMA OF THE LUNG (LELC): TWO CLINICAL CASES IN CAUCASIAN PATIENTS

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Lymphoepitelioma like carcinoma of the lung (LELC) is a rare entity, generally seen in Asians, with very few cases described in the literature in Caucasian patients. The authors present two cases of lymphoepithelioma like carcinoma of the lung (LELC). The first is a 65 years old Caucasian female with a right inferior lobe mass found on a routine chest X-ray following abdominal complaint. The second case refers to a 51 Caucasian female patient who presented with a right lower lobe mass during clinical follow up after she had been submitted to a total tiroidectomy a year before. Both were submitted to surgery and the histology of the excised tumors demonstrated LELC with no associated Epstein-Barr virus activity. In both cases, metastatic LECL was ruled out. These two cases are according to the literature regarding Caucasian patients and compared with NSCLC seemed to have a better prognosis.

Keywords: *Lymphoepitelioma like carcinoma of the lung. Epstein-Barr virus. Caucasian patients.*

PULMONARY ADENOCARCINOMAS HAVE A LOW INCIDENCE OF ALK EXPRESSION AND FISH MAY BE THE ECONOMICAL APPLIED METHOD

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Crizotinib needs validation of EML4-ALK fusion gene at lower cost and a rapid answer from Anatomical Pathology. WHO 2004 lung carcinomas classification and CK7, TTF1, CK5.6, CD56/chromogranin and vimentin immunohistochemical panel as well as ALK (clone 5A4, Novocastra Laboratories Ltd, Newcastle, United Kingdom) were applied to paraffin sections of 35 bronchial-pulmonary carcinomas: 20 adenocarcinomas, 6 epidermoid carcinomas, 4 pleomorphic carcinomas (mixed type adenocarcinomas with large/giant/fusiform cells), 4 neuroendocrine carcinomas (NEC) (1 combined large cell NEC with adenocarcinoma and 2 with epidermoid carcinomas; 1 SCLC chromogranin positive combined with adenocarcinoma) and 1 adenosquamous carcinoma. Bronchial-pulmonary carcinomas subtypes were determined and also 3 cases of 60 years old non-smoking females with mixed type adenocarcinomas had ALK expression in more than 50% of cells: acinar, solid, micropapillary and microacinar patterns; one glandular mucinous pattern (mucinous BA pattern) and one BA pattern, all expressing TTF-1. As, 3/20 adenocarcinomas of older women had ALK protein expression, only one with a mucinous pattern, protein positivity cases showed to comprise a very low number and FISH, as described by S. Lantuejoul, seems to be the most appropriate method, it applied without searching protein expression. It is now necessary to decide whether *KRAS* and *EGFR* mutations have to be determined together and/or select TTF-1 positive adenocarcinomas (from terminal respiratory unit) as raised by this approach.

Keywords: *Bronchial-pulmonary carcinomas. ALK. FISH.*

BRONCHIAL-PULMONARY CARCINOMAS MAY BE SUBTYPED BY PET

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As bronchial-pulmonary carcinomas induced survival remains poor, between 6% and 14%/men and 7% to 18%/women and treatment orientation is influenced by clinical staging and morphological classification in biopsies of 70% of the cases, other complementary methods may preview prognosis as we try to explore through PET. This study comprised 41 surgical specimens: Adenocarcinomas (18), Epidermoid Carcinomas (12) and the heterogeneous groups of Large Cell Neuroendocrine Carcinoma (3), Small Cell Lung Carcinoma (1), Large Cell Carcinoma (2), Adenosquamous Carcinoma (2) and Pleomorphic Carcinomas (3) to compare maximum 18F-Fluordesoxiglucose (FDG), a clinical parameter based in PET, with immunohistochemistry to preview diagnosis and prognosis. There were significant differences (P=.006) between TTF-1 positive and negative Adenocarcinomas: 18F-FDG capture was lower in TTF-1 positive cases, indicating lower metabolic activity while TTF-1 negative Adenocarcinomas had similar and higher metabolic activity as Epidermoid Carcinomas; the other histological types showed FDG capture similar and between the two above defined

groups. PET 18F-FDG analysis may be a clinical parameter to establish differences between Adenocarcinomas and Epidermoid Carcinomas, where TTF-1 negative Adenocarcinomas are biologically similar to Epidermoid Carcinomas, requiring a different medical approach as well as molecular pathology particular interpretation. These results strain the recognition of bronchial TTF-1 negative Adenocarcinomas because they are different from the terminal respiratory unit TTF-1 positive Adenocarcinomas.

Keywords: *Bronchial-pulmonary carcinomas. Diagnostic. PET.*

MALIGNANT PLEURAL MESOTHELIOMA-CASE REPORT: THE TUMOR THAT APPEARS WHEN WE FORGET IT

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Introduction: Malignant pleural mesothelioma (MPM) is a universally fatal and aggressive tumour that leads to pleural effusion (PE). Because of long latency time and not being a frequent cause of PE, it can be forgotten in the etiological investigation, leading to inadequate procedures and delayed diagnosis.
Objective: Illustrate, based on a clinical case, the relevance of considering MPM as a cause of PE. Show a case submitted to multimodal therapy, with current survival of 20 months.
Case report: Patient with 51 years old, non-smoker, cook, with a history of hysterectomy for cervical tumour and chronic gastritis associated with H. pylori. In Jan/2011 she went to the emergency department for pleuritic chest pain since 4 days ago, dyspnoea on exertion, cough with yellow sputum, headache, runny nose, nasal congestion and fever. CBC and laboratorial chemistry were unchanged. Chest radiograph revealed an image compatible with PE. The patient underwent thoracentesis: pH 7.4, exudate. She was admitted in the ward; antibiotics and supportive measures were prescribed. Pleural blind biopsies were performed, but were complicated with pneumothorax, requiring placement of chest tube. In the meantime, we had the result of pleural fluid (PF) cytology of the 1st thoracentesis, showing adenocarcinoma but with non-defined origin by immunohistochemistry. Given the lung expansion and absence of PE, we proceeded to pleurodesis with talc slurry. The following day the patient begins fever, coughing, mucopurulent expectoration. New antibiotic was prescribed. We performed chest CT: mediastinal nodes; on the right side: loculated PE, pleural thickening, small pneumothorax and pulmonary condensation probably by compression. The patient underwent VATS for diagnosis and therapeutic. Biopsies showed pleural mesothelioma. Additional studies were performed: PET — uptake in right pleura, without other changes; cranial CT scan — no secondary lesions; physiological staging compatible with extrapleural pneumonectomy proposal, which was performed. Pathology: epithelioid mesothelioma, without invasion of lines of resection; without lymph nodes metastasis. Adjuvant RT was accomplished. After 2 months, the patient was revaluated with PET: absence of local recurrence, but multiple hepatic nodules and collarbone metastases. We started chemotherapy with carboplatin and pemetrexed. CT scan after 4 cycles showed partial response though with renal toxicity. Two additional cycles with pemetrexed were performed. With an exhaustive questioning it was found that the patient had worked decades ago in containers insulated with asbestos. The patient has a current survival of 20 months.
Conclusion: The methods used to study the aetiology of exudative PE are usually insufficient for the diagnosis of MPM. In the favourable context, they may even induce clinical error. Cytology of PE is particularly problematic. Given the natural history and epidemiology, MPM should always be considered as a possible cause of PE. Medical thoracoscopy should be a preferred method of investigation in doubtful cases. In well selected patients, multimodal therapy may be associated with good survival without significant morbidity.

Keywords: *Malignant pleural mesothelioma. Diagnosis.*

NODULAR SCLEROSIS HODGKIN LYMPHOMA — WHAT A CHALLENGE!

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Classical Hodgkin Lymphoma (HL) is a heterogeneous group of tumors characterized by scanty neoplastic diagnostic cells (Reed-Sternberg cells) in an inflammatory background. Nodular Sclerosis Hodgkin Lymphoma (NSHL) accounts for 70 percent of classical HL cases. In NSHL, areas of necrosis may be present, and diagnostic Reed Sternberg (RS) cells may be rare. The majority of classical HL patients present with painless localized peripheral lymphadenopathy, typically involving the cervical region. A 32-year-old man was admitted for 1-week history of cough with scanty purulent sputum and left pleuritic chest pain. Patient denied episodes of fever, weight loss or night sweats. Physical examination showed fever and no peripheral lymphadenopathies. Analytically with leukocytosis and elevation of C-reactive protein. Chest X-ray showed well defined opacity in the upper left hemithorax. Chest CT revealed a 9 cm bulky lesion occupying almost the entire left upper lobe, a 2.5cm node in the left lower lobe, two nodules in the upper and middle right lobes and multiple mediastinal adenopathies. Flexible and rigid bronchoscopies with biopsies were performed without conclusive results. Percutaneous needle biopsy and EBUS with transbronchial needle aspiration were inconclusive. The patient was submitted to surgical lung biopsy, which yielded the diagnosis of NSHL with pulmonary involvement. PET scan revealed supra-diaphragmatic disease, lymph nodes and bilateral lung disease. Bone marrow biopsy showed no involvement by the disease. The patient's advanced stage HL refers to a clinical stage IV A according to Cotswold Modification of Ann Arbor Staging System and an International Prognostic Score (IPS) risk of 3 (fair risk). Combination chemotherapy with Doxorubicin, Bleomycin, Vinblastine, and Dacarbazine (ABVD) was initiated. After 2 cycles of chemotherapy the patient repeats PET, which revealed partial regression of disease. The biopsies obtained by bronchoscopies were inconclusive probably because of scarcity of RS cells and the fact that fine needle aspiration alone only allows cytological evaluations and does not provide enough tissue or information about the structural composition. The patient was also submitted to large core needle biopsies usually preferred in these cases but without conclusive results as well. This case emphasizes the need for an excisional biopsy in order to obtain enough histological material to make a correct diagnosis. Atypical presentation and subsequent procedures which failed to yield results, including core needle biopsies, made this diagnosis a challenge, culminating in a surgical lung biopsy required to reach diagnosis.

Keywords: *Hodgkin lymphoma. Excisional biopsy. Staging.*

ENDOBRONCHIAL METASTASES-2 CLINICAL CASES

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Introduction: Endobronchial metastases from extrathoracic tumors are not frequent and generally seen in extensively disseminated disease. The most common primary malignancies associated are breast, colorectal and renal carcinomas.

Objective: to describe two clinical cases of colorectal cancer with endobronchial metastasis.
Results: *Case 1.* A 56 year-old men diagnosed with adenocarcinoma of rectum 6 years earlier that underwent neoadjuvant chemoradiotherapy followed by abdominoperineal rectum amputation and postoperative chemotherapy. Three years after the diagnosis he was found to have a metastatic lesion in the lung for which he underwent metastasectomy followed by chemotherapy. Six years after the initial diagnosis he was admitted for hemoptysis; investigation showed a mediastinal adenopathic conglomerate and an endobronchial lesion in the right main bronchi that was biopsied revealing its metastatic origin. Endobronchial treatment with laser and lesion excision was performed and chemotherapy was again started. At present, 10 months after the endobronchial metastases diagnosis, he is still under chemotherapy and is asymptomatic. *Case 2.* A 76 year-old men with prostate and colon cancer diagnosed 4 and 3 years earlier respectively. He underwent total colectomy and seven months after surgery hypercaptant nodules were found in both lungs suggesting pulmonary metastases. No adjuvant treatment was performed due to poor cardiac function. Three years after the initial diagnosis he was admitted for pleural effusion. The study revealed an endobronchial lesion in the left inferior lobar bronchi whose initial biopsies were inconclusive (necrosis). Rigid bronchoscopy was performed and the diagnosis of endobronchial metastasis of colon adenocarcinoma was established. He was referred to shared decision making consultation.
Comment: Endobronchial metastases from colorectal cancer are rare and occur more frequently when allied with pulmonary metastasis. A prolonged interval between the primary tumor and endobronchial metastasis is usual and its appearance is generally associated with a poor survival. These two cases emphasize the need to maintain this diagnosis in mind when dealing with patients with endobronchial lesions and a history of colo-rectal cancer.

Keywords: *Endobronchial lesion. Metastasis. Colorectal cancer.*

TYPE AB THYMOMA-CLINICAL CASE

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Introduction: Epithelial thymic tumours, which comprise the majority of thymomas, constitute around 30% of anterior mediastinal masses in adults. These are slow growth tumours and their prognosis depends on the macroscopic or microscopic invasion of adjacent structures. Surgery is the primary treatment for these tumours, with complete resection one of the most important factors of prognosis.
Case report: The authors present the case of a 66 year-old man, non-smoker, with a history of right nephrectomy because of a clear cell renal carcinoma, in August/2009. He was referenced to Pulmonology in November/2011 because of a mediastinal mass detected on follow-up CT scan. He had no respiratory symptoms or other complaints. CT scan revealed a mediastinal mass, in the pre-vascular space, lobulated, heterogeneous, with gross calcifications, with stable features and dimensions in consecutive CT scans (since November/2010), consistent with a benign lesion, advancing as more likely diagnostic hypothesis: teratoma or mediastinal lymphadenopathy secondary to metastasis of renal neoplasia. PET scan showed hypermetabolism in the lesion located in pre-vascular space, compatible with malignancy. Bronchofibroscopy revealed no significant alterations, with bronchial lavage cytology negative for malignancy. No image-guided biopsy was performed because of risk, given the location of the lesion. He was referred to the Thoracic Surgery for diagnosis/resection. He underwent sternotomy in March/2012-it was visualized a mass with invasion of brachiocephalic trunk, with endoluminal tumor, invasion of pericardial fat and phrenic nerve. It was performed excision of the tumor, resection of the venous brachiocephalic trunk, pericardial fat and left phrenic nerve. The pathological examination revealed a type AB thymoma with extensive vascular invasion. It was decided by the Department of Oncology to start chemotherapy, with cyclophosphamide, doxorubicin and cisplatin, and radiotherapy. With respect to respiratory function, the patient has a restrictive ventilatory syndrome, secondary to diaphragmatic hemiparesis, consequence of the need for surgical removal of the phrenic nerve. He transiently needed home oxygen therapy and is in a respiratory rehabilitation program.
Comments: The authors describe this case because of diagnostic hypotheses that gave rise, since radiologically the mediastinal mass was consistent with a benign lesion, but it was verified intraoperatively that it was a tumor mass invading noble structures-a type AB thymoma.

Keywords: *Anterior mediastinal mass. Differential diagnosis. Teratoma. Type AB thymoma.*

GIANT THYMOMA-CASE REPORT

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Introduction: The study of a mediastinal mass is a challenge, since its etiology may be caused by a multiplicity of pathologies.
Case report: The authors describe the case of a 53 years old female patient, ex-smoker, who went to the emergency department due to a pleuritic chest pain and non-productive cough with five days of evolution. Also reported weight loss (10 kg) with a year of evolution. The patient was subfebrile and has decreased breath sounds in the lower two-thirds of right hemithorax. Mild anemia, leukocytosis and increased C-reactive protein were observed. Chest x-ray revealed an opacity occupying the lower two-thirds of the right hemithorax. Diagnostic thoracentesis was done with drainage of serohematic fluid, lymphocytic predominance (65%), ADA and glucose normal values and a biochemical study compatible with exudate. Chest CT showed a mass with a diameter of 12×15 cm occupying most of the right hemithorax, in the continuity of the anterior wall, conditioning marked shift of the mediastinum to the left, shaping the intermediate bronchus, heart and lower vena cava; right pleural effusion; without hilar or mediastinal lymphadenopathy. The patient was admitted to the pulmonology department where was submitted to another thoracentesis and pleural biopsy whose study was negative for malignant cells, and immunophenotyping of pleural fluid revealed a T lymphocytes predominance. Transthoracic needle biopsy showed a polymorphic lymphoid and polymorphonuclear population; immunophenotypic study revealed an atypical lymphoid population. Because of the inconclusiveness of the study the patient was submitted to a surgical lung biopsy that showed changes that were consistent with thymoma. Thoracic MR was performed to evaluate the possibility of a resection of the thymoma, with no evidence of invasion of adjacent structures. The patient underwent surgical resection of the mass with 1257 grams which histopathology revealed a type AB thymoma, predominantly type B1 without macroscopic or microscopic capsular invasion (Masaoka stage I). Currently the patient is followed up in oncology outpatient clinic, with no indication for adjuvant treatment.
Conclusions: Thymomas account for about 20 percent of mediastinal neoplasms. Clinical manifestations mainly arise from compression of neighboring structures, the most common being chest pain, dyspnea and superior vena cava syndrome. Complete surgical resection is the main treatment, and the most significant

predictor of long-term survival. The majority of AB thymomas behave in a benign manner, being the five-year survival rate for Masaoka stage I tumour between 94-100%. Despite the large size of the tumor, this case has an excellent prognosis.

Keywords: *Mediastinal mass. Giant thymoma.*

INTESTINAL METASTASIS OF LUNG CARCINOMA: REPORT OF 3 CASES

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Introduction: Lung cancer often presents metastatic disease ant the other lung (50%), liver (37%), adrenal glands (31%), bones (29%), kidneys (18%) and brain (12%) are the organs most commonly involved. Intestinal metastases are rare (0.1-3%) and only about a hundred cases are described in the literature. It is important to consider the differential diagnosis with primary neoplasm of the intestinal tract. However, the prevalence may be greater, based on autopsy data, which identified several asymptomatic cases. Clinically, intestinal metastasis can manifest as bowel perforation (35.9%), stenosis (27%), intussusceptions (21.8%) or bleeding (15.4%). The prognosis of lung cancer patients with intestinal metastasis is poor, with a low survival (3 weeks-4 months). The treatment is surgical, and may result in a significant increase of survival.
Aim/methods: The authors describe 3 clinical cases of patients with carcinoma of the lung and intestinal metastasis.
Results: *Case 1.* A 49-year-old woman with a history of active smoking. She was diagnosed with adenocarcinoma of the lung (stage IV) in 2009 and received chemotherapy. Due to disease progression, with increasing liver and bone metastasis she underwent four lines of chemotherapy, providing stable disease. In December 2010 has intestinal occlusion by intraluminal mass with intestinal perforation and peritonitis. The tumor was ressected and diagnosed as a metastasis of lung cancer. The survival after enterectomy was 2 months. *Case 2.* A 75-year-old man with history of smoking in the past. He was diagnosed with adenocarcinoma of the lung (stage IIb) in 2005. The patient underwent right upper lobectomy and adjuvant chemotherapy, with disease remission. In November 2011, presented asthenia, weight loss and anemia. In imaging exams, he had a mass in right upper lobe and thickening at the level of the jejune. Subsequently, he developed intestinal obstruction, requiring tumor and segmental bowel resection. Histological examination of the surgical specimen showed metastasis of lung carcinoma. The patient died 2 months after enterectomy. *Case 3.* A 57-year-old man with a history of active smoking. In October 2011, he was diagnosed with non-small cell carcinoma of the lung in stage IV and had 4 cycles of first line chemotherapy. In January 2012, the patient presented abdominal pain, nauseas and vomiting associated with food intolerance and weight loss. Abdominal CT and ultrasound showed jejune intussusception. We proceeded to tumorectomy and segmental bowel resection. The histology was compatible with metastatic carcinoma of the lung. The survival after enterectomy was 2 weeks.
Conclusion: With the increased survival of patients with lung cancer, it is necessary to take into account the possibility of occurrence of metastasis in distant places least prevalent. The intestinal metastasis from lung cancer is rare, but should be considered in patients with suggestive symptoms. Surgical treatment, although palliative, can result in a significant increase in survival in some patients, however it was not observed in any of the cases described.

Keywords: *Lung cancer. Intestinal metastasis.*

PULMONARY ADENOID CYSTIC CARCINOMA, A CASE REPORT

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Background: Adenoid cystic carcinomas are a differentiated malignant epithelial neoplasm, more common in the salivary glands, but can also be found in the lung. However, its incidence in the lung is very rare. In lung, they usually occur between the 4nd and 6th decade of life, with no gender predominance, classically have slow growing, with indolent clinical course, and low malignant potential. However they can have a more aggressive course, with tendency for local recurrence and metastatic potential.
Case report: The authors present a case of a 60 years old man, admitted through the emergency room with prolonged community-acquired pneumonia and hemoptysis. Chest radiography showed an opacification of the left lower lobe of the lung, with ipsilateral mediastinal shift and patient had respiratory failure, which led to his admition. Complementary study showed, a mass (4.5x4cm) causing collapse of left lower lobe in the CT of the chest. The bronchoscopy confirmed the presence of an endobronchial lesion whose bronchial biopsies were positive for adenoid cystic carcinoma. The PET-scan showed a single left hilar lesion suggestive of neoplasm, with no signs of metastasis. The case was discussed in oncological pulmonology multidisciplinary meeting, and patient was accepted for thoracic radiotherapy sessions with curative intent (60Gy), already concluded. Currently the patient is asymptomatic, without poor prognostic factors, in surveillance at intervals of 3 months.
Conclusion: Adenoid cystic carcinoma of the lung, although rare, is an entity that should be considered in those patients with more indolent clinical course. Although the usually surgical treatment, thoracic radiotherapy is a viable treatment alternative.

Keywords: *Carcinoma. Adenoid. Cystic. Lung.*

LATE METASTASIS

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Introduction: Uterine leiomyosarcoma are rare, accounting for 1% of all malignant uterine neoplasm. These tumors are characterized by a very poor prognosis, with a high rate of local recurrence as well as a high rate of recurrent metastases. Lung metastasis is the second location most frequent, after peritoneal cavity. The familiar appearance of such disease is characterized by a large number of parenchymal nodules, but unilateral location of one mass does not rule out metastatic lung disease.
Aim/methods: Because the long disease free-interval, the authors report the case of a patient hysterectomized by a leiomyosarcoma, that 26 years after presented lung metastasis.
Results: It's a 77 year old female with no history of smoking. The past history revealed a hystero-oophorectomy for uterine sarcoma about 26 years ago. She complained of cough and dyspnea over the past 2 weeks. The patient was admitted to the emergency department for worsening of symptoms. Her general condition was good, but she had reduced lung sounds in the lower half of the left hemithorax. The PaO_2 in room air was 73 mmHg. Chest X-ray demonstrated a nodular image of greater density in the lower left hemithorax. She was admitted for further diagnostic

studies in the Pneumology department. Thoracic CT showed an opacity with 6.7 x4, 8cm in the left lower lobe, with relatively well-defined limits. The transthoracic pulmonary biopsy revealed a malignant tumor with mesenchymal origin, but conclusive diagnosis was not possible, because the sample was insufficient. The mass was subsequently removed by left lower lobectomy In the surgical specimen was identified a solid and spheroid mass, with 8 cm and with multiple foci of necrosis. Histological examination of tumor samples allowed the diagnosis of a pleomorphic leiomyosarcoma (compatible with uterine leiomyosarcoma metastasis). There was no infiltration in the mediastinal nodes and the other complementary exams didn't demonstrate evidence of tumor elsewhere on the body.

Conclusion: The lung is a frequent site of hematogeneous metastasis of many diverse tumors including soft-tissue sarcoma. In our case the pulmonary metastasis manifested past 26 years, the longest disease-free interval wee known (after review of literature in the English language). The disease-free interval for this case was 26 years, which was, to our knowledge (through literature review) the longest among previous reports of pulmonary metastasis of uterine leimomyosarcoma. This case suggests that the long term follow-up is necessary in the case of uterine leiomyossarcoma after operation. Pulmonary metastasectomy is the only treatment modality that can provide a care for the patients.

Keywords: *Uterine leimomyosarcoma. Pulmonary metastasis. Disease-free interval.*

VINORELBINE IN NON-SMALL CELL LUNG CANCER: SUCCESSFUL CLINICAL CASE

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Introduction: The use of vinorelbine is approved as therapy for non-small cell lung cancer (NSCLC), as combination or monotherapy. However, studies of its use as salvage therapy showed a poor response rate.

Case report: The authors describe a case of a 45-year old man, ex-smoker, pool technician, with no history of previous diseases. He was diagnosed in November 2008 with NSCLC stage IIIA (mass with 5x6 cm in RUL, with invasion of the first three ribs and right hilar lymphadenopathy with uptake in PET-CT). Evaluated by a thoracic surgeon and was refused to surgical treatment. He started a scheme of concomitant chemoradiotherapy with carboplatin and gemcitabine. Because of local progression in June 2009, a second QT line with docetaxel was proposed, which caused however a serious anaphylactic reaction. Further treatments included pemetrexed for 3 months and erlotinib afterwards. In October 2009 develops jaundice due to an adenocarcinoma of the periampular region (collocation of stent by CPRE). Because of recurrent progression of lung cancer, initiated a 4th line scheme with oral vinorelbine in December 2009. In the first 6 months we observed a partial response with significant reduction of the lung mass size and no evidence of distant metastasis. Treatment was maintained with vinorelbine for a 22 month period (completed 30 cycles) with stable disease, good hematological tolerance and excellent performance status. In August 2011 local progression was observed. Since this date, he had progressive disease despite new chemotherapy schemes. Currently he presents zubrod 3, with significant increase of lung mass dimensions that causes obstruction of right main bronchus conditioning atelectasis and frequent episodes of hemoptysis.

Conclusion: This clinical case highlights the use of vinorelbine as a rescue treatment option in NSCLC that, in particular situations, may have a favorable response with good tolerance.

Keywords: *Vinorelbine. Non-small cell lung cancer.*

PRIMARY PULMONARY LYMPHOMA OF MALT TYPE: AN UNUSUAL PRESENTATION

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Primary pulmonary lymphoma (PPL) is very rare, represent only 0.5 to 1% of primary pulmonary malignancies. Bronchus-associated Lymphoid Tissue (BALT) lymphoma B, a extranodal marginal lymphoma, is the most common type of PPL. Considered to originate from mucosa-associated lymphoid tissue (MALT) of the bronchus, BALT lymphoma comprises more than two-thirds of all primary non-Hodgkin's lymphoma (NHLs) of the lung and about 1% of all NHLs. We describe the case of a 67-year-old man, a nonsmoker, retired metalworker, with no relevant previous diseases, presented with a 3-month history of fatigue, weight loss and dyspnea on effort. Chest radiograph showed *left*-sided *pleural effusion*. Admitted at a pneumology ward for study. Pleural fluid and biopsy presented unspecific inflammation characteristics. A CT scan revealed left lower lobe diffuse bronchovascular densification, with large volume pleural effusion associated and multiple bilateral supra and infra centrimetric pulmonary nodules, predominantly located at lower and middle lobes, yet coexisting small pericardial effusion and multiple axillary, supraclavicular, mediastinal and abdominal adenopathies, as well as enlarged spleen with homogeneous round morphology. Plasma immunoglobulins and ß2-microglobulin were increased. Bronchoalveolar lavage was innocent and transbronchial biopsy histological examination revealed a chronic inflammatory infiltrate with moderate activity and predominance of T-lymphocytes. Transthoracic aspiration biopsy and axillary lymph node surgical biopsy were inconclusive. Diagnosis was made by axillary lymph node dissection, which morphological and immunohistological study revealed marginal zone B-cell lymphoma of MALT-type. The patient was oriented to Hematology consultation for further study and treatment. Other primary tumor localizations were excluded and bone marrow biopsy showed no cytogenetic alterations. Chemotherapy was started with a favorable response. PPL MALT type affects both sexes equally. Age of onset is approximately 50 to 70 years. Presents an indolent course and is associated with chronic inflammatory lung or systemic diseases. Nearly half of the patients are asymptomatic at diagnosis. B symptoms are uncommon (<25%). Major radiographic abnormalities represented either lung nodules or airspace consolidation with or without air bronchogram. Pleural effusion (<10%) or adenopathies are unusual and pericardial effusion is very rare. The diagnosis is based on histological examination of lung biopsy material. The survival outcome is excellent with different treatment modalities. It is therefore a case of BALT lymphoma with atypical clinical and radiological presentation, with polyserositis and generalized lymphadenopathy. Diagnosis can be difficult due to nonspecific cellular features of the samples obtained, often interpreted as an infectious or chronic inflammatory infiltrate in cases of low-grade. Clinical suspicion is very important as well as persistent diagnosis research.

Keywords: *Primary pulmonary lymphoma. Lymphoma of MALT type.*

HODGKIN LYMPHOMA PRESENTING AS A PULMONARY CAVITATED MASS

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Lung cavitated lesions have a wide differential diagnosis which includes various infectious diseases (gram-negative bacteria, anaerobic bacteria, mycobacteria, and fungal agents: actinomycosis, histoplasmosis, aspergilosis), neoplastic diseases, granulomatous

diseases (Wegener's Granulomatosis) and others (pneumoconiosis, silicosis etc). The pulmonary involvement in lymphoproliferative diseases and in particular Hodgkin's disease can occur in 15 to 40% of cases and is usually associated with direct invasion by lymphoid follicles or peribronchiolar adenopathies adjacent to the lung parenchyma. The primary pulmonary Hodgkin's Lymphoma is a rare disease and less than 100 cases have been reported in literature. To establish the diagnosis of primary Pulmonary Hodgkin's Lymphoma the following criteria must be met: a histological result compatible with Hodgkin's disease must be found, and the disease must be confined to the lung with or without minimal hilar lymph node involvement at the time of diagnosis and at 3 months follow-up. We report a case of a Caucasian female patient, 24 years old, non-smoker, presenting with asthenia, pruritus, and night sweats for 6 months and a non-productive cough, left chest pain and fever. She was admitted to the hospital after identification in plain radiograph of a cavitated lesion and was transferred to our Hospital with a presumptive diagnosis of Pulmonary Tuberculosis. She was submitted to various bronchoscopies, but all results were negative for infectious agents and biopsies only revealed a chronic inflammatory process. Initially the thoracic CT scan only revealed a large lung cavitated lesion. The patient was placed on anti-tuberculous therapy empirically. However, she became worse clinically and the disease progressed with large mediastinum and supraclavicular lymph node involvement, and pleural effusion after 4 weeks. A mediastinum lymph node biopsy confirmed the diagnosis of mixed cellularity Hodgkin's Lymphoma. Although this case does not fulfill all diagnostic criteria for the diagnosis of primary Hodgkins lymphoma due to a rapid lymph node evolvement during hospital stay, this is an interesting case of lung involvement by Hodgkin's lymphoma and reminds the importance of considering this as a differential diagnosis for lung cavitated lesions.

Keywords: *Hodgkins lymphoma. Lymphoproliferative diseases of the lung. Pulmonary cavitation. Neoplasms of the lung. Differential diagnosis of pulmonary cavitation.*

X RADIATION EFFECTS IN SMALL CELL LUNG CANCER-STUDY IN CELL CULTURE

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Introduction: Small cell lung cancer (SCLC) represents 15% to 20% of all cancers originated in the lung, being a major cause of death from this disease and the most aggressive. SCLC has increased is prevalence in the world population being the tobacco the primary cause, which not only enhances the appearance of the disease but induces also resistance to chemotherapy. The diagnosis is done in most cases in advanced stages with metastases, which complicates the treatment and the selection of the therapeutic protocol. The main treatment usually is surgery followed by chemotherapy and/or radiotherapy. Although good results are obtained in an early stage during treatment in response to chemotherapy and/or radiotherapy, in most cases disease recurrence happens. Mutations in genes that encode proteins involved in the regulation of apoptosis as p53, a tumor suppressor protein, generally activated after DNA damage, it may induce cell cycle blockage. Loss of p53 function could lead to increased growth rate and decreased activation of apoptotic pathways. The radiation induces direct damage (DNA, RNA, proteins and lipids) or indirect mainly through the radiolysis of water yielding reactive oxygen species that can damage macromolecules and alter cell signaling. Also induces changes in the properties of cell membranes, causing changes in their permeability and eventual lysis. Moreover, the radiation can induce activation of p53 by blocking the cell cycle, predominantly at the checkpoint G1/S.
Aim: Evaluate the effects of radiation X (RX) in viability and proliferation, as well as characterize death and cell cycle in H69 SCLC cells.
Material and methods: We evaluated cell proliferation and viability of H69 cells (ATCC; p53 Mut, 171:GAG→TAG) in the absence (control) and after exposure to RX 4 MeV with different doses (0.5Gy, 15Gy and 30Gy), every 24 hours for a period of 96 hours. Death and cell cycle were analyzed 48h after irradiation by flow cytometry using double staining with annexinV/propidium iodide (PI) and PI/RNase, respectively.
Results and conclusion: Results suggests that RX effect on cell line H69 is dose and time dependent. Exposure to 15Gy and 30Gy induces decreased cell proliferation, inducing apoptosis and cell death preferably by blocking cell cycle in S phase. This can be explained by the loss of constraint point controlled by p53, which is mutated in this cell line.

Keywords: *Small cell lung cancer. X radiation. Cell cycle.*

PLEURAL EFFUSION AS GASTRIC ADENOCARCINOMA PRESENTATION

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We describe the case of a 51 year-old Caucasian male patient, smoker, who presented to the emergency room with a dry cough, dyspnea, asthenia and anorexia with a weight loss of 7 Kg in the last month. On physical examination the patient was evidently emaciated and there was a marked decrease on breath sounds in the lower third of both hemithorax. The arterial blood gas showed a partial respiratory failure with a PaO_2 of 50 mmHg. Laboratory data showed moderate leukocytosis with CRP elevation and the chest x-ray revealed a heterogeneous hypotransparency at the lower third of both pulmonary fields, obliteration of costophrenic angle and an apparent bilateral Ellis-Damoiseau line. The patient was admitted under the diagnose of community-acquired pneumonia, complicated by bilateral effusion, and started empirical antibiotic therapy. The clinical course was not favorable, with progression to global respiratory failure and ventilatory exhaustion, being thus transferred to the intensive care unit. The thoracentesis revealed a serous-hematic pleural fluid, consistent with an exudate, negative for malignant cells. The placement of bilateral chest drains resulted on massive left hemothorax. The urgent thoracotomy preformed showed an extensive pleural nodularity, histologically compatible with adenocarcinoma, with immunohistochemistry suggestive of primary gastric tumor metastasis. The endoscopy showed an irregular elevated lesion in the distal esophageal and gastric mucosa consistent with an infiltrative lesion. The biopsies confirmed the diagnosis of gastric adenocarcinoma. The patient died a few days later. Pleural effusion is rarely the primary manifestation of a gastric adenocarcinoma. This metastatic pattern seems to result from the migration of malignant cells through submucosal lymphatic vessels and appears more often in signet ring cell carcinoma. To our best knowledge, the only published series on literature comprises only 4 cases.

Keywords: *Pleural effusion. Gastric adenocarcinoma.*

THYROID AND COLON METASTASIS FROM LUNG CANCER: A CASE REPORT

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Brain, liver, adrenal glands and bone are the most common sites of metastatic disease in patients with lung cancer. The clinical prevalence of symptomatic gastrointestinal metastasis of cancer is only 0.2 to 0.5%. Therefore a number of clinical cases of small bowel metastasis have been reported, while clinical cases of colonic metastases have so far only rarely been reported. Although richly vascularized, the thyroid is an infrequent site for lung metastasis. We report a case of a 74-year-old female, non-smoker. The patient was diagnosed with adenocarcinoma of the lung (EGFR+, Stage IV with brain metastasis) four months before the admission and treated with radiosurgery Gamma Knife and oral corticotherapy. She presented with a history of melenas in the last two months and severe anemia was observed at the admission. Upper gastrointestinal endoscopy demonstrated no lesions however the colonoscopy showed a pseudo-polypoid mass in the hepatic angle of the colon. The mass was biopsied and the immunohistological findings suggested to be colon metastasis from the primary lung carcinoma. At the time of the admission a thyroid enlargement was noted and fine needle aspiration biopsy was performed and the findings were compatible with lung origin. Because the life expectancy of patients with lung cancer is longer than in the past we should pay attention to the clinical manifestations of metastasis in atypical places.

***Keywords**: Colonic metastasis. Thyroid metastasis. Primary lung cancer.*

BRONCHOGENIC CYST, A. CLINICAL CASE

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Bronchogenic cysts are rare congenital lesions that arise from abnormal budding of the primitive tracheobronchial tube from the embryonic foregut, between the 5th and 7th weeks of gestation. They can be located in the mediastinum, along the traqueobronchial tree when the budding occurs during early development or located within the lung parenchyma (15% to 20% of cases), when they arise later during gestation. They are lined by columnar ciliated epithelium and their walls often contain focal areas of hyaline cartilage, smooth muscle, and bronchial glands which confirm the diagnosis. The authors present a case of a 68-year old female, retired (secondary school teacher), and a former smoker with a previous hysterectomy due to endometrial adenocarcinoma. She was referred to Pulmonology in 2003 for asthenia, anorexia, weight loss and dyspnea on moderate exertion. Her medical examination was normal. Exams performed showed opacity in the right lung with 4, 8 cm diameter and no abnormalities in blood tests or in bronchoscopy. The patient was proposed for diagnostic and therapeutic surgery and underwent atypical resection of the superior right lobe. Two hemorrhagic cystic areas were found and were described histopathologically as an organizing pneumonia with xantomatous inflammatory infiltrate (lipidic pneumonia/ focally organized pneumonia). She refused further investigation and, as she was asymptomatic, was maintained in follow-up. Although an irregular opacity with fibrous retraction persisted on her thoracic scan, it was considered surgical sequelae. In April 2012 she complained of fever, chills, dry cough and sore throat and the lesion increased (from 30 to 43mm) and had a hypermetabolic profile in PET evaluation. Lung cancer was suspected and fiberoptic bronchoscopy was performed that showed diffuse inflammatory signs. Bronchial biopsies showed normal bronchial mucosa and cytology was negative for cancer cells. She underwent another surgery with resection of the lesion whose anatomic-pathological evaluation revealed a cyst with purulent content, lined with bronchial epithelium and cartilage within its wall, compatible with bronchogenic cyst. The patient had no postoperative complications, and is currently under surveillance and asymptomatic.

Discussion: Mostly described in the pediatric population, bronchogenic cysts may cause symptoms by compressing nearby structures (dyspnea, dysphagia, cough, pain or mimic pulmonary infections). In adults, cysts are often asymptomatic and diagnosed during the patient's chest evaluation. In this case the diagnosis was made only after the cyst became symptomatic since the CT lesion was thought to be secondary to surgery. This evolution supports some authors' opinion that in most cases, symptoms will arise if there is no treatment and recommend surgical removal, even in the absence of complaints, given the likelihood of symptoms and complications.

***Keywords**: Pulmonary mass. Bronchogenic cyst. Treatment.*

A STRANGE CASE OF RECURRENT WHEEZING

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Introduction: The differential diagnosis of recurrent wheezing, especially in adulthood, is usually confined to asthma and other obstructive airway diseases such as COPD. The correct assessment of symptoms, medical observation and judicious use of diagnostic exams can prevent error or misdiagnosis with other pathologies, including neoplastic disease.

Results: We describe a 21 years old male patient, non-smoker, with a history of allergic rhinitis and recurrent wheezing in the 1st childhood, who presented with repeated episodes of breathlessness, dry cough and wheezing, predominantly nocturnal, which motivate multiple trips to the emergency department. In one of these trips, he also reported pleuritic chest pain. At observation, he was hemodynamically stable, with tachypnea and signs of respiratory distress, subcutaneous crackles at the cervical region and bronchospasm. During the diagnosis algorithm a chest CT was performed, that showed pneumomediastinum, subcutaneous emphysema, a pediculated image on the distal trachea-mucous impaction vs exophytic lesion-and parenchymal infiltratates suggesting a bilateral pattern of bronchogenic spread-hemorrhage vs infection. Due to clinical worsening, he was referred to our hospital, where he performed rigid bronchoscopy, which demonstrated, 2cm proximal from the carina, a decrease of 90% of the trachea's lumen by a multilobulated partially pediculated tumor. The patient underwent therapy with YAG laser (40 watts, continuous mode, with a total power of 8664 watts), with partial improvement and patency of the trachea and a HOOD 16/50 tracheal prosthesis was placed. The histological diagnosis was low grade muco-epidermoid tumor of the trachea. After multidisciplinary discussion, the patient underwent segmental tracheal resection with top-to-top anastomosis by thoracotomy, uneventfully. The revaluation fiberoptic bronchoscopies showed good healing and patency of the trachea. The edges of the surgical piece were free of malignancy.

Conclusion: The correct and timely referral to a tertiary hospital allowed the combined treatment by rigid bronchoscopy and conventional surgery for a rare tracheal cancer. Also of note is the need for correct categorization of patients with "wheezing",

especially in the presence of atypical symptoms, given the ensuing diagnostic implications.

Keywords: *Muco-epidermoid tumor of the trachea. Bronchoscopy. YAG laser. Thoracotomy.*

GERMINATIVE CELL TUMORS SUPPORT TUMORAL CLASSIFICATION THROUGH CELL LINEAGES: IMMUNOHISTOCHEMICAL STUDY

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The morphology of germ cell tumors (GCT) may correlate with embryogenesis, organogenesis and tissue maturation. Studies that support carcionogenesis based in cellular differentiation may be explored in germinal cell tumors to understand cell lineages and neoplastic development. New classifications according with a specific imunohistochemical panel would identify pluripotent stem cells, adult stem cells and preview morphology. The following antibodies AE1/AE3, CK7 and LP34 (cellular maturation), CDX2, TTF1 and PLAP (organogenesis) and Oct3/4, Nanog and Vimentin (embryogenesis) were applied to 34 benign and malignant formalin-fixed paraffin-embedded GCT, concerning 17 males and 17 females (age range 16-73 years).The immunohistochemical results were scored semi-quantitatively by the percentage of positive cells as 0% (0), <10% (1+),10-50% (2+) and >50% (3+) considering cytoplasmic or nuclei expression/positivity. Oct3/4 expression in nuclei of seminomas cells is different from embrionary carcinoma cells that expressed either nuclear and cytoplasmic positivity; Nanog gene expression was seen only in 2 cases concerning non-differentiated mesenchyme. Vimentin came out as a particular antibody in between Oct3/4 expression and CDX2/TTF1 cellular maturation, indicating its value in the transition between organogenesis and adult stem cells maturation. Intestinal differentiation revealed by CDX2 expression and TTF1 sensitive pulmonary alveolar epithelium were consistently absent in all epidermoid cysts of the ovary. PLAP glycoprotein showed to have positive expression in the majority of embryonic carcinomas and also identified smooth muscle cells. Cytokeratin AE1/AE3 was not discriminatory because it was expressed in all cases and corresponded to a non-specific epithelial marker. We may stress that the cellular maturation spectrum seen since embryogenesis till adult stem cells in GCT is similar to malignant cell lineage development in carcinogenesis, according with the actual grades of differentiation, and also validates epithelial-mesenchimal transition observed in less differentiated tumors/ carcinomas in all organs, including bronchial-pulmonary carcinomas.

Keywords: *Tumoral classification. Germ cell tumors. Immunohistochemistry.*

BRONCHOPULMONARY CARCINOID TUMORS: A CASE OF A TC WITH N1 GANGLIONAR METASTASIS. REVIEW OF THE LITERATURE

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Carcinoid tumors (CT) are of neuroendocrine origin and around 25% are bronchopulmonary. Despite their indolent behavior they're considered malignant tumors and classified as typical (TC) and atypical (AC) according to histological findings. According to literature, AC demonstrate a more aggressive clinical course and are known for a higher metastatic ability. For any given T stage and among central carcinoids, 90% of N0 are TC and 40% either N1 or N2 are AC at presentation. This metastatic potential is reflected by 10-year survival rates for TC and AC (88% vs 49%), and death of recurrent disease being more frequent in AC patients (71% vs 16%). Surgery is the gold standard while chemotherapy has revealed a low response rate.

Case report: 21 years old male patient, admitted for moderate hemoptysis. The patient was diagnosed an atypical lung carcinoid tumor by bronchial biopsy. CT scan showed areas of atelectasis/ condensation of the upper left lobe and a partially occlusive endobronchial mass located on the main left bronchus (MLB). Surgery: After bronchotomy we found the tumor arising from the LLLB. Lower left lobectomy was performed with bronchoplasty after clear bronchial margin achievement. Lymphadenectomy was also performed. Pathological analysis confirmed the diagnosis and showed an N1 lymph node metastasis. The histopathological differential diagnosis of TC and AC is difficult justifying distinct survival rates and clinical behavior reports for CT. Multicentric studies should be performed in order to establish Portuguese reality.

Keywords: *Carcinoids. Metastasis.*

AN EXCEPTION TO SILHOUETTE SIGN. CLINICAL CASE REPORT

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The authors present the case of a 28-year-old woman, smoker since the age of 21 (about 7 cigarettes per day), with chronic sinusitis without other known diseases, including respiratory diseases. The patient was observed in the emergency department for a complaining of cough with mucopurulent sputum, dyspnea, myalgia, pleuritic chest pain and fever (39 ºC). The observation of the chest showed pectus excavatum and pulmonary auscultation showed symmetrical breath sounds, with no adventitious sounds. Blood sample results showed leukocytosis and elevated CRP. Chest radiography showed homogeneous opacity in the right lower third, with cardiac's silhouette sign. Arterial blood gases (21%)-pH 7.42, pCO_2 29.6, pO_2 72.9, HCO3 20.9, Sat 95.1%. A diagnosis of community acquired pneumonia was made, and sent home with amoxicillin/ clavulanic Acid and Azithromycin. She was observed after course of antibiotic treatments, presenting with clinical and analytical improvement, however without radiological improvement, reason why she was sent to the pneumology consultation. During the investigation chest CT showed: subsegmental atelectasis with slight linear densification and cylindrical bronchiectasis in the left lower lobe; bilateral breast prostheses; without consolidation in the right hemithorax. Currently the patient remains in follow-up in Pneumology to study and treat the bronchiectasis. Breast implants have been available since 1960, and its prevalence has grown, especially in the last decade, cosmetic reasons and for breast reconstruction after mastectomy for cancer. Being a new interface between the lung and the chest wall, the prosthesis may interfere with standard chest radiography image, rendering its interpretation in to a clinical challenge. This case illustrates the importance of obtaining a complete medical history and physical examination, as well as obtaining posterio-anterior and profile chest radiography whenever possible. Thus, knowledge of the presence of breast implants in this patient, could help in the clinical interpretation and placing of differential diagnoses.

Keywords: *Breast implants. CAP. Bronchiectasis.*

FOREIGN BODY ASPIRATION IN THE ADULT

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The foreign body aspiration (FBA) is a serious and potentially fatal accident that can occur at any stage of life, but it's much more common in children. Early diagnosis of FBA is essential, since any delay in its recognition and treatment can result in permanent squeal or fatal damage. The presentation may be characteristic of FBA or be nonspecific with no signs on the physical examination which requires a high degree of suspicion to avoid the delay. The authors present the case of a 27 year old man, appealing to the emergency department (ED) after a dental procedure. At ED referring slight discomfort at the oropharynx, dry cough, dyspnea, dysphagia or pain. Hemodynamically stable, saturation, pulmonary auscultation and exam of oropharynx were normal. In this context the patient was subjected to posterior anterior chest radiography (PA) which revealed the presence of a foreign body apparently located in the esophageal high level. Later was discharged from ED with indication to return if worsening of symptoms. After two days with the same discomfort at chest, aggravated by coughing, the patient returned to the ED where he performed a new chest radiograph (PA), similar to the previous one. There were no changes in the objective test. We performed endoscopy (EGD) that did not reveal the presence of a foreign body, putting the chance of it being located in the trachea. We conducted a new chest radiograph (PA and lateral) after endoscopy showed that the progression of the foreign body to the Left Main Bronchus (LMB) that was submitted by the Fibrotic bronchoscopy. It removed the foreign body (dental drill) out of the lingula. In this case and since it was a foreign body with extremely sharp tip the patient was admitted and started antibiotic treatment with piperacillin and tazobactam to prevent possible mediastinitis. After 5 days of treatment and since no analytical and clinical abnormalities were shown the patient was discharged and forwarded to Pulmonology.

***Keywords**: Foreign body. Bronchoscopy. Mediastinitis.*

SPONTANEOUS PNEUMOTHORAX AND UNILATERAL HYPERINFLATION: AN UNPRECEDENTED CAUSE

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Introduction: The association of spontaneous pneumothorax and lung cancer is rare and its pathophysiological mechanisms are not clear.

Case report: We report the case of a female patient, 30 years of age, with no history of smoking, which presented with complaints of recent left chest pain and a history of cough over the past 2 months. Chest x-ray showed a left side pneumothorax that was resolved by insertion of an intercostal tube. Two months later she returned with new onset hemoptysis. Chest computed tomography revealed a small left side pneumothorax and also, increased left lung volume, contralateral mediastinum shift and increased right lung attenuation. These findings were more notorious with expiration and maintained after resolution of the pneumothorax. Furthermore, a lesion was visible in the left main bronchus, which bronchoscopy showed to be typical carcinoid tumor. An octreotide scan revealed uptake solely in the lesion of the left main bronchus and surgery was performed with excision of the left main bronchus and lobectomy of left upper lobe. Pathology confirmed a typical carcinoid tumor and showed no signs of emphysema or pleural blebs. One year after surgery the patient is asymptomatic, had no new pneumothorax and her chest computed tomography shows resolution of the mediastinum shift and of the differences in the lungs attenuation.

Discussion: Spontaneous pneumothorax as a complication of lung cancer is rare, comprising only 0.05 to 1.4% of all pneumothoraxes and occurring in just 0.05 to 0.46% of the patients with pulmonary malignancy. Moreover, to our knowledge, there was no previous description of a bronchial carcinoid tumor presenting, or being complicated, with pneumothorax. The proposed mechanisms for pneumothorax in cancer patients have been the formation of a bronchopleural fistula secondary to pleural invasion or necrosis and, less frequently, the rupture of alveoli, emphysematous bullae or subpleural blebs due to hyperinflation of the peripheral airways caused by the tumor acting as a check-valve. In the presented case, the tumor in the left main bronchus, functioned as a variable obstruction leading to the entrapment of air and unilateral left hyperinflation, which associated with expiratory maneuvers caused an increase in alveolar pressure with eventual rupture of alveoli and pneumothorax. This is a very rare case, with no previous description, of a bronchial carcinoid tumor presenting with a recurrent spontaneous pneumothorax and unilateral hyperinflation explained by a large check-valve mechanism.

***Keywords**: Pneumothorax. Lung neoplasms. Carcinoid tumor.*

WHEN INVASIVE VENTILATION FAILS

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Extracorporeal membrane oxigenation is a prolongued cardio-respiratory support procedure, wich intends to help the failing lungs and/or heart unresponsive to optimized conventional measures. The authors describe a clinical case of a 42 year-old non-smoker male, police officer, previously healthy, admitted to hospital for productive cough, dyspnoea and vomiting for the previous 4 days. He had been medicated with amoxicillin and clavulanic acid with no response. On physical examination he showed increased respiratory rate and prolonged expiratory time. The blood tests performed showed elevation of positive acute phase parameters, negative HIV 1/ 2. Influenza H1N1, blood and sputum cultures were also negative. Blood gases analysis revealed hypoxemia. The chest X-ray showed bilateral interstitial pulmonary infiltrates. Empirical treatment with ceftriaxone and clarithromycin was started with clinical worsening and need for invasive mechanical ventilation. He was admitted to an ICU. Even though his clinical condition continued to deteriorate, reason why he was transferred to another ICU for extracorporeal membrane oxygenation (ECMO). He developed peripheral eosinophilia and had elevated eosinophil counts in bronchoalveolar lavage. The diagnostic hypothesis of eosinophilic pneumonia or hypereosinophilic syndrome were considered and methylprednisolone started with a favorable response. Several complications related to ECMO technique were managed successfully and tracheostomy was performed due to a difficult weaning. He returned to our hospital ICU and after to the Pulmonology Department were he started a rehabilitation program with progressive improvement and tracheostomy closure.

***Keywords**: Extracorporeal membrane oxygenation. Invasive mechanical ventilation. Eosinophilic pneumonia.*

MOUNIER-KUHN SYNDROME-2 CLINICAL CASES

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Introduction: TheMounier-Kunh (MK) syndrome or tracheo-bronchomegaly is a rare clinical and radiologicalentity. It is thought to be related to congenital deficiency of muscular and elasctc layer of the trachea and main bronchi, leading to its dilation and even

to the formation of diverticula between the cartilaginous rings. Characteristically has distinct clinical forms of presentation.
Case report: *Case 1*. Male patient of 45 years, non-smoker. Complaints of dyspnea, cough and sometimes with hemoptysis with a few years of development and with recurrent respiratory infections. Chest radiograph showed enlargement of the diameter of the trachea and pattern suggestive of bilateral bronchiectasis. The TAC-chest showed increased caliber and and change in normal configuration of trachea and main bronchi, as well as diffuse varicose bronchiectasis. Bronchoscopy identified dilated trachea with recesses and diverticula, suggesting tracheobronchomegaly, dynamic collapse of the trachea and main bronchi with abundant purulent secretions. The respiratory functional study demonstrated increased residual volume (2.94 L-162.59%) with an obstructive pattern and positive bronchodilation: FEV_1 2.11 L (67.2%-post-bronchodilator), FVC of 2.77 L (73.8% post-BD) and FEV_1/FVC of 76.16% (post-BD). The patient showed improvement in symptoms and in number of exacerbations with the introduction of bronchodilator therapy and integration in pulmonary rehabilitation program. *Case 2*. Male patient of 62 years, smoker and history of Peyronie's disease, gastric ulcer with partial gastrectomy and monoclonal gammopathy. Had symptoms of dyspnoea, productive cough and occasional wheezing. On chest radiograph showed enlargement of the diameter of the trachea and signs suggestive of pulmonary emphysema. A chest CT scan showed dilatation of the trachea and main bronchi, centrilobular emphysema predominantly paraseptal, basal cylindrical bronchiectasis in the upper, medium and basal lobes. The broncofibrospia showed the presence of increased caliber of the trachea and protrusion of cartilage more evident in the 1/3 inferior left side wall .Functionally presented a slight decrease in alveolar-capillary diffusion of CO (DLCO-SB: 65.1 and DLCO/VA: 62.7). Bronchodilator therapy was initiated and the patient was sent for smoking cessationconsultation.The patient abandoned smoking habits, with improvement in symptoms.
Conclusion: This syndrome may be an occasional finding in asymptomatic individuals but when symptomatic is characterized by recurrent pneumonia, with eventual progression to chronic productive cough, hemoptysis or occasional progressive dyspnea. Other complications include massive hemoptysis, spontaneous pneumothorax, digital clubbing and respiratory failure. The MK syndrome may be associated with bronchiectasis, emphysema and pulmonary fibrosis as a form of presentation. Although rare should always be suspected in patients with recurrent respiratory infections and bronchiectasis with chronic expectoration production. An evaluation of the airway anatomy by chest CT scan is important to the diagnosis.

Keywords: *Tracheobrochomegaly. Bronchiectasis. Emphysema.*

HEMOPTYSIS, ETIOLOGY AND MANAGEMENT

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Aim: Evaluate the etiology, diagnosis and treatment of patients admitted for hemoptysis in a pulmonology department.
Methods: Retrospective study of patients admitted for hemoptysis in the Pulmonology Department of a Tertiary Hospital between July 1st, 2011 and July 31st, 2012. We analyzed the demographics, etiology, diagnostic procedures performed, the most common therapeutic approaches and mortality rate.
Results: From a total of 893 patients admitted, 32 patients were hospitalized for hemoptysis. Mean age: 65±14 years. Male sex prevailed (81%). The average length of stay for hemoptysis was 13 days, comparing to a length of stay of 11 days in the Department in the same period. Lung cancer, bronchiectasis and pneumonia were the most prevalent diagnoses (accounting for 28%, 16% and 16%, respectively), followed by other respiratory tract infections (9%), chronic obstructive pulmonary disease (6%), interstitial lung disease (6%), tuberculosis sequelae (3%), aspergilloma (3%), intrabronchial rupture of aortic aneurysm (3%), thyroid cancer with lung metastasis (3%) and iatrogenic (3%) . The cause of bleeding remained unknown after diagnostic workup in only one patient (3%). Concerning diagnostic investigation, chest CT was performed in 72% and bronchoscopy in 75% of patients. Relevant therapeutic interventions included antifibrinolytics in 94% of patients and somatostatin in 9%. In 12% of patients, bleeding was controlled using topical instillation of adrenalin via fiberoptic bronchoscopy. The mortality rate observed in hospitalized patients with hemoptysis was 16%, hemoptysis being the direct cause of death in 9%. The mortality rate in our Department during the study period was 11%.
Conclusion: Hemoptysis remains a troublesome symptom representing 3.6% of all admissions. The main causes in hospitalized patients in our ward were lung cancer, bronchiectasis and pneumonia. Mortality was slightly higher in this subset of patients, comparing to the overall mortality in the same period.

Keywords: *Hemoptysis. Etiology. Management.*

DYSPNEA: DISEASE, SIGNAL OR SYMPTOM?

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Introduction: *American Thoracic Society* defines dyspnea as a subjective experience of breathing discomfort including qualitatively different sensations with variable intensity. In USA it represents one of 10 principal reasons for recourse to Emergency Department (ED). The cognitive alteration in the perception of symptoms amplifies the interaction among physiological, psychological, social and environmental factors and it can induce physiological and behavioral secondary answers, identified by others. Being an unspecific symptom/signal, the identification of the etiology is a challenge. The arterial blood gas analysis (ABGA) doesn't allow to distinguish the etiology of the acute dyspnea, but it can be used in stratification of the risk.
Aim: To understand the clinical, social and environmental profile of patients who resorted to ED of adults of Sousa Martins Hospital screened by flowchart of dyspnea of Manchester Triage System, with very urgent priority (orange).
Material and methods: Retrospective observational study (1/7/2011 to 30/6/2012). In 869 patients 571 were selected with number of consecutive process. The criteria of exclusion applied were: absence of register of the variable in *Alert®*, respiratory alkalosis with hyperoxia and traumatic situations. In patients with more than one resort to ED it was considered the last episode. It was made the framing in groups: I- acute disease again; II- exacerbation of chronic disease; III- isolated symptom without diagnosis of specific disease; IV- signal referred by the others/ registered in evaluation of the patient without diagnosis of specific disease.
Results: Of 59701 episodes of urgency were registered with 3748 (6.3%) screened by flowchart dyspnea. The very urgent priority was attributed in1661 cases (44.3%). It was obtained a sample of 491 patients, 46.4% men and 53.6% women, with the average age of 82years. Most patients live at home (63%). In 53% it was identified a caretaker and 1.2% lives alone. Most registered pathological antecedents were: cardiovascular (67.6%), metabolic (35%), respiratory (33.4%) and neurological (18.9%). Two or more antecedents were registered in 71% of situations. The occurrence of dyspnea was related in 40.9% with exacerbation of chronic disease and in 38.9% of acute disease. 21,6% of patients didn't have diagnosis. The ABGA showed in 15.7% of patients compensated global respiratory insufficiency and in 11.6% respiratory acidemia.

It was made the hospitalization in 63.9% and the hospital discharge in 29.9%. In 4.5% it happened the death in ED. It was verified alteration of pH in 68.4% of the admissions in Intensive Cares Unit (ICU) and in the dead.
Conclusions: The study reflects a geriatric population, coming in majority from home, depending from others and high number of comorbidities justifying the increase of the incidence of dyspnea in view of other studies. The etiology of dyspnea was in most situations related to exacerbation of the chronic disease. In 21.6% it wasn't attributed diagnosis, similar value to other studies. The alteration of pH was a frequent finding in admissions in the ICU and in the dead, what is according to a recent study in which it was associated with mortality.

Keywords: *Dyspnea. Emergency. Manchester triage.*

A CASE OF THORACIC ENDOMETRIOSIS

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Case report: The authors present a case of a 42 years old Caucasian woman that worked at a hotel reception. She was a smoker and denied any other addiction. The patient was unaware of any previous disease and the only medication she was taking were oral contraceptives that she decided to interrupt a week ago. The patient was apparently wealthy until the day she came to the emergency room complaining about a sudden pleuritic pain in the right hemithorax, without irradiation or relief measures. There was no history of trauma, fever, cough, sputum, hemoptysis or dyspnea. The patient denied a similar episode in the past. She was hemodynamically stable, with decreased breath sounds on the right with hyper-resonant sound and vocal vibrations diminished on the right side. The chest X-Ray showed a peripheral hypertransparency at the right hemithorax compatible with a primary spontaneous pneumothorax. The thoracic drainage was placed with progressive decrease in the pneumothorax chamber. At the 18th day of drainage there was no resolution and the patient was transferred to the thoracic surgery department. The patient underwent video assisted thoracoscopic surgery where fenestrations and implants were visualized and were biopsied. The histopathology revealed that those implants were in fact endometriosis implants. In this context, the patient was treated with goserelin for 2 months. Due to gastrointestinal intolerance that medication was substituted by oral contraceptives taken continuously without any complains.
Conclusion: Endometriosis is defined as the presence of endometrial glands in stroma outside the confines of the uterine cavity and musculature. Growth and maintenance of endometriotic implants is dependent upon the presence of ovarian steroids, and therefore endometriosis occurs almost exclusively among women of reproductive age or among those on estrogen replacement therapy. Thoracic endometriosis is an uncommon condition that in 70-73% of the cases is presented by catamenial pneumothorax.

Keywords: *Thoracic endometriosis. Catamenial pneumothorax.*

RARE OR UNDER-DIAGNOSED?

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Idiopathic tracheobronchomegaly or Mounier-Kühn syndrome is a rare disorder characterized by pathological dilatation of the trachea and major bronchi. The clinical manifestation spectrum is broad and nonspecific, however, recurrent respiratory infections is a usual presentation. Computerized tomography (TC) scan of the chest is important for the diagnosis as well as the evaluation of complications, such as tracheal diverticulosis and bronchiectasis. Treatment is mainly supportive. We report a case of a 76 years-old, ex-smoker male with a history of recurrent respiratory infections, bronchiectasis and respiratory failure, who was admitted with the diagnosis of necrotizing bilateral pneumonia and exacerbation of chronic respiratory failure. The TC scan of the chest and bronchoscopy revealed tracheal diverticulosis, extensive bilateral bronchiectasis and tracheobronchomegaly. Therefore, we admitted the diagnosis of Mounier-Kühn syndrome. In conclusion, Mounier-Kühn syndrome should be considered in differential diagnosis of recurrent respiratory infections and of bronchiectasis. The nonspecific clinical manifestations of this disorder, associated with a increasing number of literature cases reported, lead us to question whether this syndrome is so rare as is supposed or whether is under-diagnosed.

Keywords: *Mounier-Kühn syndrome. Tracheobronchomegaly.*

A RARE CAUSE OF HEMOPTYSIS

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Massive hemoptysis is a relatively uncommon phenomenon and can be attributed to many causes including infection, malignancy, trauma, cardiovascular diseases and iatrogenic sources. We present an unusual case of an 88-year-old man with a previous history of COPD, silicosis, treated lung tuberculosis, arterial hypertension and ischemic heart disease that was admitted to the Pneumology Unit with mild self-limited hemoptysis. Bronchoscopic examination did not show any evidence of infection or tumor. A thoracic computed tomography scan with intravenous contrast showed a thoracic aortic aneurysm (4.7×3.3×5 cm) adhered to the left lung. Just after the diagnosis the patient had massive hemoptysis, developed cardiovascular collapse and evolved to death. Upon autopsy, a thoracic aortic aneurism that adhered solidly to the left lung and ruptured to the left lung bronchus was described. This case describes a very rare clinical event of an aortic aneurysm that ruptures into the lung leading to acute, massive and fatal hemoptysis. The patient had no further episodes of hemoptysis

Keywords: *Hemoptysis. Thoracic aortic aneurysm.*

PULMONARY SEQUESTRATION IN ADOLESCENT

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The authors submit the case of a 15 years-old patient, black race, student, non-smoker, previously healthy. He recurred to the emergency department for nonproductive cough and exertional dyspnea, for a month, and at the evaluation he had decreased breath sounds at the inferior third of the right lung with increased transmission of vocal vibrations and with normal laboratorial tests. On the complementary study with chest X-ray it was revealed a condensation in the inferior third of the right lung, that didn't resolve with antibiotic so he underwent a thorax CT that showed an irregular condensation with air bronchogram in the right inferior lobe (RIL) that after contrast injection was shown an arterial vessel coming from the right side of the descending thoracic aorta, irrigating part of the RIL, suggesting intralobar pulmonary sequestration, of non-frequent location. He was referred to a Pulmonology Techniques Appointment, being submitted to a fibrobronchoscopy (that was normal) and was afterwards referred to a Thoracic Surgery Appointment, being submitted to an ablation of the abnormal artery and RIL lobectomy that underwent without complications. Pulmonary sequestration is a rare bronchopulmonary malformation, consisting in a non-functioning pulmonary mass, that receives its arterial blood supply from the systemic circulation and does not communicate with the tracheobronquial tree from a normal bronchus. The diagnosis can be made in the prenatal period, using fetal imagiology techniques.

A more accurate diagnosis should be made prior to the surgical resection of the anomaly, that constitutes the consensual treatment of the symptomatic forms of presentation.

Keywords: *Pulmonary sequestration. Adolescent.*

MULTIPLE PULMONARY CYSTS IN SMOKING PATIENT

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The authors submit the case of a 42 years-old female patient, Caucasian, operations assistant, smoker of 40 pack years, with gastroesophageal reflux disease and uterine polyps. She had been consulted for 4 years for hemoptoic cough and begins with pleural pain, loss of 4 kg, asteny, anorexia, night sweats, keeping hemoptoic cough for a month. She underwent a thoracoabdominal CT scan that revealed moderate emphysema and bilateral cystic formations, non-calcified and an increase of the dimensions of the left lobe of the liver, without other relevant changes. For persistent complaints, she went to the Emergency Department, being medicated with antibiotic and was referred to a Pulmonology Techniques Appointment, being submitted to a fibrobronchoscopy and respiratory function tests that were normal. She began joint pains (small and large joints, with inflammatory and migratory characteristics), accentuated fatigue, repeated the CT (without changes), reason why she began corticotherapy and was referred to a surgical lung biopsy, being obtained the diagnosis of Langerhans cell histiocytosis. The patient was advised not to smoke (that she didn't abide to) and she started corticotherapy transitorily in low dosages with clinical improvement, having regular follow-up Pulmonology appointments, being clinically stable (without corticotherapy) and without functional and imagiological worsening. The authors present this case for the rarity and difficulty of the diagnosis, being this only obtained by lung biopsy, and because, although not having a curative treatment, it is possible to minimize the symptoms and evolution of the disease with corticotherapy, smoking cessation and cytotoxic drugs, reason why we should invest actively in its diagnosis and treatment.

Keywords: *Cysts. Smoker. Arthralgia.*

OESOPHAGEAL RUPTURE-A DIAGNOSE NOT TO FORGET

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The authors present the clinical report of an 83 yd male patient, non-smoker, autonomous on his daily living. Previously asymptomatic, he presented to ER of a hospital with nauseas, emesis followed by epigastric pain for the last 8 days. An acute gastroenteritis was diagnosed and medication started, but without clinical improvement. Two days before re-observation at another hospital the patient started fever, dyspnoea, productive cough and anterior thoracalgia with irradiation to the back. The patient was vigil, calm and oriented. Eupneic and acyanotic at rest. There was a reduction of breath sound and crackles on the lower 1/3 of his right hemithorax. Laboratory data showed mild leukocytosis with marked neutrophilia and CRP elevation. On arterial blood gases he was hypoxemic and the chest X-ray presented a homogenous hypotransparency of the lower 1/3 of the right field. The patient was admitted under the diagnose of community acquired pneumonia and stared on empiric IV antibiotics. During his stay, a pleural effusion developed with progressive worsening of respiratory failure and the patient transferred to the Intermediate Care Unit. The placement of a central venous catheter caused an hydropneumothorax and the insertion of a chest drain, revealed a purulent content, compatible with empyema but with an unexpected rise of amylase and glucose. However clinical stability allowed the patient's transference to the Pulmonology ward. The unfavourable clinical and laboratorial evolution, associated with the constant change on the macroscopic aspect of the drained fluid determined its re-analysis, which showed a rise on amylase from 17000 to 29700 UI/L. The Chest CT scan revealed a loculated piopneumothorax and the presence of pneumo-mediastinum suggesting oesophageal rupture, confirmed after contrast. The patient was transferred to Pulido Valente Hospital's ICU and underwent a conjunct surgery by thoracic and general surgeons. An oesophageal fistula at distal oesophagus was found and closed. The esophageal-pleural fistula is a rare condition associated with high morbimortality, particularly when late diagnosed. This case report illustrates the importance of anamnesis as an essential element of diagnostic thinking, the relevance of the pleural effusion study and favorable evolution of a serious clinical condition.

Keywords: *Esophageal fistula. Piopneumothorax.*

IS DYSPNEA PERCEPTION INFLUENCED BY EXERCISE TRAINING?

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Introduction: Dyspnea is a dominant feature in patients with chronic respiratory disease, causing exercise capacity limitation and quality of life impairment. In these patients, exercise training (TE) has been shown to be effective in functional rehabilitation.
Aim: Evaluate the impact of a TE program on perception of dyspnea and fatigue during exercise.
Methods: We performed a retrospective study in patients with moderate to severe obstruction and airflow limitation in exercise capacity documented in cardiorespiratory exercise test, who integrated a TE program. The combined TE consisted of 20 sessions on bicycle (4 cycles of 3 minutes at intensity equal or above the anaerobic threshold, alternating with 2 minute periods of lower intensity) and 10 minutes in arm ergometer. Aerobic workout was included at the beginning and end of each session. Dyspnea was assessed by MMRC scales before and after TE and modified Borg scale at the beginning and end of each training session and 6-minute walk test, performed before and after the TE program. In the data analysis we used the IBM SPSS 20 program, Wilcoxon test.
Results: 13 patients completed the training, 12 male, aged 57.7±9.4 years. Eleven patients had COPD, 1 had idiopathic pulmonary fibrosis and another with bronchiectasis. The functional study included: $\%FEV_1$ 45.2±13.6, %DLOC 59.2±17.5, and at baseline PaO_2 70.5±10.7 and PaCO2 41.3±5.6.

	mMRC	Borg PM6m	
		Dyspnea	Fatigue
Antes TE	2.2±1.1	1.7±1.5	1.2±1.5
Após TE	1.8±0.8	1.1±1.3	0.5±0.8
P	0.16	0.04	0.26
		Borg dyspnea	Borg fatigue
First training session		1.3±1.1	2.0±1.6
Twentieth training session		0.8±0.7	0.8±0.6
P		0.03	0.04

Conclusions: TE programs positively influenced the perception of dyspnea and fatigue during exercise. Other studies may frame these results in a positive perception of respiratory chronic diseases and improve health expectations.

Keywords: *Exercise training. Dyspnea. Impact.*

THE IMPACT OF HOSPITALIZATION ON SMOKING BEHAVIOR IN PATIENTS WITH PRIMARY SPONTANEOUS PNEUMOTHORAX AND BACTERIAL PNEUMONIA

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Introduction: Primary spontaneous pneumothorax (PSP) and bacterial pneumonia (BP) are acute respiratory diseases that frequently affect younger patients and are responsible for disabling symptoms, hospitalization and invasive treatments. Smokers have a higher risk of PSP and severe BP. The hospitalization represents an opportunity for secondary prevention and orientation.
Objective: To analyze and compare the impact of hospitalization for PSP and BP in smoking habits.
Methods: Between 2007 and 2010, 132 patients with PSP and 254 patients with BP aged ≤55 years were admitted in a pulmonology ward, of which 90 and 96, respectively, were selected according to the following criteria: smokers, without lung pathology or prior history of drug abuse. A confidential telephone questionnaire survey and a retrospective medical record analysis were performed in order to assess smoking habits, the recognition of the relationship between smoking and the concerned diseases, the treatment and orientation during and after hospitalization.
Results: It was possible to perform a survey to 43 (53%) patients with PSP and 38 (47%) patients with PB. At the time of the interview, 29 (35.8%) patients did not smoke, 16 (55.2%) of whom were abstinent since hospitalization. 61 (75.3%) patients stopped smoking after hospitalization, of these, 45 (73.7%) relapsed 4 months (0.25-18 months) after. At the time of the survey, 52 (64.2%) were smoking 14+8 cigarettes/day (CD), significantly less than before admission (20+9 CD) (P<.001). 71 (87.7%) reported having been advised to quit smoking during hospitalization and 48 (59.3%) were aware of the relationship between smoking and its pathology. Nicotine replacement therapy was prescribed to 8 (9.3%) patients; 15 (18.5%) were sent to smoking cessation consultation, of whom 7 (46.6%) didn't attend. Patients with BP presented higher age (40.4+9.7 vs 28.5+8.6 years, P<.001), packs year (30+18 vs 12+10 A, P<.001) and length of stay (11.9+6.8 vs 5.5+2.3 days, P<.001). There weren't significant differences with respect smoking behavior when comparing the two diseases groups. The smoking relapse was not significantly lower in patients with PSP undergone more invasive treatments such as medical or surgical thoracoscopy, or in patients with recurrent pneumothorax. The 3 patients with pneumonia requiring admission to intensive care unit and invasive ventilation didn't relapse smoking habits (100 vs 66.7%, P=.01).
Discussion: A significant number of patients remained abstinent for several months and there was a significant reduction in the smoke load after hospitalization. Pathology, smoke load and the duration of hospitalization had no influence on smoking behavior, as well pneumothorax treatment modality. The admission to intensive care unit seems to be a significant event for the cessation. Most of the patients recognized a relationship between smoking and the hospitalization and have been advised to quit smoking. International guidelines propose a structured program which incorporates post-discharge follow-up.

Keywords: *Pneumothorax. Pneumonia. Smoking prevention.*

EVOLUTION OF SMOKING BEHAVIOR AFTER EARLY WITHDRAWAL OF A SMOKING CESSATION PROGRAM

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Introduction: Tobacco is the leading preventable cause of morbidity and mortality. Despite the recognized effectiveness of treatments currently available, the results fall short of the desirable. Poor adherence to treatment programs greatly compromises the therapeutic success. Repeated interventions in smoking cessation, even on smokers who are not prepared to quit, increase the probability of future attempts succeed. Smokers who drop out of treatment programs are assigned as having failed the intervention. It is not known the evolution of smoking behavior of smokers who leave the intensive support queries.
Objective: Clinical and epidemiological characterization of smokers with early withdrawal in a smoking cessation program. Evaluation of tobacco consumption and motivation to quit on smokers after 6 or more months they attended the consultation.
Methods: Retrospective study based on consultation of the clinical process and applying a telephone questionnaire to smokers who began a program of smoking withdrawal in CHVNG in 2010 and 2011, having, however, abandoned the same up to a maximum of 3 interventions.
Results: In the period considered were made 417 first consultations. 191 smoking were included (67% men), with an average age of 12±46.3 years. The average wait time for integration in the program was 3.6±2.9 months. The population main co-morbidities were respiratory, cardiovascular and psychiatric. Most smokers (59%) had a history of prior attempt smoking cessation. The smoking load average was 38±26 packs-year. The motivation for the cessation was of 7.4±2 (numeric scale analog) and 8±1.5 (Richmond) and the dependency of 5.3±2.2 (Fagerström test). The telephone interview was the 124 individuals, 82 (66%) males. The average age is 47±11 years. Fifteen (12%) of smokers contacted left smoking, 10 in the first month after the last query and none with accompaniment. Increased motivation was the reason cited by 2/3 of the respondents to the success in smoking cessation. About 40% of the total number of respondents referred to cessation or temporary reduction of consumption after the abandonment of the query. About 88% (109) of respondents still smokes at the time of the investigation. Approximately half of these did not change the consumption, one-third has decreased the amount of cigarettes per day with the remaining have increased. Among the current smoking, 37 showed interest in quitting, recognizing they would need specialized follow-up.
Conclusion: The early abandonment of the treatment program is disturbing and is one of the key factors for failure. Although not confirmed, prolonged abstinence is not negligible. About 30% of smokers want to quit smoking and recognizes the need for specialized support. It seems to be apparent the impact of treatment in these results, although it is not possible to quantify it. Improved information about chronic character and the need for treatment and follow-up of smoking addiction can reduce dropouts and relapses. It is also recommended to improve the referencing and accessibility to treatment programs.

Keywords: *Tobacco. Smokers. Withdrawal. Consultation.*

PROFILE OF USER SESSIONS FOR SMOKING CESSATION: SMOKING VERSUS ABSTINENT AT 6 MONTHS

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Since 2002 medical sessions in Smoking Cessation have been increase in Portuguese Health Centers, providing an agglomerate of medical records that has rarely been explored. This study compares users of these heath sessions that have successfully stop

smoking vs users of the same sessions that have not stop smoking at 6 months in terms of: socio-demographic data, smoking history (past), smoking behaviors (present), and cessation perspective (the future). The sample includes 395 participants, with average age of 41.58 years (±12.68); 35.7% females. The data were obtained from medical records of sessions in smoking cessation. The main results indicate that 12.9% of the participants are without smoking at 6 months. The comparison between those who have stop smoking and those who have not shown that the user profile with more success is that who have: older age; consumes less cigarettes per day, takes less capacity to reduce the number of cigarettes per day; perceive less family support to stop smoking. These data provide information to health professionals who develop medical session in Smoking Cessation, indicating elements of the users profile with (un)success in smoking cessation, providing guidance for planning responses that increase the efficiency of these medical sessions.

Keywords: *Epidemiological. Help. Abandonment of tobacco use.*

SMOKING CESSATION PROGRAM ABANDONMENT: THE SMOKERS PERSPECTIVE

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Introduction: The act of smoking is a behavior explained by the combination of physiological, psychological, cognitive and social reasons. The intensive intervention is associated with higher rate in smoking behavior change. Motivation is the key determinant of success and is the first criterion for referral to consultation intensive support. Non-adherence to smoking cessation program is one of the main factors related to its failure. Knowing the patient's point of view may help to improve the response to their expectations.
Objective: To determine the smokers reasons for early withdrawal from smoking cessation program.
Methods: The study was based on clinical file consultation and a telephone interview to smokers that, having started smoking cessation program in CHVNG/E during 2010 and 2011, abandoned it, performing at most, 3 medical appointments. There were included only smokers who responded to the telephone questionnaire.
Results: We interviewed 124 smokers, 82 were male. The average age is 47±11 years. About 48% of smokers referred the lack of motivation as the main responsible for the abandonment of the program, 21% the program didn´t meet their expectations and 18% invokes social reasons. The medical imposition to integrate the program was often identified by smokers who abandoned for lack of personal motivation. In the group of patients whose program didn´t met expectations, ignorance of the smoking cessation process, the need to program the interventions, the role and limitations of pharmacological therapy, the need for psychological intervention and behavior change were identified as factors associated with failure. The change of residence, labor commitment, costs associated with hospital taxes, transportation and the proposed medication were the most mentioned among those who invoked social reason for leaving the program.
Conclusions: Lack of motivation was appointed as the main reason responsible for the early quit from smoking cessation program. The particular false expectations about the role of drugs and ignorance regarding the treatment program is another obstacle to adherence and success. Significant is the weight of social issues that are beyond the capacity of direct intervention of the medical team. It is necessary to improve information for smokers and health professionals. Any modality therapy will fail if not part of a prevention program and tobacco control that ensures accessibility to treatment. The effectiveness of any program will be engaged in a personal and social context adverse to the smoking cessation.

Keywords: *Smoking cessation. Abandonment. Motivation.*

THORACIC IMAGING AND CARDIOVASCULAR CO-MORBIDITIES IN COPD

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Introduction: COPD is a disease with systemic involvement, associated with co-morbidities, responsible for high morbidity and mortality rates. Cardiovascular disease is responsible for 30% of deaths in these patients.

Table 1

Cardiovascular comorbidities	> 3 hospitalizations	Without hospitalizations
Total	39	50
Male	89.7%	86%
mean age	64.6±10.6 years	68.1±10.0 years
Mean FEV_1 (% post BD)	33.3±12%	32,9±9%
Stage GOLD III/IV	25.7%/74.3%	28%/72%

	> 3 hospitalizations	Without hospitalizations	Difference
CV comorbidities			
Isquemic heart disease	37.5%	15.7%	21.8% (*P*=.016)
Cardiac insufficiency	70%	56.9%	13.1% (*P*=.18)
Cardiac dysrhythmia	25%	17.6%	7.4%
Hypertension	55%	47.1%	7.9%
Chest CT scan			
Pulmonary hypertension	8.2%	4.1%	4.1% (*P*=.318)
Cardiomegaly	4.1%	1.4%	2.7% (*P*=.218)
Vascular disease	20.5%	17.8%	2.7% (*P*=.257)
Echocardiography			
Pulmonary Hypertension	10.3%	9.1%	6.2% (*P*=.361)
Cardiomegaly	15.6%	19.5%	(*P*=.836)
Decreased EF	23.6%	5.6%	18% (*P*=.0001)

Objective: Determine the role of thoracic imaging in the evaluation of cardiovascular co-morbidities in patients with COPD.
Material and methods: Retrospective cohort study (2009-2010) including patients with COPD without or with 3 admissions for exacerbation of the disease. We considered demographics and co-morbidities data, changes in echocardiography and Chest ST scan (cardiomegaly, pulmonary hypertension, and vascular disease). Statistical analysis was conducted in SPSS v 20, assuming a confidence level of 95%.
Results: We analyzed 39 patients with at least 3 hospitalizations during the study period and 50 patients without hospitalization. The mean Charlton index is significantly higher in patients with >3 hospitalizations (2.71 Vs 1.92, *P*<.05). In the study population (n=89), 70 (78.7%) of the patients had cardiovascular co-morbidities. Of all patients with co-morbidities (n=70), chest CT scan identified in 7% of the cases cardiomegaly, 24.6% pulmonary hypertension and vascular disease in 45.6%. The difference in the occurrence of cardiovascular co-morbidities between the two groups was more relevant in ischemic heart disease (37.5% vs 15.7% (*P*=.016). When comparing changes in chest CT scan between the two patient groups there were not statistically significant differences. The ejection fraction of the left ventricle showed statistically significant variation in the patient group with 3 hospitalizations (*P*=.0001). Chest CT scan and echocardiogram showed a concordance of 74% in relation to pulmonary hypertension (Table 1 p. 136).
Conclusions: In this study cardiovascular changes identified in the Chest CT scan shown an important contribution and a complement to other findings in the evaluation of cardiovascular co-morbidities of COPD.

***Keywords**: Imaging. COPD. Cardiovascular comorbidities.*

DECLINE OF THE FEV_1: A HETEROGENEOUS MECHANISM IN THE COPD?

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Introduction: The parameter FEV_1 has been used as a marker of the natural history of the COPD and it is described heterogeneity of the rate of annual decline of FEV_1 in patients with COPD.
Objective: To evaluate the annual variability of FEV_1 in patients with COPD.
Method: It was carried out a retrospective study that included 103 patients with COPD (78% males; meanage 62.5±9.7 years old), that had performed multiple spirometric evaluations for a period of 10 years. Foreach patient it was analysed age, initial FEV_1, GOLD Stage and the rate of annual decline of FEV_1 (*software SPIROLA-Longitudinal Spirometry Analysis Date Version 3.0*).
Results: The average rate of decline of FEV_1 varied significantly depending on the severity of the obstruction, being the highest value in GOLD stages I and II, comparing to GOLD III and IV (−48.4±39.7 mL/year; −16.6±64.7 mL/year; −12.7±31.8 mL/year; 3±14.6 mL/year respectively). The standardized regression coefficients showed that the variable baseline FEV_1 significantly influenced the rate of annual decline in FEV_1 (P-value=0.00, α=0.05). The analysis of fast declinators (≥40 mL/year vs <40 mL/year) revealed a mean baseline FEV_1 significantly higher in patients with higher rates of decline of FEV_1 (≥40 mL/year: 1.98±0.77 L).
Conclusion: The rate of decline of FEV_1 among patients with COPD is highly variable, with increased rates among patients with early stages of the disease. This study emphasyses the role of serial sprirometries to detect fast declinator patients.

***Keywords**: Spirometry. COPD. FEV_1.*

SPIROMETRY REFERRAL FROM PRIMARY CARE UNITS TO PNEUMOLOGY DEPARTMENTS

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Background: A larger national network of spirometry will have a wide range of health benefits, facing the expected morbidity and mortality linked with the smoking prevalence in Portugal.
Objective: Earlier diagnosis of COPD would support measures in order to improve the physical performance and health status of COPD patients and would prevent the evolution trend of the disease.
Material and methods: Individuals were selected by GPs for performing spirometry. Among them, were chosen those who fulfilled these criteria: age ≥40 years old with a smoking history >10 pack years and/or professional activity with a known respiratory risk and/or mention to symptom(s): chronic cough or sputum or exercise dyspnea (descriptive: fatigue, short breath). The enregistrement of demographic data, occupation, smoking history, previous pathology and therapeutics were done by a cardiopulmonologist tehcnician. We used the Vitalograph Spirometry equipment with software SPIROTRAC IV/V. Predicted values were obtained from the European tables. The highest values of FVC and FEV_1 were selected after three spirometric tests with a variation coefficient lesser than 5%. It was considered bronchial obstruction when the baseline spirometry registered a FEV_1/FVC ratio<0.70 and the diagnosis of COPD with a FEV_1/FVC ratio<0.70 after bronchodilatation test with a short-acting beta 2 agonist (GOLD guidelines). Statistical analysis was based on the validation of all records and investigation by SPSS software. Results are expressed by its absolute and percentage values, as well as their average and standard deviations values (SD). As a mean of comparison tests, we used the parametric analysis by the "Student's t" and "factor Anova 1" and in the case of qualitative data to χ^2 (chi square), always with a significance threshold lower than 0, 05.
Results: Subsgroups analyzed by smoking history.

	Non smoker	Number of pack-years <10	Number of pack-years ≥10	Number of pack-years ≥30
n	1245	156	3353	1376
Age (≥40 years)	65.8±12.0	57.2±12.1	61.0±11.9	60.3±10.2
Gender, male	25.7%	45.5%	55.4%	80.7%
Pack-Years	0	4.8±2.7	39.8±21.7	50.3±19.6
Obstructed	24.4%	23.1%	32.6%	44.5%
COPD	20.3%	15.4%	27.5%	38.5%

Conclusions: Results analysis should take into consideration the fact that the study was not a random screening, but a scheduled Spirometry series from patients with respiratory risk, oriented by the GP towards the Pulmonology department. It is generally agreed that with the use of Spirometry the obtained COPD prevalence exceeds the prevalence level based only on clinical data. The airflow limitation that is not fully reversible —"post-BD FEV_1/FVC<0.70, with or without symptoms— as has been advocated by GOLD for diagnosis of COPD, overestimates the prevalence of COPD in the elderly, including non-smokers older than 65 years. Anyway, the study does not fail to clearly confirm that COPD is under diagnosed in Portugal.
Graphic design supported by Novartis Farma, S.A.

***Keywords**: COPD. Spirometry in primary care.*

PULMONARY REHABILITATION FOR COPD PATIENTS IN PORTUGAL

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Background: Chronic obstructive pulmonary disease (COPD) is a nationally and internationally highly prevalent disease, but it is currently underdiagnosed and inadequately treated. Although pulmonary rehabilitation (PR) is a standard treatment for COPD, it is still largely unrecognized by both health care professionals and patients. In Portugal there are twelve institutions with PR programs, but are family doctors aware of these programs and of the guidelines for the treatment of the COPD patient? Do these programs have the necessary capacity to respond to all patients who might benefit from PR?
Objectives: to understand how COPD patients are presently participating in PR in Portugal; to know the perceptions of health professionals about PR; to identify the determinant factors of COPD patients' accessibility to PR.
Methods: in this observational cross-sectional study, a questionnaire was produced and applied to a sample of 79 coordinators of Family Health Care Units-for assessing the knowledge, behavior and perceptions of primary care physicians regarding the treatment and management of the COPD patient-and another questionnaire to 6 of the 12 PR programs' coordinators-for assessing the programs' capacity and response, as well as the participation of COPD patients in each. Resulting data was processed using descriptive statistics and correlation tests among variables.
Results: less than half the family doctors attended training sessions on COPD and there seems to be a low level of knowledge about PR in general, which is strongly correlated (P=.000) with the also low referral of COPD patients to PR (9%). The supply of PR is considered by most participating family doctors and PR coordinators as either *Insufficient* or *Very insufficient* and the number of COPD patients who annually participate in PR in responding institutions is, overall, rather low (<80 patients). Moreover, there is an important set of structural, patient and physician-related determinants which hinder the referral to and participation of COPD patients in PR.
Conclusions: there is a significant difference between the number of COPD patients in primary care registers and the number of patients participating in PR annually. Only a small percentage of COPD patients who would benefit from PR is presently having access to this intervention in Portugal, which might be explained by different factors, such as the knowledge of physicians with regards to PR, patients' economic difficulties, among others.

***Keywords**: COPD. Pulmonary rehabilitation. Participation.*

BRONCHIECTASIS — WHY NOT CYSTIC FIBROSIS*?*

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Introduction: In developed countries, Cystic Fibrosis (CF) is the most common cause of bronchiectasis (BC) in children and should be considered in the assessment of adults with this disease. Although, in its classic form, CF is usually diagnosed in childhood due to chronic respiratory symptoms and malabsorption of early onset, this genetic disorder presents a large clinical heterogeneity that results not only due to the severity of the mutations involved but also as a result of the influence of modifying genes and environmental factors. CF atypical manifestations may be subtle, often limited to a single organ, such as repeated lung infections and BC, and frequently present normal or borderline levels of chlorine in sweat.
Case report: The authors present the case of two brothers who were diagnosed with CF in the 5th decade of life. The case began in April 2012, when a male patient, 58 years-old, ex-smoker 20 UMA, was admitted in the Emergency Department for hemoptysis, referring mucopurulent sputum beginning two days before. On auscultation he had bibasilar crackles and showed partial respiratory failure. The chest radiograph showed bilateral bronchovascular strengthening and bronchoscopy revealed purulent and bloody secretions in the left bronchial tree without active bleeding. Bacteriological examination of bronchial aspirate identified oxacillin-sensitive *Staphylococcus aureus*. A CT scan of the chest revealed BC in both upper lobes anterior segments. As the patient had a personal history of infertility and a family history of a sibling with BC the hypothesis of CF was considered and a sweat test was carried out. Consequently, the sibling, now 52, was reassessed. He had been in follow-up in the Pulmonology Clinic for BC since 1995 and had already undergone bronchial artery embolization for hemoptysis. He referred daily sputum production with repeated respiratory infections although without hospitalizations. He had bronchial colonization by *Staphylococcus aureus*. Functional studies revealed mild respiratory obstruction of the small airways and thoracic CT showed extensive bilateral BC in the upper lobes. When questioned, he also referred ~~to~~ infertility and so a sweat test had also been requested. In both cases the sweat tests were positive, confirming the diagnosis of CF and further study was carried out to identify the genetic mutations.
Discussion: CF is a disease of surveillance strategies with well-defined clinical, microbiologic and therapeutic steps which translate into significant benefits for patients, particularly in terms of survival. Thus, although patients with diagnosis in adulthood generally have a better prognosis because they present mostly with pancreatic sufficiency and minor lung disease, it is important to establish the diagnosis as early as possible not only to start appropriate treatment but also because of the implications for genetic counseling. The sweat test should, therefore, be included in the etiological investigation of BC even in adult patients.

***Keywords**: Bronchiectasis. Cystic fibrosis.*

DIAGNOSING CYSTIC FIBROSIS — STILL A CHALLENGE

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Background: Cystic fibrosis (CF) is an autosomal recessive and multiorganic disease. The most common mutation, among Caucasians, is the delta-F508. About 70% of diagnoses are performed before 2 years of age. Pulmonary disease is the leading cause of morbidity and mortality.
Case report: A 23 years-old man, with no relevant history, showed a productive cough since 13 years-old. It was assumed the diagnosis of asthma and bronchiectasis (2007-Table 1). In 2010, he showed unfavorable evolution (2010, Table 1) and performed two sweat tests, which were positive (77/82 mmol/L). Genetic testing revealed homozygosity for the delta F508 (confirming the clinical suspicion) and heterozygosity in the parents. The patient was referred to a CF-specialized consult in Hospital São João and presented favorable evolution (2012-Table 1) with the beginning of treatment.

Table 1

	2007	2010	2012
Symptoms (cough, dyspnea, limited physical activity, recurrent infections, GERD)	+	+++ IMC 15.69	BMI 16.0 Increased exercise tolerance Without nasal/digestive symptoms
% FEV_1/% TLC/% RV	67.3/81.6/252.7	42.2/65.0/264.0	45.9/96.3/267.9
Blood gas analisys	Normal	PaO_2: 73	PaO_2: 67.8
Bacteriology	MSSA	MSSA	MRSA+MSSA+*Achromobacter*+*Aspergillus*
Thoracic HRCT	Cylindrical bronchiectasis in RML	Cylindrical bronchiectasis in RML, RLL e LLL	Diffuse bilateral bronchiectasis Pancreatic fat infiltration
Echocardiography	Not done	Right ventricular dysfunction	Not done
Abdominal Ultrasonography	Normal	Mild hepatic steatosis	Not done
Others	Prick tests+for mites	Normal alpha 1-antitrypsin Pansinusitis (CT)	Osteopenia Pancreatic insufficiency Vitamin deficit (D, A, E)

Conclusion: Diagnosing CF in adulthood requires a high degree of suspicion, since nowadays CF is no longer exclusively a pediatric disease. It remains a challenge in clinical practice for medical specialties in general and in particular for Pulmonology.

Keywords: *Cystic fibrosis. Diagnosis. Adulthood.*

A CASE OF CYSTIC FIBROSIS

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Introduction: Cystic fibrosis (CF) is the most common fatal autosomal recessive disease among Caucasian populations, with a frequency of 1 in 2000 to 3000 live births. The usual presenting symptoms and signs include persistent pulmonary infection, pancreatic insufficiency, and elevated sweat chloride levels. However, many patients demonstrate mild or atypical symptoms, and clinicians should remain alert to the possibility of CF even when only a few of the usual features are present.
Case report: The authors present a case of a 44 years old Caucasian woman, non-smoker, employee at a supermarket. At 2004 she was followed in consult for recurrent respiratory infections due to bronchiectasis of the right upper lobe. In this context, the patient was subject to right pneumonectomy complicated with a methicillin sensitive *S. aureus* empyema. The patient remained asymptomatic until the year 2009, when she came to the hospital complaining about dyspnea at small exertion and a productive cough with a four months evolution. There was no history of fever, chest pain, hemoptysis or orthopnea. Objectively, the patient had a body mass index of 13 Kg/m^2, was hemodynamically stable and afebrile. The patient presented resting global respiratory failure with severe hypoxemia (pO_2: 50 mmHg; pCO_2: 54 mmHg) so it was decided to start long-term oxygen therapy. There were made a transthoracic echocardiogram and pulmonary function tests that showed secondary pulmonary hypertension and a severe restrictive pattern, respectively. It was still requested a thorax CT that revealed residual bronchiectasis at the left lung. When trying to identify the bronchiectasis etiology it was requested an alpha 1 antitrypsin dosing and phenotyping as well as a sweat testing. The last one was positive in two different occasions. To confirm the cystic fibrosis diagnosis was done a research for mutations at the cystic fibrosis trans membrane conductance regulator (CFTR) gene which concluded that our patient was a combination of two different mutations: F508del and 3272-26A>G.
Conclusion: The phenotypic expression of disease varies widely, primarily as a function of the specific mutation (or mutations) present. There are more than 1300 different mutations in the CFTR gene with potential to cause disease. The F508del is a severe mutation while the 3272-26A>G mutation is a mild one and these two combined constitute a phenotype with middle severity which explains the late diagnosis in this case.

Keywords: *Cystic fibrosis. Adult. Bronchiectasis.*

QUALITY OF LIFE AND RESPIRATORY REHABILITATION: ANALYSIS OF A PROGRAM

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Introduction: Chronic respiratory diseases are frequently associated with a significant and measurable loss of quality of life, particularly the symptomatic and debilitating forms. Respiratory rehabilitation is a global and multidisciplinary intervention directed at this population, with demonstrated benefits in the improvement of patient's symptoms and quality of life as well as shortening their hospitalization time.
Objectives: Evaluation of the gains in quality of life at the end of a respiratory rehabilitation program developed in a department of pulmonology.
Methods: A one year long prospective study was performed, including patients involved in a respiratory rehabilitation program in a department of pulmonology. The program lasted at least 12 weeks, with twice weekly attended sessions including exercise training in a treadmill, cycle ergometer and arm cycle ergometer, muscle strength training, education and psychosocial and nutritional intervention. The quality of life was evaluated at the beginning and at the end of the program, by the use of the questionnaires Saint George Respiratory Questionnaire (SGRQ), EuroQuol and Hospital Anxiety and Depression Scale (HADS), Portuguese versions. In parallel, and according to the program protocol, the exercise capacity, nutritional state and lung function were also evaluated in all patients, at the beginning and end of the program.
Results: Twenty three patients were included (22 males), with an average age of 67.8 years (47 to 80). The most frequent diagnosis was COPD (92%), including 5 patients in GOLD stage II, 8 in GOLD III and 8 in GOLD IV. At the beginning of the program the average score on SGRQ was 51.8±6.0, and this was not significantly modified at the end of the program (51.0±10.5). This lack of significant change was observed in all components of the questionnaire, although there was a tendency for a decrease in the symptoms component (56.6±20.4 at the beginning and 48.6±26.1 at the

end). Concerning EuroQuol questionnaire, no significant changes were also observed with the respiratory rehabilitation program (10.1±2.0 at the start and 10.0±2.1 at the end). The results of the HADS did not change with the rehabilitation program as well (anxiety component-7.76±3.9 at the start and 7.9±3.8 at the end; depression component-6.45±3.5 at the start and 7.1±3.8 at the end). Finally there was no significant correlation between the results of the quality of life questionnaires and the lung function at the beginning of the program. These results are in contrast with the observed improvement in exercise capacity with the program, where a significant increase in the endurance time was noted at the end of the program (from 07:03±03:54 to 21: 8±09: 03; *P*<.05).
Discussion: The results of our study of the effects of respiratory rehabilitation on patients the quality of life are in contrast with previously reported studies and with the changes in the exercise capacity of the same population. This may be due to a lower sensitivity and objectivity of the questionnaires compared to the functional evaluation of exercise, as well as some difficulties in the filling of the questionnaires by the patients. Future studies should test the use of questionnaires with higher sensitivity and lower filling complexity.

Keywords: *Respiratory rehabilitation. Quality of life. Questionnaires.*

EVALUATION OF BENEFITS OF A RESPIRATORY REHABILITATION PROGRAM: BEFORE, AFTER AND A YEAR LATER

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Introduction: Respiratory rehabilitation (RR) is a multidisciplinary and holistic intervention directed to patient with chronic respiratory disease. Evidence has demonstrated its multiple benefits. However, currently, it is known that these decrease after the end of the program.
Objectives: To assess physical activity, respiratory function and nutritional status of patients who joined a program of RR, on 3 occasions: at the beginning, after completion and ≥12 months after its completion. Quantify, characterize and discriminate maintenance or loss of benefit after 12 or more months of the end of the program.
Material and methods: The program was conducted in a hospital. After the program patients were encouraged periodically to maintain the level of physical activity. Before joining the program patients underwent a comprehensive assessment that included, among others: assessment of physical activity (APA)-specific endurance test (SET*) and proof of 6-minute walking (P6MW); respiratory functional evaluation; assessment of dyspnea; nutritional assessment. This assessment was repeated at the end of the program and at ≥12 months after completion and we proceeded to comparison of results. *SET (Cooper CB, ATS, 2007) is a constant exercise test, held on treadmill, with previously calculated load in incremental test.
Results: Sixteen patients completed the RR program ≥12 months (mean: 18, min 12, max: 27 months), with mean age of 69.1±9.86 years, 15 males, 10 ex-smokers and 15 with COPD. At the end of the program we found benefit in APA, with increasing of endurance time (median: 7′35″ vs 23′20″, *P*<.005). Twelve or more months after the end of the program, endurance time decreased significantly (median: 23′20″ vs 5′30″, *P*<.005), for similar values to the pre-rehabilitation. The P6MW did not reflect the benefits of the program. After the RR program, we verified benefits in quantification of dyspnea and body composition, also lost ≥12 months after completion of the program, but without statistical significance.
Conclusions: Benefits of RR were reflected in the increase of endurance time, which suggests that Cooper's endurance test can be a privileged form for that evaluation. Twelve or more months after the end of the program there is a loss of benefit. Effective strategies for maintaining benefits must be defined.

Keywords: *Rehabilitation program. Respiratory rehabilitation. Revaluation.*

CHARACTERIZATION OF A POPULATION IN RESPIRATORY REHABILITATION PROGRAM

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Introduction: All patients with chronic respiratory disease appear to benefit from pulmonary rehabilitation (RR), which is a fundamental component in the management of these patients. The co-morbidities are common, interfering with disease progression and therapeutic efficacy.
Objectives: To characterize the population in a respiratory rehabilitation program (RRP). Demographic data, education level, socioeconomic status, accessibility to the program, nutritional status and co-morbidities where evaluated.
Methods: Retrospective study of 34 patients admitted in a RRP, in the Pulmonology Department of Hospital Geral-CHUC, from March 2010 to May 2012. Analysis of the major comorbidities based on cardiac evaluation (with electrocardiogram, transthoracic echocardiogram, and SCORE index), lipid profile, nutritional status (with bioimpedance balance-TANITA, BC-545) and index of anxiety and depression with the HADS questionnaire.
Results: Patients had a mean age of 65.8±1.17 years, 91.2% male. The majority (88.2%) with smoking habits. With respect to respiratory disease, 88.2% had chronic obstructive pulmonary disease (COPD) (16.6%-GOLD II, 30%-III and 53%-IV). The mean BODE index was 4.7±1.9. Regarding educational qualifications: 63% were schooled until 4th grade, 29.6% until 6th, and 3.7% (n=1) had a university education, only one patient illiterate. Regarding the socio-economic status, 48.2% of patients reached a monthly social support (419.20€). Patients lived at a mean distance of 38.7 Km from the rehabilitation center, and 27.6% lived at a distance greater than 50 km. Comorbidities were identified in 94% of the patients. Dyslipidemia in 53% of patients, a mean total cholesterol of 5.1±1.06 mmol/L (N<5.2 mmol /L). With regard to cardiovascular disease, 47.8% had arterial hypertension, pharmacologically controlled. Dysrhythmia was identified in 14.7% of cases, 32.4% had an abnormal echocardiogram, and the mean SCORE (*European Cardiology Society*) mortality at 10 years for cardiovascular disease was 2±1%. There was anxiety and depression in 11.7% and 8.8% of the patients, respectively. In the review of the clinical process, 11.8% of the patients had diabetes mellitus type 2. Other comorbities have been identified, like obstructive sleep apnea syndrome, ischemic peripheral vascular disease, cancer (oropharynx, palate, prostate) and systemic lupus erythematosus. In the nutritional assessment, a mean BMI of 27.5±4.4 kg/m^2 and 18.5±2.5 kg/m$^{2\ of}$ lean body mass index, 44.1% of the patients were overweight, 20.6% obese and 17.6% had depletion of muscle mass.
Conclusion: In this population there was a high incidence of comorbidities. The most prevalent were dyslipidemia, excess of weight and arterial hypertension. The identification and treatment of the co-morbidities are critical when addressing a RRP, thus contributing to a greater therapeutic success. The low

socio-economic status, low education level, and the distance to the rehabilitation center difficult the accessibility to the RRP.

Keywords: *Respiratory rehabilitation. Comorbidities. COPD.*

GENDER DIFFERENCES IN ALPHA-1 ANTITRYPSIN DEFICIENCY

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Introduction: Gender differences in airway behavior and in the clinical manifestations of airway disease have been described.
Objective: To compare gender differences in the clinical presentation of alpha-1 antitrypsin patients (α_1AT).
Methods: This retrospective study included patients followed in the Hospital São João outpatient department, aged >18 years that had α_1AT deficient alleles (phenotypes ZZ and SZ) confirmed by phenotype determination by IPATIMUP, between 2002 and 2010.
Results: Forty six patients were included, 69.6% were males and 30.4% were females. Twenty two patients (47.8%) were homozygote (ZZ) and twenty four (52.2%) were heterozygote (SZ). The mean age at diagnosis was lower in females (38.5 vs 42.3 years), as well as smoking history (7.1% vs 65.6%, *P*<.05) and mean duration of symptoms (7.4 vs 10.4 years). SZ women had higher levels of α_1AT, but without statistical significance (66.6 mg/dL vs 58.1 mg/dL, *P*=.1). Women had an higher number of asthma diagnosis (28.6 vs 16.1) and less COPD (21.4 vs 65.4, *P*=.007). ZZ patients there was a predominance of bronchiectasis in females (60% vs 25%) and emphysema in males (87.5% vs 20%, *P*=.011); lung function was significantly better in women: FEV_1 (79.06 vs 38.8, *P*=.026), FEV_1/FVC ratio (68.7 vs 43, *P*=.017), DLVA (92.6 vs 46.4, *P*=.021) and pulse oximetric saturation nadir during the 6MWT (91.8 vs 81.6, *P*=.009).
Conclusions: Gender differences were found, regarding clinical presentation, radiographic and respiratory function. However, an heavier smoking history can be an explanation for these differences in male patients. Further studies are needed to establish the true contribution of gender in this pathology and its implications in terms of diagnostic and therapeutic strategy.

Keywords: *Alpha-1 antitrypsin. Gender.*

HUMAN ANTI-MOUSE ANTIBODIES: LOOKING FOR THE INEXISTENT PULMONARY THROMBOEMBOLISM

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Background: D-dimer determination is an important tool for the diagnostic approach of low-to-moderate pre-test probability of pulmonary thromboembolism (PE), in the emergency department(ED). We report the second ever published case of a patient that, after several presences in the ED, was repeatedly submitted to angio-CT scans after evidence of a high d-dimer value, explainable by immunological interference by presence of human anti-mouse antibodies (HAMA).
Case report: A 58 year old patient went for three times to a central hospital ED for moderate effort dyspnea, cough and chest pain. At the first episode, lab tests revealed a strikingly elevated d-dimer value, with normal fibrinogen levels and no other confounding abnormalities like erytrocitosis, hyperbilirrubinaemia or inflammatory markers. He underwent an angio-CT scan that was negative for PE, although documenting emphysema. After some days he returned to the ED and, again, was submitted to d-dimer test followed by a new angiography by chest CT. By the time of his third presence in the ED, the possibility of an immunological phenomenon involving a heterophile antibody was hypothesized. The patient was further studied outpatiently by his Pulmonologist. There was no hypertrigliceridaemia, medication with heparin or dicumarinic or elevated rheumatoid factor. The case was discussed with the Clinical Hematology Lab and further immunological testing was accomplished.
Conclusions: The possibility of false-positives in d-dimer ELISA testing should be present.Although rare, the presence of anti-animal specific antibodies interferes in the diagnostic study, leading to unnecessary angio-CT scans with higher cost and the risk of high dose of electromagnetic radiation. This kind of antibodies may arise after therapies with animal immunoglobulins and possibly by environmental contact with certain animals.

Keywords: *Pulmonary thromboembolism. Heterophile antibody. D-dimer.*

PULMONARY EMBOLISM LIKE FORM OF PRESENTATION OF BEHÇET DISEASE

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Introduction: Behçet's disease is a chronic inflammatory multisystemic disease characterized by orogenital recurrent ulcerations, eye and skin lesions, among others, in particular cardiovascular manifestations. The prevalence of cardiovascular involvement varies between 7.7% and 43%, including the presence, amongst others, pulmonary embolism, intracardiac thrombosis and pulmonary artery aneurysms.
Case report: We report the clinical case of a male patient aged 38 with multiple admissions for respiratory infection in 2008, with chest pain, dyspnea, fever, cough, with occasional hemoptysis and erythema nodosum. A chest radiograph had a predominance of peripheral hypotransparent vaguely triangular with external base subpleural, which raised the suspicion of thromboembolism, confirmed by a CT angiography on the admission. This also detected with a mass 30x22mm, adherent to the free wall of the right ventricle, compatible intracavitary thrombus, later confirmed by cardiac MRI. The complementary diagnostic exams (CDE) for initial etiologic research revealed: Doppler echocardiography of the lower limbs without change; study of peripheral blood for autoimmune thrombophilic alterations or negative. Oral anticoagulation was prescribed but the patient failed to follow-up appointments. In 2010 he returned to be admitted through the emergency department with clinical respiratory infection, erythema nodosum and recurrent and lush oral ulcerations in the last year, confessing not to have fulfilled the proposed therapy. The CDEs revealed: VS of 93 mm h 1st, proteinuria (<1 g/24 h) without changes in renal function, antiphospholipid antibodies, ANCA's and ANA's negative with other autoantibody negative for collagen diseases; mild hyperhomocysteinemia 18 mmol/L; HLA-B51. The echocardiogram revealed a decreased thrombus for 22×10 mm and thoracic CT showed thromboembolic pulmonary arteries with aneurysmal dilatations in the apical segment of the right lower lobe. The Doppler ultrasound of the lower limbs with evidence of previous thrombosis with recanalization. The pathergy test was positive and there were superficial phlebitis of the upper limb secondary to venipuncture. He was treated with prednisolone 60mg with significant clinical improvement after 1 month of treatment but the patient has left the follow-up.
Conclusion: This is a patient with recurrent respiratory infections, secondary to thromboembolic phenomena as a result of intracardiac thrombus. This patient meets the criteria of The International Criteria for Behcet's Disease (ICBD) for Behcet's disease: have recurrent ulceration of the oral mucosa, erythema nodosum, positive pathergy test and vascular manifestations, heart and lung,

with phenotyping HLA B51 frequent in this pathology. The right ventricular thrombus and pulmonary artery aneurysms are rare and signs of poor prognosis, they may precede the other manifestations.

Keywords: *Behcet. Pulmonary thromboembolism. Intracardiac thrombus.*

DRUG-RELATED COMPLICATIONS IN POST PULMONARY TRANSPLANT PATIENTS

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Introduction: Pulmonary transplantation is an important therapeutic option for end stage respiratory diseases. Although its success is definitely related to the lifelong use of immunosuppressive agents they have a high risk of side effects, complications and morbidity.
Aim: To determine the prevalence, type and severity of drug related complications in post-pulmonary transplanted patients.
Methods: Retrospective analysis of the clinical processes of patients followed in the Centro Hospitalar de São João who underwent pulmonary transplant since 2005.
Results: 32 patients were included, 19 (59.4%) were male, mean time of follow-up post-transplant was 30.78±17.8 months and mean age at time of procedure was 44.91±12.8 years. The initial immunosuppressive therapy used in all patients was corticoid, cyclosporin and azathioprin. Six patients still maintain that scheme, while for others adjustments were made due to acute rejection (n=16), suspicion of chronic rejection (n=4) and adverse effects/immunosuppressive complications (n=14). Amongst those, we highlight a malignancy diagnosis in 2 patients (non-Hodgkin lymphoma and cutaneous lymphoma), gastric intolerance (n=4), renal impairment (n=2) and hirsurtism (n=2). Leucopenia was diagnosed as an antibiotic prophylaxis side effect in 2 patients, treatment discontinuation was not necessary. During the follow up period, 8 viral infections and 3 fungal infections were diagnosed, and 17 hospital admittances for severe bacterial infection treatment occurred. For patients presenting such infections immunosuppressive therapy was statistically longer than for patients with no infections (P=.025), and no other parameter (age, diagnosis, type of transplant, pre-transplant treatment) was found to be infection related. Regarding the adverse effects, requiring specific treatment but not treatment cessation, the most common were dyslipidemia (n=12, 37.5%), diabetes (n=10, 31,5%) and hypertension (n=4, 12.5%) .
Conclusion: High incidence of adverse effects and complications prevail during immunosuppressive therapy in post-transplanted patients. Close monitoring assures proper diagnosis, treatment targeting and timely adaptation of immunosuppressive and prophylactic therapy.

Keywords: *Lung transplant. Immunosuppression. Adverse effects.*

OVERCOMING ANATOMIC INCOMPATIBILITY IN LUNG TRANSPLANTATION

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The compatibility between the dimensions of the donor lung and the receptors' chest cavity is one of the main obstacles that we face in lung transplantation. In pediatric patients or in young adults with small chests, as is typical in patients with cystic fibrosis, this represents an even more vital problem because of shortage of donors in pediatric age. Since the 90's a small number of centers worldwide, perform lobar transplants, not only from cadaveric but also from living donors. We present the clinical cases of two patients with cystic fibrosis, that were on the waiting list for lung transplantation, in a high urgency situation, that were submitted to lobar implants from adult cadaveric donors after ex-vivo lobectomies. One of them an 18-year old, with progressive deterioration of his clinical status, with long-term oxygen therapy (LTOT) and non-invasive ventilation (NIV) that also had an asymmetric *pectus carinatum* with a clearly smaller right hemithorax. We performed, an upper bilobectomy on the donor lung and implanted only the right lower lobe, and then the complete left donor lung was implanted on the left side. Extracorporeal circulation (ECC) was necessary for the second implant but the post-operatory period had no complications. The patient was extubated 36 hours after surgery and was discharged from the hospital on the 31st day. His FEV1 after one month was 1900 cc and 2910 6 months later. The other patient, a 13-year old, had been committed to the hospital for the two previous months under LTOT and NIV, also with progressive worsening of her clinical status. Considering the size of her chest we had to perform a lower lobectomy on the right and upper lobectomy on the left donor lungs, implanting the upper and middle lobes on the right and the lower lobe on the left side. ECC was also necessary for the second implant. The patient was extubated on the 7th day and was discharged 26 days after surgery. The FEV1 on the first month was 1290 cc and 1750 cc on the fifth month. Both patients had an uneventful evolution and are clinically well 20 and 8 months after their transplants without need of re-admission.

Keywords: *Lung transplantation. Size mismatch. Lobar transplant.*

RESPIRATORY AMYLOIDOSIS, FROM SURPRISE TO PANIC

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Amyloidosis is a disease of protein metabolism characterized by the deposition of amyloid substance in several organs, causing different degrees of dysfunction. Bronchopulmonary envolvement is a rare situation either as a focal form of the disease or as part of a widespread systemic amyloidosis. We present three different clinical situations with challenging differential diagnosis and treatment solutions: A 68 year-old female, Caucasian, smoker CT 55UM, history of COPD. Presenting one-year history of fatigue, without weight loss, ECOG PS 1. Chest-CT showed "two cm nodule in right upper lobe (RUL)"; Lung function tests (LFT) with bronchial obstruction (BD negative), DLCO/VA 97%; Fiberopticbronchoscopy (FB) was normal; PET "RUL lesion (SUV 2.07), not allowing exclusion of malignant lesion". In this context she underwent right upper lobectomy which histology revealed nodular amyloidosis of the lung. The study for systemic or secondary causes of amyloidosis was negative. A 62 year-old nonsmoker man, Caucasian, past history of arterial hypertension and repeated respiratory infections. In November 2009 he was send to consultation for haemoptysis, dysphonia and fatigue. The physical examination revealed dysphonia and wheezing. The study showed: Immunofixation with monoclonal IgG Kappa fraction; LFT with severe bronchial obstruction (BD negative); FB showed "thickened vocal cords and a bulky obstructive mass on the right wall of the upper third of

the trachea with multiple scattered raised plaques on the right". Improvement was achieved with endobronchial treatment. The biopsy revealed amyloid deposits. The laryngeal biopsy revealed laryngeal amyloidosis. Performed laryngeal microsurgery with CO_2 laser with improvement of dysphonia. The patient is proposed for systemic chemotherapy antimyeloma. A 75 year-old nonsmoker woman, Caucasian, with past history of malignant Histiocytosis subjected to right lower lobectomy in 1963, and repeated respiratory infections. In 2003 cough, haemoptoic expectoration, headache and polyarthralgia. Chest-CT "bilateral residual lesions"; skull x-ray "two lytic lesions on the left parietal bone"; FB "ulcerated plaques in both trees ecchymotic proximal bronchial; Biopsy confirmed tracheobronchial amyloidosis. The patient remained in follow-up with recurrent episodes of haemoptysis, after refusing to carry out further exams. In January 2012, in the context of haemoptysis, she developed respiratory failure requiring mechanical ventilation. Osteomedular biopsy revealed aberrant dispersed plasmocytes suggestive of multiple myeloma; serum immunofixation "discreet presence of fraction monoclonal IgM Lambda". The diagnosis of amyloidosis associated with monoclonal gammopathy and the presence of clinical complications led to therapy antimyeloma.
Discussion: Tracheobronchial envolvement is the most common presentation of pulmonary amyloidosis with a wide range of symptoms such as dyspnea, wheezing, cough, haemoptysis and recurrent respiratory infection. Systematic tissue sampling is always necessary for a correct diagnosis and planning of a multidisciplinary treatment.

***Keywords**: Amyloidosis. Management approach.*

PLEURAL EFFUSION AS INITIAL PRESENTATION OF RHEUMATOID ARTHRITIS-CASE REPORT

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Introduction: Pleural effusion is a common extra-articular manifestation of rheumatoid arthritis (RA), particularly in middle-aged men with positive rheumatoid factor and rheumatoid nodules. The authors present a case of diagnosis of RA in a patient with pleural effusion and without joint symptoms.
Case report: We report the case of a Caucasian 51 years old male patient, tobacco smoker of 44 pack-year, with personal history of treated pulmonary tuberculosis 15 years ago. He was referred to our consultation in 2009 to investigate a right pleural effusion detected on a thoracic CT scan that also showed a left pleural nodular thickening, interlobular interstitial thickening and a nonspecific micronodule. No further investigation was made due to patient's refusal. In 2012 he was again referred to our consultation to investigate the right pleural effusion, with no history of fever or any symptoms. A thoracocentesis was made with removal of a thick purulent/milky liquid with pH 7.0. The diagnosis of empyema was made and treatment was initiated with thoracic drainage and empirical antimicrobial therapy. Biochemical analysis of the pleural fluid showed exudates features, with glucose<10 mg/dL, LDH 2986 U/L, ADA 200 U/L and cholesterol 205 mg/dL; microscopic examination showed lipid and crystalline material; Gram stain and cultures were negative; cytological examination showed rare mesothelial cells and rare lymphocytes, without neoplastic cells. Pleural biopsies revealed fibrosis and marked inflammatory infiltrate without granulomas or neoplastic tissue. On physical examination we found two painless and voluminous tumefactions, one on the left wrist and another on the left elbow. Extensive tenosynovitis of the extensor digitorum communis of the index, middle and ring fingers and elbow bursitis with important synovitis were documented in ultrasonographic study. The radiological examination of superior and inferior members showed periarticular soft tissue swelling, changes of the shape and density of the articular surfaces and loss of joint lines distinction. Blood tests revealed mild anemia (Hg 12.4 g/dL), leukocytosis (13.3×10.9/L) with no neutrophilia, high CRP (2.76 mg/dL) and ESR (34 mm h) and positive rheumatoid factor (31.3 UI/mL) and anti-citrullinated protein antibody (>250 UA/mL). According to American College of Rheumatology/European League Against Rheumatism (2010) we made the diagnosis of RA-score of 9/10 (involvement of more than 10 joints, high-positive rheumatoid factor/anti-citrullinated protein antibody and abnormal ESR/CRP). The purulent pleural fluid, with very low pH and glucose levels and high LDH levels, in the presence of negative bacterial smears and cultures, suggested the presence of rheumatoid sterile empyematous pleural effusion.
Conclusion: This is a case of an unusual presentation of RA which illustrates the importance of considering the diagnosis of RA in a patient with pleural effusion with uncommon characteristics.

***Keywords**: Pleural effusion. Sterile empyematous pleural effusion. Rheumatoid arthritis.*

ANTISYNTHETASE SYNDROME

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The Antisynthetase Syndrome (ASyS) is a subset of the spectrum of the idiopathic inflammatory myopathies (IIMs) and is characterized by the sudden onset polimiositis or dermatomiosites of sudden onset, fever, non-erosive symmetric arthritis, Raynaud phenomena and interstitial lung disease (ILD) with the presence of auto-antibodies specific to aminoacyl-tRNA synthetases. The presence of these autoantibodies correlates with the lung involvement and has important clinical and prognostic implications. The authors report the case of a 44 year-old female nurse, previously asymptomatic, who presents to her respiratory physician with myalgia, arthralgia, dry cough and striking exercise impairment, which, after clinical investigation proved to be an antisynthetase syndrome with anti-Jo1 positive autoantibodies. The timely diagnose and the start of immunosuppressive treatment with steroids and azathioprine led to a favourable clinical evolution, regression of the imagiologic features and great improvement in her lung function. This case report illustrates the relevance of a high suspicion index for the ASyS on ILD patients. The importance of the diagnose concerns its prognostic and therapeutic implications. Searching anti-synthetase autoantibodies, namely anti-Jo1 and the determination of CK are useful procedures on patients with ILD. The treatment with steroids and immunosuppressive therapy is necessary on most patients.

***Keywords**: Anti-synthetase syndrome. Lung interstitial disease. Autoimmune disease.*

MISCELLANEOUS PNEUMOCONIOSIS AND HARD METAL LUNG DISEASE, ASSOCIATED WITH SILICA-RELATED SJOGREN

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Background: Welding is associated with relevant inhalatory exposure to various gases and particles, most commonly to hard metals, silica and iron. Intense and long exposures to silica can

rarely be associated with the emergence of Gougerot-Sjogren Syndrome. We report the case of a welder with a long lasting exposure to hard metals, silica and asbestos, diagnosed with parenquimatous lung disease due to hard metal lung disease, silicosis, asbestos-induced pleural disease and probable pulmonary involvment secondary to Sjogren.

Case report: A 75 year old patient, welder for over 35 years reporting welding of metal alloys, polishing with sandblasting and regular isolation of tools with asbestos. He started with grade II/III MRC effort-dyspnea and persistent dry cough, three years before the diagnose. He came to cyclically develop liquenified scaly skin lesions on his limbs, Reynaud phenomenon and sicca syndrome. Radiographically there was a reticular interstitial pattern. Chest CR showed egg-shell calcification of mediastinal lymph nodes and fibrotic abnormalities as interlobular reticulation, traction bronchiectasis and volume loss, along with focal areas ground glass/consolidation over bronchovascular bundles. Pleural thickening was also evident. BAL disclosed a total cell count of 130 cells/mm^3, with 40% lymphocitary predominance—ratio CD4/CD8 0.68—and 16% neutrophils, with negative microbiological and cytopathological studies. Immunophenotyping presented a lymphocitary predominance of T CD8 and B cells. The inorganic fraction was sent to determination of hard metals and silica having shown high levels of silica, copper, cobalt, chromium, rubidium and zinc. B2 microglobulina was 6.3 mg/dL, complement was normal, serum ACE 127 U/L, and there was a IgG polyclonal hypergammaglobulinemia. He presented a typical serological profile of primary Sjogren on auto-immunity testing, and a moderate restrictive syndrome with moderately diminuished diffusion capacity and low distance accomplished at 6MWT. Shirmer's test was positive for xeroftalmia. Surgical lung biopsy evidenced areas of fibroblastic proliferation with macrophage cells with antracosic pigmentation and birrefringent particles, as well as several giant multinucleated cells, along bronchovascular bundles and interlobular septa. There was associated diffuse pleural fibrosis and alveolar macrophagic accumulation. A diagnose of mixed pneumoconiosis to silica and asbestos and hard metal lung disease, probably associated with primary Sjogren with parenquimatous involvment and xerotrachea. He was started on oral corticotherapy with partial improvement of dyspnea and complete remission of the skin lesions.

Conclusions: This case exemplifies the remarkable expression of miscellaneous interstitial disease, resulting from the convergence of long lasting inhalatory exposure to silica, asbestos, hard metals, associated with the probable participation of lymphocitary infiltration of Sjogren. The data correlation from imaging, pathology and immunological study was important. Welders are at high risk for developping hard metal lung disease overlapped with pneumoconiosis.

Keywords: *Pneumoconiosis. Sjogren. Hard-metal lung disease.*

IDIOPATHIC PLEUROPARENCHYMAL FIBROELASTOSIS: AN UNKNOWN ENTITY

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Introduction: Pleuroparenchymal fibroelastosis is a rare entity, first described in 2004. It is characterized by pleural and subpleural parenchymal fibrosis, mainly in the upper lobes. The etiology and pathophysiology are unknown. The clinical course of disease is progressive and prognosis is poor. No therapeutic options other than lung transplantation are currently available.

Case report: Female patient, 66 years old, Caucasian, non-smoker, with history of esophageal hiatus hernia. Asymptomatic until a year ago, when she complaint of dyspnea on exertion and non-productive cough. A chest x-ray performed at that time revealed a reticular densification with peripheral distribution and predominance in the upper lobes. Lung function tests showed moderate reduction in diffusion capacity. High resolution chest CT showed pleural and subpleural thickening with moderate fibrotic changes in the marginal parenchyma, mainly in the upper lobes. Subsequently, a bronchoscopy was performed and bronchoalveolar lavage revealed lymphocytosis (28.4%) with CD4 predominance (CD4/CD8 ratio of 14.5) and transbronchial and bronchial biopsies had no evidence of malignancy or granulomas. The patient was submitted to CT-guided transthoracic lung biopsy in two distinct locations, complicated by pneumothorax with bronchopleural fistula and subcutaneous emphysema. Histological evaluation found a marked fibroelastosis and a patchy lymphoplasmacytic infiltrate. In conclusion, it is a patient with clinical, imaging and histology compatible with idiopathic pleuroparenchymal fibroelastosis. Therapy with azathioprine was started, and the patient is clinically, functionally and imagiologicaly stable until now.

Conclusions: Idiopathic pleuroparenchymal fibroelastosis is a distinct clinicopathologic entity, usually connected with a poor prognosis, which should be properly identified in order to be correctly addressed.

Keywords: *Interstitial lung disease. Pulmonary fibrosis. Fibroelastosis.*

CONSTRICTIVE AND GRANULOMATOUS BRONCHIOLITIS — A RARE HISTOLOGICAL PATTERN OF HYPERSENSITIVITY

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Introduction: Bronchiolitis is a generic term that includes a group of diseases with different etiologies and distinct anatomopathological patterns, generally reflecting the presence of inflammation in the small airways. They can be classified as specific, like constrictive bronchiolitis and granulomatous bronchiolitis, or non-specific. Constrictive bronchiolitis is a rare entity characterized by inflammation and fibrosis with collagen deposition, conditioning narrowing or occlusion of the lumen of the bronchioles. Granulomatous bronchiolitis is a disease which is normally associated with Crohn's disease or secondary to chemotherapy, and is histologically characterized by the presence of epithelioid granulomas near the alveolar ducts.

Case report: The authors describe a case of a female patient, 76-year old, referred to the Pulmonology Department because of a pulmonary infection associated with nonspecific radiological findings. She had a history of Graves disease, hypertension, dyslipidemia, atrial fibrillation, heart failure, osteoarticular disease and had worked for several years in wood-crafting without the use of protection devices. The patient reported persistent complaints of nocturnal wheeze, dyspnea and dry cough with insidious onset and progressive worsening. The initial pulmonary function tests were normal with DLCO also within normal limits. The CT Chest scan revealed the presence of ground glass areas with air entrapment and bronchiectasis bilaterally. The autoimmunity study, immunoglobulins, and skin tests were negative. Due to persistent complaints, bronchoscopy

with bronchoalveolar lavage was performed. No endobronchial alterations were found, and the bronchoalveolar lavage presented low cellularity. The trans-bronchial biopsies only led to a nonspecific histological diagnosis of pneumonia. Due to the difficulty in obtaining a final diagnosis and the clinical and functional worsening, the patient was submitted to a surgical biopsy which allowed the final diagnosis of constrictive and granulomatous bronchiolitis. After this result, treatment with corticosteroids was initiated, and the patient maintains close follow-up in the Pulmonology Department.
Conclusion: We describe a case with a histological diagnosis of bronchiolitis presenting morphological markers of constrictive bronchiolitis, like the presence of peri-bronchiolar collagen, and of granulomatous bronchiolitis, as the presence of epithelioid granulomas. These entities by themselves are rare. However, there are cases reported in the literature with the association occurring after chemotherapy or secondary to pulmonary metastization. In this particular clinical case the causal relationship between the two entities seems to be a hypersensitivity reaction, manifesting as a rare anatomopathological form of Hypersensitivity Pneumonitis. Corticotherapy is usually the first therapeutic option. However, many of these situations are resistant to corticosteroids, and the association of other immunosuppressors is controversial, primarily due to the absence of specific studies.

Keywords: *Constrictive and granulomatous bronchiolitis.*

AIRWAY-CENTERED INTERSTITIAL FIBROSIS — A CASE REPORT

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Introduction: Airway-centered interstitial fibrosis (ACIF) is an interstitial lung disease, recently described, characterized by bronchiolocentric pattern of pulmonary lesion, which shows peribronchiolar fibrosis and chronic inflammatory infiltrate which extends into the adjacent interstitium, typically without granulomas. Patients with ACIF are mostly female and usual complaints are progressive dyspnea and dry cough. The etiology of ACIF is unknown and there are no identified risk factors associated with the occurrence of this disease.
Case report: The authors report the case of a 69- year old woman admitted to the Emergency Room with dyspnea, progressive asthenia, productive cough and purulent sputum with a 4-day history. She was afebrile and presented with type 1 (Hypoxaemic) respiratory failure that responded to oxygen. Chest X-ray showed a bilateral nonspecific interstitial infiltrate and so she was admitted for treatment of a respiratory infection and to study a possible Interstitial Lung Disease. The patient had a history of hypertension, atrial fibrillation treated with amiodarone and a left mastectomy submitted to cobaltotherapy 30 years ago. Adding to this she had a daily exposure to chemicals from household cleaning and was a non-smoker. Laboratory tests showed a subclinical hypothyroidism, tumor markers within the normal range, except for CA 125 (35 U/mL), negative autoimmunity and precipitins and protein electrophoresis with a slight monoclonal peak in the Beta-Gamma transition. Chest CT scan revealed a ground glass pattern in all lung lobes, with only a few small areas of normal lung. Further study included a bronchoscopy that showed signs of inflammation grade I and II and bronchoalveolar lavage (BAL) showed 11% lymphocytes and a CD4/CD8 ratio of 4.93. Pulmonary function testing was normal (FEV1 97.5%, FEV/ FVC 81.7%, DLCO 87.3%). In April 2012 a surgical biopsy of the middle lobe was carried out and histological study showed morphology consistent with ACIF. The patient was followed up in to the Interstitial Lung Disease outpatient clinic and in May 2012 started on corticosteroid therapy with 45 mg/day of deflazacort. There was improvement of respiratory symptoms, is currently on a dose of 30 mg/day and is continuing to show clinical improvement.
Conclusion: ACIF is an interstitial lung disease poorly described in which the most common symptoms are nonspecific and shared by several respiratory. As the main differential diagnosis is hypersensitivity pneumonitis, a careful medical history, assessing exposure to antigens and the determination of precipitins, are essential to reach a swift diagnosis. Treatment recommendations are corticosteroids although clinical response is often poor and mortality is usually high.

Keywords: *Dyspnea. Airway-centered interstitial fibrosis. Interstitial lung disease.*

EOSINOPHILIC PNEUMONIA: A CLINIC CASE

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Introduction: The Chronic eosinophilic pneumonia (CEP) is a rare disorder characterized by alveolar and interstitial eosinophilic infiltration of unknown cause. It can occur at any age, but the peak incidence is in the fifth decade of life, predominantly in females at a ratio of 2:1. Patients show symptoms a few months before diagnosis, such as progressive dyspnea, cough, fever, fatigue, night sweats and moderate weight loss. The cough, which is usually dry at first, can be productive with mucoid disease progression, is an almost universal symptom. Dyspnea is usually mild or moderate. Hemoptysis and extrapulmonary symptoms are not common. About two thirds of the patients have a history of atopy. In different studies, prior history of asthma was present in 52% of cases. Spontaneous remission of PEC rarely occurs and may even evolve into standard pulmonary fibrosis. The following case describes the clinical presentation and treatment response typical of this uncommon disease and therefore of special interest to professional pulmonology.
Discussion: A 36 year old technical salesman, nonsmoker, with no history of drug or occupational exposure with personal history of atopic asthma to dust mites in the house since childhood with a strong component of rhinoconjunctivitis. He has been treated with long-acting bronchodilators and inhaled corticosteroids which he does on a regular and correct manner. He was in good general condition, eupneic, acyanotic, without clubbing. Normal pulmonary and cardiac auscultation. In October 2010 the patient develops a predominantly nocturnal dry cough, uses the Otorhinolaryngologist as if it being treated for asthma but later on goes to his family doctor where he performs a chest x-ray that reveals pulmonary infiltrates in the apex, bilateral, confirmed by CT and analytically significant eosinophilia, however, for a strong suspicion of TP starts AB broad spectrum and is sent to the specialty where he repeats chest radiograph showing disappearance of lesions on the right and left worsening. Performed bronchoscopy finding no macroscopic changes in bronchial lavage eosinophils without bacteria alcohol acid resistant. Given the clinical and analytical findings this led to the diagnosis of CEP and initiating corticosteroid therapy is the treatment of choice. The patient maintains consultations with Pulmonology with significant improvement of symptoms.

Keywords: *Eosinophilic pneumonia. Bronchoscopy. Interstitial infiltration.*

PULMONARY LANGERHANS CELL HISTIOCYTOSIS

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Pulmonary Langerhans' cell histiocytosis is an uncommon disorder for which no accurate epidemiological data is available, that occurs more often in young (20-40 years old) patients with a smoking history and is characterized by the infiltration and destruction of the terminal bronchioles by granulomas of Langerhans' cells. The clinical presentation of the disease can be widely varied and symptoms may be minimal or attributed to smoking (25% of cases). In two-thirds of cases, symptoms like a dry cough and dyspnoea on exertion may be associated to constitutional symptoms (asthenia, fever, night sweats and weight loss) and in 10-20% of cases the primary sign may be a spontaneous pneumothorax. High-resolution CT is essential for the diagnosis, usually showing nodular lesions (2-10mm) and cavitated nodules and cysts in the middle and upper lobes of both lungs. The definitive diagnosis requires the identification of Langerhans cells granulomas in lung biopsy but due to the potential iatrogenic risk, the technique needs to be considered on a case by case basis. It is considered adequate a presumptive diagnosis based on clinical manifestations, suggesting images in HRCT and a Bronchoalveolar lavage fluid with high numbers of macrophages and identification of Langerhans' cells using the monoclonal antibody against CD1a (OKT6). Due to the possibility of false-positives in smokers (values of CD1a up to 3%) and other interstitial diseases with alveolar hyperplasia, a threshold value >5% of CD1a cells is considered to have a good specificity for the diagnosis although in some series has a low sensitivity (<25%). We report a case of a young female patient, 29 years old, with a smoking history up to 2 years ago, diagnosed with deficit of alpha1 antitrypsin in routine examination with normal lung function tests and with no need for specific therapy. During her 2-year follow-up in the Pneumology outpatient unit she had an initial HRCT in 2010 showing some emphysematous bullae, in the upper lobes. In 2012 the HRCT also showed multiple cavitated nodules dispersed throughout both lungs and centrilobular micronodules in the upper lobes. The patient denied respiratory symptoms of any kind. A Bronchoscopy was made and the bronchoalveolar fluid identified 12% of CD1a cells and many histiocytes. Although no Langerhans' cells granulomas were identified in transbronchial biopsies, the fact that the patient had quit smoking 2 years prior, the elevated number of CD1a cells and the presence of many histiocytes in the bronchoalveolar fluid allows an adequate diagnosis of Pulmonary Langerhans' cells Histiocytosis. This case illustrates the diagnostic process for this disease and is interesting due to the presence of two different pulmonary pathologies in the same patient.

Keywords: *Pulmonary Langerhans cell histiocytosis. Histiocytosis X. CD1a. OKT6.*

THE DIFFERENT SIDES OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Pulmonary hypertension (PH) is a rare disease that can be idiopathic or associated with several diseases. It is characterized by elevated pulmonary vascular resistance leading to right ventricular failure and death. It is defined as mean pulmonary artery pressure (mPAP) greater than or equal to 25 mmHg, at rest as assessed by right heart catheterization. Chronic thromboembolic pulmonary hypertension (CTEPH) is one of the most prevalent forms of HP and the only one potentially curable and it should be excluded in patients with a history of symptomatic pulmonary embolism (PE). The authors report three cases that illustrate this entity: 33 year-old female with a known history of large B cells Non-Hodgkin lymphoma in remission since 2007 and severe PE in July 2010 with an estimated PASP of 85 mmHg. In 2010 she started complaining of asthenia and dyspnea witch persisted until November 2011. At that time, she described grave functional limitation (functional class-FC-NYHA IV), lower limb edema and orthopnea and her estimated value of PASP was 83 mmHg witch lead to the hypothesis of PH. The CTEPH diagnosis was confirmed after right heart catheterization (mPAP: 52 mmHg) and CT angiography (thrombotic material in the left pulmonary artery in almost its entire length). The patient was considered for pulmonary endarterectomy. A 35 year-old female with a history PE in 2004 who underwent pulmonary thrombectomy whose outcome was suboptimal, with NYHA class III and important postoperative residual PH (mPAP: 52 mmHg). Since there was no history of drug use, respiratory, heart or hereditary diseases, the diagnosis was reviewed and changed to Pulmonary Arterial Hypertension and the patient started triple vasodilator medication with an initial symptomatic improvement. However, after several months of optimized vasodilator therapy at the maximum tolerated dose, the patient maintained a stable but unsatisfactory state and was considered for lung transplantation. A 34 year-old female, with an irrelevant history. In 2007, because of a 12 years old complaint of fatigue that recently progressively aggravated, she was submitted to a ventilation-perfusion scintigraphy that documented changes consistent with PE. She was diagnosed with having protein C and S deficit and PH (mPAP: 30 mmHg). Pulmonary angiography showed diffuse vascular disease, mostly at the level of the right pulmonary artery. CTEPH was considered and the patient underwent pulmonary endarterectomy (2009) with symptom remission and normalization of pulmonary hemodynamics. With these cases, the authors wish to draw attention to the fact that, despite the presence of personal or family history of PE/DVT is very suggestive of the diagnosis of HPTEC, this is not mandatory. Moreover, in some cases pulmonary thromboembolic disease can be just one component of HP which will persist even after pulmonary endarterectomy, which is curative in other situations. The authors also wish to highlight that the presence of elevated PASP should raise the suspicion of PH, especially in patients with risk factors, requiring confirmation by right heart catheterization.

Keywords: *Pulmonary hypertension. Thromboembolism. Diagnosis.*

ALVEOLAR HEMORRHAGE AS FIRST CLINICAL MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

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Introduction: Diffuse alveolar bleeding is a rare complication of Systemic lupus erythematosus (SLE). Incidence of this condition is low, reaching about 2% of SL patients, more prevalente in female (4:1). Alveolar bleeding is associated with high mortality, fluctuating between 45% and 60%. Pathogenesis is not well understood yet, but most of data correlates with immunocomplexes deposits.

Methods: A 44 years old woman, with a background of hypothyroidism and two thyroid nodules, obesity, hypertension and a history of nephritis ate eleven years old appears with an one week of dyspnoea, persistent cough with hemoptoic sputum.

Chest radiography shows a reticulonodular bilateral and enhanced bronchovascular plot.
Results: CT scan shows ground glass pattern. The bronchoalveolar citology showed intra-alveolar bleeding; serologies for other viruses and tuberculosis were made and they were negative. She was medicated with metilprednisolone. It was request anti-nuclear antibody wich was positive (mottled pattern). Further evaluation showed positive anti-SSA e anti-SSB as well positive scintigraphy salivary glands and capillaroscopy. She is actually medicated with immunosuppressive therapy.
Conclusions: Pulmonary bleeding secondary to SLE is an unusual and serious complication and it could be the first sign of the disease. In most cases, diagnosis is already settled months before. However, 20% occurs as first sign, complicating diagnosis. Therapeutic delaying could compromise disease progress, hence precocious diagnosis is essencial, being corticotherapy the usual treatment.

Keywords: *Alveolar haemorrhage. SLE. Corticotherapy.*

IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION VS. PULMONARY CAPILLARY HEMANGIOMATOSIS

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Introduction: The pulmonary capillary hemangiomatosis (PCH) and pulmonary veno-occlusive disease (PVOD) are rare nosological entities, which should be considered in the differential diagnosis of pulmonary arterial hypertension (PAH). The diagnosis should be suspected based on clinical presentation and suggestive findings on lung computed tomography (CT). The diagnosis is histopathological through lung biopsy.
Results: The authors document a case of a man of 47 years, admitted for progressive exercise dyspnea, with 2 months of evolution, accompanied by episodes of syncope. After performing a transthoracic echocardiogram suggestive of pulmonary hypertension (PH), with severely impaired right ventricular function, he was referenced to our center in WHO class IV. He began his diagnostic algorithm, with highlight to the lung CT, which shows a centrilobular micronodular pattern, with ground glass opacity, interlobular septal thickening and presence of mediastinal lymphadenopathy, features not suggestive of idiopathic PAH. In conjunction with other tests, various etiologies were excluded, doubt remains between idiopathic PAH, PCH and PVOD. At admission various criteria of severity and poor prognosis were present and sequential vasodilator therapy with sildenafil, bosentan and inhaled iloprost was started. After obtaining clinically stability lung biopsy was performed with histological confirmation of PCH. Under triple therapy sustained improvement was observed over 1 year of follow up. We stress the clinical improvement for WHO Class II, the decreasing of biochemical biomarkers and increased functional capability documented by functional tests. After the first year he initiated progressive clinical deterioration over 12 months despite titration of therapy with prostanoids, at this stage in ev continuous infusion. He was hospitalized again in WHO class IV and was discharged clinically referred to cardio-pulmonary transplantation; after 8 months on waiting list, he died in hospital from right ventricular dysfunction.
Conclusions: The importance of making this differential diagnosis should be the consideration of treatment options (considering titrations in inpatient treatment due to frequent complications in these patients and early lung transplantation) and to establish the prognosis.

Keywords: *Pulmonary capillary hemangiomatosis. Idiophatic pulmonary arterial hypertension.*

ESADA: EUROPEAN SLEEP APNEA DATABASE

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The European Sleep Apnea Database ESADA is a European multi-center long term observational Cohort. At present 22 sleep centers are participating including the Sleep Laboratory of the Pneumology Department, of Santa Maria Hopsital (HSM). The ESADA project aims to create a multinational on-line database containing diagnosis, treatment and co-morbidities of patients with obstructive sleep apnea (OSA). The results are supposed to give insight in the pathogenesis of OSA, its cardiovascular risk and help to develop future strategies for the optimization of diagnosis and treatment and ultimately the creation of European standards. Depending on the availability of study members eligible patients with OSA will be included after observation by a physician trained in sleep disorders. The diagnostic and therapeutic data will be registered on-line, e.g. anthropometric data, daily sleepiness parameters (SDE), diagnosis, therapeutic procedures as well as the associated co-morbidity and type of pharmacological therapy. We describe here the rationale, procedure and advantages of the European database in respect of the above mentioned endpoints.

Keywords: *OSA therapeutic. OSA diagnosis. OSA mortality/ morbidity.*

SLEEP APNEA TREATMENT AND HYPERSOMNOLENCE. THE DIFFICULT ASSESSMENT OF RESIDUAL SLEEPINESS. A PUPILLARY SLEEPINESS TEST BASED STUDY

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Introduction: Excessive daytime sleepiness (EDS) is a common feature in patients with obstructive sleep apneas (OSA). However, EDS is not observed in all patients and there is at best a mild correlation between respiratory or sleep parameters and the prevalence of EDS. Global EDS is estimated by the Epworth sleepiness score (ESS) while the Stanford sleepiness scale (SSS) measures the acute level of sleepiness. Although extensively tested, both scales are subjective and therefore sensitive towards over or underestimation depended on patient's motivation. The pupillographic sleepiness test (PST) uses slow oscillation of the pupil, the so called pupillary unrest index (PUI) to estimate objectively the acute sleepiness.
Methods: We investigated 39 patients with a respiratory disturbance index >5 h at three time points (TP): 1) after the diagnostic night, 2) after a CPAP titration night and 3) following 3 month of APAP therapy. Following each night with polysomnographic recordings (PSG) the patients filled the SSS and ESS questionnaires and performed the PST. Results are demonstrated as mean±standard deviation. Additionally, the differences between TP1 towards TP2 and TP3 were calculated. Changes in the PUI were considered relevant if the subtraction revealed a difference of more than 2 points (P). Thus, compared to baseline, a positive value indicates a decrease in the PUI or less sleepiness, while a negative value stands for the opposite.
Results: Mean PSG and sleepiness data of are displayed in Table 1. The changes of the PUI measured objective sleepiness are listed in table 2. After the first night of CPAP therapy the majority of patients show no changed PUI. After 3 month of therapy, however, the majority demonstrate an improve in the PUI.

Table 1.

TP	SE (%)	SWS (%)	REM (%)	Arousal (/h)
1	81±1.8	10.3±1.3 *#	12.2±1.1 *#	54.0±3.4*#
2	78.4±2.2	19.8±1.8	27.3±12.2	30.0±2.6
3	83.3±1.8	16.44±1.9	16.5±0.8	21.8±1,5

TP	RDI (/h)	ODI (/h)	T90 (/h)	ESE (/h)	ESS (/h)	PUI [mm/min]
1	53.3±4.4[a,b]	38.9±4.1[a,b]	12.6±3.3[a,b]	10.5±0.8[a,b]	2.42±0.2	6.1±0.7
2	12.6±2.2	6.6±1.7	1.8±0.3	7.3±0.7	2.22±0.2	6.6±0.6
3	6.1±1,5	7.2±2.8	0.7±0.5	5.4±0.5	1.9±0.1	6.3±0.6

SE: sleep efficiency, SWS: slow wave sleep, RDI: respiratory disturbance index, ODI: oxygen desaturation index, T90: time of SpO_2<90%, ESS: Epworth sleepiness scale, SSS: Stanford sleepiness scale, PUI pupillary sleepiness index.
[a]TP1 vs TP2<0.05.
[b]TP1 vs TP3<0.05.
Neither SSS nor PUI reached statistical significance.

Table 2. Number of patients with a change of the PUI of more than 2P. Following the first night of CPAP titration the majority of patients did not demonstrate any relevant change.

TP	Difference PUI<−2P	Difference PUI −2 to +2P	Difference PUI >+2P
TP1 vs TP2	9	20	8
TP1 vs TP3	3	9	23

However, 3 month of APAP was accompanied with a clear increase of the patients showing a reduction in sleepiness. We also analyzed the change of PUI against the differences in the SSS at TP1 compared to TP2 or TP3. Note that a negative value in the SSS indicates increased sleepiness, while a positive result indicates increased vigilance.

Table 3. Change of Stanford sleepiness scale (SSS) in dependence of a 2P change of the PUI. A negative change of the PUI was accompanied by a decrease in the SSS while an improvement of the PUI >2 also demonstrated a reduction in the subjective sleepiness.

Alterações na ESE	Number of PUI<−2P	Number of PUI −2 to +2P	Number of PUI>+2P
F1 vs. F2	−0.56	+0.05	+1.13
F1 vs. F3	−0.66	+0.44	+0.61

Discussion and conclusion: In this study we evaluated the effect of OSA treatment on the subjective and objective sleepiness. On the first glance the ESS appears to exhibit best correlation with the PSG results. However, strictly speaking the ESS should analyze sleepiness over a period of 3 weeks. In fact the improvement of several items of the ESS e.g. "driving a car" is not only unlikely but impossible after one night of CPAP treatment. Nevertheless these items were frequently marked as decreased in the questionnaires. The two tests of acute sleepiness PUI and SSS did not change significantly. This might be based on two factors. First: The adaption problems at the first 2 TPs (first night effect and first time with nasal mask) might cause in some patients a paradoxical sleepiness not explained by respiratory values. Second: The PUI is a mean value over 8 time frames. By falling asleep during the test the following epochs are scored zero and thus the patient, although clearly sleepy, demonstrates a lower total PUI than real. By avoiding absolute values and analyzing only the change of PUI we could demonstrate that the number of patients with a decrease in objective measured sleepiness increase after 3 month adaption time, and that relevant changes of the PUI are associated with a decrease in subjective sleepiness. In conclusion: the PUI might indeed be a fast and uncomplicated test to measure acute sleepiness, but absolute values should be used with caution.

Keywords: *Obstructive sleep apnea. Excessive daytime sleepiness. Polysomnographic. Recordings. Pupillographic sleepiness test.*

SLEEP APNEA AND MADELUNG'S DISEASE

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Introduction: The authors present four clinical cases which describes Madelung's disease associated with Sleep Apnea Syndrome (SAS) in patients with previous palliative surgeries and with symptoms of fatigue and daytime sleepiness with previously known diagnosis of multiple symmetric lipomatosis (MSL) and with a history of smoking and alcoholism accented. The Symmetric lipomatosis Multiple (LSM), also called Madelung's disease, has an unknown etiology, often associated metabolic disorders and chronic alcoholism, characterized by symmetrical and progressive accumulation of adipose tissue of non-encapsulated benign character, cervical, face and/or chest, producing severe disfigurements. Its prevalence is highest in the Mediterranean area, the male reaches in 30:1 ratio. There are 2 types according to their location, the type I fatty tissue builds up around the neck, shoulders and upper back, in type 2 lipomas extend throughout the body, including the thighs, is considered a rare disease with an unknown incidence. Moreover, the Sleep Apnea Syndrome is a condition characterized by the change in the flow of inspired air, where there is total or partial collapse of the upper airways, affecting multiple health problems, including cardiovascular, without forgetting the social implications secondary the framework of drowsiness and chronic fatigue. The definitive diagnosis is made by polysomnography or cardiorespiratory polygraphy sleep according to clinical situations. Patients in the study were sent to the query pulmonology, referenced by your family doctor for presenting changes during sleep including snoring and periods of apnea with daytime symptoms mentioned above. The initial approach in all cases was similar to the initial

collection of the history and application of Epworth Sleepiness Scale that in all cases identified excessive sleepiness; research was performed in 2 cases with polysomnography, cardiorespiratory polygraphy in 1 case and oximetry night in 1 case, identifying all situations in apnea-hypopnea index higher. Following refers to three of the cases studied began CPAP successfully passed y 1 precozmente for oncologic pathology. Being the Madelung's disease a rare disease are few published cases when associated with SAS, although the evolution of Madelung's disease is different for each patient, the benefit of treating associated diseases and in this case the SAS remarkably improves the quality of life for patients.

Keywords: *Madelung. Apnea. CPAP.*

PATIENTS WITH OBSTRUCTIVE SLEEP APNEA SYNDROME DIAGNOSED IN SCREENNG STUDIES: CPAP ADHERENCE

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Introduction: In the treatment of Obstructive Sleep Apnea-hypopnea Syndrome (OSA) the elective treatment is ventilation therapy with continuous positive airway pressure (CPAP). Several trials have indicated a high prevalence of OSA in patients with coronary artery disease and/or diabetes. There are very few data published regarding the eventual CPAP adherence in patients diagnosed in screening studies of OSA in this population.
Objective: Evaluate the CPAP adherence in patients with OSA diagnosis made in screening studies of patients with Acute Coronary Syndrome (ACS) and Type 2 Diabetes *Mellitus* (DM).
Methods: Restrospective study with 188 patients with the diagnosis of OSA and indication to use CPAP between 2009 and 2010. Patients with other associated diagnosis such as COPD and Obesity-hypoventilation Syndrome have been excluded. The study sample was divided into two groups: group diagnosed by screening (GDS) with 45 patients (17 diagnosed in the context of ACS and 28 in the context of DM) and, group referred to the appointment (GRA) with 143 patients. Every patient underwent polysomnography or cardiorespiratory polygraphy, began treatment with CPAP and underwent the same follow-up according to the Respiratory Sleep Pathology Appointment protocol. The demographic characteristics (gender, age, Body Mass Index, Epworth Scale score, Apnea-Hypopnea Index), adherence or refusal of therapy with CPAP (at the beginning, first, third and last appointment) of the two groups were compared using chi2, Fisher's, T student or Mann-Whitney tests, according to the indications. The statistical analyses were performed using SPSS v 18.
Results: In the characterization of groups it has been there was a statistically significant correlation only between Epworth Scale (GDS 6.9±3 vs GRA 10±4, $P<.0001$), with no differences between gender, age, Body Mass Index or Apnea-Hypopnea Index). There haven't been significant differences between groups regarding refusing CPAP at the beginning of treatment (GDS 2 patients vs GRA 3 patients, $P=.595$), first month (GDS 3 patients vs GRA 5 patients, $P=.400$), third month (GDS 2 patients vs GRA 11 patients, $P=.739$) and at the last appointment (GDS 1 patient vs GRA 2 patients, $P=.562$). There haven't been statistically significant correlation between the groups concerning the adherence to CPAP in the first month (GDS 4.1±2 hours vs GRA 4.7±2 hours, $P=.868$), third month (GDS 4.2±2 hours vs GRA 4.9±2 hours, $P=.104$) and at the last appointment (GDS 5.5±2 hours vs GRA 6±1.6 hours, $P=.162$).
Conclusions: The patients with ACS and DM diagnosed and treated with CPAP in the context of a screening study of OSA had a similar CPAP adherence comparing to the patients that were referred to the Respiratory Sleep Pathology Appointment. The lowest score of sleepiness in the group of patients diagnosed in the screening did not have a negative influence in the CPAP adherence.

Keywords: *CPAP. Adherence. Screening.*

EFFECT OF DEEP BRAIN STIMULATION ON A PARKINSONIC PATIENT WITH OBSTRUCTIVE SLEEP APNEA SYNDROME — A CASE REPORT

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Introduction: Parkinson's disease (PD) is associated to several sleep disturbances among which obstructive sleep apnea syndrome (OSAS). The loss of tone in upper airway muscles, in PD patients, is one of the major factors leading to sleep apnea.
Case report: A 68 year-old woman, retired dressmaker, non-smoking. She reported a history of hyperthyroidism, advanced stage of idiopathic PD and insomnia. She was usually medicated with levothyroxine 250µg 1id, selegiline 1.25 mg 1 id, ropinirole 8 mg 1 id, levodopa 25 mg/carbidopa 100 mg 8 id, venlafaxine 75 mg 2 id, bromazepam 3 mg 1 id and trazodone 150 mg 1id. The patient was sent to Sleep Apnea Consultation of the Hospital Center of Gaia/Espinho in 2009 because she had long duration snoring, nocturnal arousals with asphyxiation feeling, symptoms of restless legs and non-repairing sleep with excessive day sleepiness (EDS)-Epworth Scale (ES) scored 15/21. She performed a cardiorespiratory sleep study with apnea-hypopnea index (AHI) of 21.4 h with a minimum O_2 saturation of 74%. The sleep hygiene measures were explained to the patient and she began treatment with nasal continuous positive airway pressure (A-CPAP) during the night. The patient always presented poor therapeutic compliance caused by reduced tolerance of the ventilator, with complaints of dryness of the mucous membrane and sleep fragmentation caused by a feeling of high pressure in the airway. From 2009 to 2011 the PD worsened with increased stiffness, bradykinesia and tremors. The sleep circadian rhythm also was affected. In November 2011 the patient was submitted to a surgery for implantation of a deep brain stimulator of the subthalamic nucleus (DBS-STN) in the Hospital of São João. After the surgery it was possible to reduce the dosage of dopaminergic medication and there was a clear improvement in the motor symptoms of PD, with no muscular stiffness, mild bradykinesia and mild neck dystonia. In the Sleep Apnea Consultation, after the implantation of the DBS-STN, the patient reported an improvement of the snoring, absence of witnessed apneas, disappearance of nocturnal arousals, lack of restless legs symptoms and of periodical limbs movements during sleep. She presented repairing sleep without EDS with a score of 3 on the ES. Before this clinical picture, the patient performed another polysomnography with an AHI of 4.1h, minimum O_2 saturation of 94% without detection of periodical limbs movements during sleep. The significant improvement in sleep quality of the patient was interpreted in the context of the implantation of the DBS-STN and, once there were no longer criteria of OSAS the patient was discharged from the Sleep Apnea Consultation.
Conclusion: In PD, the long term deep brain stimulation of the subthalamic nucleus improves the motor symptoms and, in the majority of cases, the reduction of the dosage of dopaminergic medication. Furthermore, there are recent studies which show a subjective and objective improvement of the sleep quality with

this treatment, as observed in this patient. In this case report, the authors intend to highlight the possibility of implantation of a DBS-STN in PD patients with criteria for its use and with OSAS, with the possibility of improvement for both the pathologies.

Keywords: *Obstructive sleep apnea syndrome. Parkinson´s Disease. Deep brain stimulation.*

OBSTRUCTIVE SLEEP APNEA SYNDROME — NOT EVERYTHING IS WHAT IT SEEMS

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Introduction: There are several pathologies which lead to nocturnal arousals, such as heart failure, asthma and obstructive sleep apnea syndrome (OSAS). This last one should be equated when patients present snoring and witnessed apneas.
Case report: A 42 year-old woman, textile worker, married, resident in Vila Nova de Gaia, was sent to the Sleep Apnea Consultation of the Hospitalar Center of Gaia/Espinho because she had snoring and excessive daytime sleepiness (EDS). The patient had a history of renal lithiasis, dislipidemia and overweightness. She reported no use of chronic medication. The patient reported loud snoring, witnessed apneas and the occurrence of nocturnal arousals with asphyxiation feeling. She referred nightmares with occasional hypnopompic hallucinations. She denied cataplexy and sleep attacks. On physical examination, the body mass index was 32Kg/m^2, the cervical perimeter was 38cm and the abdominal perimeter was 96cm. The patient scored 13 in the Epworth Scale (ES). She brought the result of a cardiorespiratory sleep study that showed an apnea-hypopnea index (AHI) of 0.8 h with an oxygen desaturation (OD) of 4.7 h. Since there was a high clinical suspicion of OSAS, the study proceeded, performing pulmonary function tests and arterial blood gas that didn't reveal significant changes. She performed an echocardiography which revealed left ventricle global hypokinesia, with moderately depressed systolic function and ejection fraction of 32%, without changes in right ventricular function, consistent with the diagnosis of dilated cardiomyopathy. She performed a polysomnography that showed an AHI of 1.7 h, absence of significant peripheral desaturation and higher state of wakefulness. Before the result of the echocardiography, the patient was oriented to the Cardiology Consultation and began to take carvedilol 6.25 mg 1id and perindopril 4 mg 1 id. When the medication was initiated, there was a significant improvement in the patient's symptoms. She no longer referred the nocturnal arousals with the asphyxiation feeling and the hypnopompic hallucinations, and she was able to have a restful sleep, without EDS (ES: 5). The patient was discharged from the Sleep Apnea Consultation. Currently, the patient is being followed in the Cardiology Consultation and the etiology of the cardiomyopathy remains unknown.
Conclusion: The authors present this case report with the objective of highlighting the fact that not all of the patients that present snoring and nocturnal arousals have OSAS. In the case of this patient, the heart failure, undiagnosed in the past, was the cause of the paroxysmal nocturnal dyspnoea, described by the patient as arousals with feeling of asphyxiation. The fact of not having a restful sleep caused EDS with an elevated score in the ES. The performing of a polysomnography excluded OSAS and the disappearance of the patient's symptoms after the beginning of cardiomyopathy medical therapy reinforces the hypothesis that heart disease is the cause of the symptoms.

Keywords: *Obstructive sleep apnea syndrome. Nocturnal arousals. Heart failure.*

AGREEMENT STUDY OF THE FEV_1/FVC RATIO IN TWO DIFFERENT SEASONS, IN THE SCOPE OF THE STUDY "AIR QUALITY, EXPOSURE AND HUMAN HEALTH IN INDUSTRIALIZED URBAN AREAS (INSPIRAR)

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Introduction: Chronic obstructive lung disease (COPD) constitutes a public health problem and there are several spirometric diagnostic criteria. The agreement of these different criteria, taken at different times of the year, is not well established.
Aim: To evaluate the agreement of the fixed ratio (FEV_1/FVC<70%) and the lower limit of normal (LLN) criteria, between two measurements taken at different times of the year.
Methods: In the scope of the INSPIRAR study, we studied prospectively the same group of workers from Estarreja, in two different seasons of the year: June 2011 (V_1) and February 2012 (V_2). Spirometry with bronchodilation was performed to all participants. We evaluated the frequency of COPD, established by spirometric criteria, for each season. The considered criteria were the existence of a FEV_1/FVC ratio<70% and having an FEV_1/FVC ratio below the LLN, after administration of a bronchodilator. Agreement was assessed trough Cohen's kappa coefficient.
Results: A total of 116 workers were evaluated in both seasons, majority were males (85%), with a mean age of 44±10 years. The frequencies of FEV_1/FVC ratio<70% (V_1= 6.0%; V_2=5.2%) and FEV_1/FVC ratio below the LLN (V_1= 4.3%; V_2= 6%) did not differ in the two seasons. The intra-individual agreement between the two measurements was moderate for both spirometric criteria (for FEV_1/FVC<70%: kappa= 0.59; for FEV_1/FVC ratio below LLN: kappa= 0.47).
Conclusion: The frequency of COPD established through spirometric criteria, remained constant during the study. However, the moderate agreement suggests the existence of some degree of intra-individual oscillation between the two seasons.
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Keywords: *FEV_1/FVC. Agreement. COPD.*

IMPACT OF WEIGHT LOSS IN SPIROMETRY VALUES IN PATIENTS WITH BRONCHIECTASIS — REPORT OF 2 CLINICAL CASES

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Bronchiectasis are abnormal and irreversible dilatations of the bronchi and bronchioles. They may present with malnutrition, associated with high morbidity. However, overweight can lead to functional loss that, in patients with bronchiectasis, can add the one resulting from structural changes in the airways and parenchyma. The authors present two case reports of patients with bronchiectasis and weight excess which improved the spirometric parameters when significant weight loss was achieved.
Case 1: A 50-year-old woman, non-smoker, evaluated at the Pneumology department because of bronchiectasis in the context of a Swyer-James syndrome, presented, at the initial evaluation, weight excess, severe obstructive ventilatory syndrome with

negative bronchodilatation, global respiratory failure and absence of atopy. The chest CT showed cystic bronchiectasis in the left side, mainly in the left lower lobe. Therapy was optimized with inhaled bronchodilators and corticosteroids, nocturnal NIV, LTOT, ambulatory O2 and respiratory physiotherapy. The patient was seen by a nutritionist without favorable results, and so placement of intragastric balloon (IGB) was decided. Prior to the procedure, BMI was 44.6, and the spirometric and blood gases (FiO2 21%) values were the following-FVC 1.12 (40.6%), FEV_1 0.74 (31.6%), FEV_1/FVC 66%; pH 7.38, pO2 59.3 mmHg, pCO_2 54.6 mmHg, Sat 91.6%. After a significant weight reduction (BMI=35.7), the values were the following: FVC 1.79 (66%), FEV_1 1.02 (44%), FEV_1/FVC 57%; pH 7.44, pO2 77.7 mmHg, pCO_2 39.7 mmHg, Sat 94.3%. Nocturnal NIV and LTOT were suspended, but ambulatory O2 was maintained because of exercise desaturation.

Case 2: A 65-year-old woman, nonsmoker, was followed at the pneumology department because of bronchiectasis of unknown etiology, and she had a previous history of right bilobectomy (middle and lower lobes) at age of 30. At the initial evaluation the patient had weight excess, a mixed ventilatory syndrome with negative bronchodilatation test, absence of respiratory failure or atopy. The chest CT showed mild bronchiectasis in the right upper lobe. The patient began inhalation therapy and respiratory physiotherapy. Because of the respiratory and also musculoskeletal complaints, the patient began regular physical activity and a nutritional program for weight reduction. Previously, she had a BMI=29.3, and the following spirometric and blood gases (FiO2 21%) values-FVC 2.02 (78%), FEV_1 1.30 (60%), FEV_1/FVC 65%; pH 7.41, pO2 90.2 mmHg, pCO_2 41.4 mmHg, Sat 97.5%. After significant weight reduction (BMI=25.3), the values were the following: FVC 1.99 (81.6%), FEV 1.50 (73.9%), FEV_1/FVC 75.38%; pH 7.40, pO2 96.5 mmHg, pCO_2 39.8 mmHg, Sat 96.7%. Although in the first case it is not possible to exclude the influence of a possibly associated obesity hypoventilation syndrome (OHS) improved at the same time, the presented clinical cases are two examples of how weight loss can lead to a favorable shift of spirometric values and it may be interesting to assess its clinical and QoL impact in patients with bronchiectasis and weight excess.

***Keywords**: Bronchiectasis. Spirometry. Weight loss.*

EFFECTIVENESS OF HOME NON-INVASIVE MECHANICAL VENTILATION IN RESTRICTIVE THORACIC DISEASES

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Introduction: Non-invasive ventilation mechanical ventilation (NIMV) is appropriate and effective for the treatment of chronic respiratory failure due to chest wall diseases (CWD) and neuromuscular diseases (NMD). It has been shown that the application of NIMV prolongs survival, improves blood gases, normalizes the sleep pattern and increments the health-related quality of life in patients with restrictive thoracic disease.

Objectives: To characterize the CWD and NMD patients with home NIMV. Analyze the effectiveness of NIMV in the blood gas exchange, lung function parameters and utilization of health care resources.

Patients and methods: Retrospective study of NMD and CWD patients with home NIMV evaluated at sleep and noninvasive ventilation unit, during a 12-month period (from January to December 2011). The following parameters were analyzed: NIVM adherence, lung function tests, arterial blood gas, exacerbations with emergency department visits and hospital admissions.

Results: A total of 27 were included in this study, 15 patients with NMD and 12 patients with CWD. The mean age was 62.2±17.9 years. Sixteen patients were female. The mean length of NIVM treatment was 55.2±51.6 months. Before treatment with NIMV the mean PaO_2 was 57.4±12.42 mmHg, the mean $PaCO_2$ was 7.9±53.4 and mean excess bases (EB) was 9.7±6.6 mmol/L. The mean forced vital capacity was 48.7±18.9% of predicted and mean total lung capacity of 69.5±10.3% of predicted. The mean daily use of NIMV was 7.5±3.3 hours and the mean percentage of NIVM use over 4 hours per night was of 75.2±28%. There was a greater adherence to home ventilation in CWD patients. After NIVM treatment there was an improvement of blood gas exchange. There was an increase of the PaO_2 in 3.9 mmHg (P=.012), a reduction of the PaCO2 in 6.0 mmHg (P=.001) and a reduction of EB in 4.2 mmol/L (P=.004). The improvement of PaCO2 was higher in CWD patients. There has no improvement in lung function parameters. It was observed a reduction in the number of hospital admissions (from 0.7 per year to 0.3 per year, P=.009) and in the exacerbations with emergency room visits (from 1.5 per year to 0.8 per year, P=.001).

Conclusions: This group of patients with restrictive thoracic disease had a good adherence to home VMNI. The VMNI was effective in improving blood gas exchange and contributed to a greater clinical stability of these patients.

***Keywords**: Restrictive thoracic diseases. Non-invasive mechanical ventilation.*

FUNCTIONAL ASSESSMENT IN CARDIOVASCULAR FAILURE

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Introduction: The assessment of exercise cardiopulmonary function is the most objective way of classifying the severity of heart failure and of determining functional capacity. It allows verifying if there are other causes of limitation, to evaluate the efficacy of the therapeutic regimen and is essential in the evaluating candidates for heart transplantation.

Objective: To evaluate the indications for cardiopulmonary exercise test (CPET) and the abnormalities found in patients referred because cardiovascular failure.

Methods: Review of clinical data and CPET performed in the of respiratory pathophysiology laboratory of CHUC-HG from January 2009 to August 2012, analyzing the following parameters: underlying disease, indication of proof, results and contribution to decision clinic.

Results: We performed 43 CPET in patients with cardiovascular failure without complications. The referral was made mainly by Cardiology (88%). The main cardiovascular pathologies that motivated the CPET were: dilated cardiomyopathy (23%), pulmonary hypertension (21%), ischemic cardiomyopathy (19%) and heart failure (16%). We evaluated 33 patients, and 8 completed at least two or more consecutive assessments with a range from 5 months to a year to monitor clinical evolution. The main objective of the CPET was objective assessment of functional capacity (72%), preoperative assessment (12%) and evaluation prior to referral for cardiac transplantation in 14%. To assess functional capacity tests 31 CPET were performed (age 56±15 years), with cardiovascular limitation in 24 tests with VO2 of 14.0±4.5 mL/kg/min. In three, despite the suspected cardiovascular pathology, no limitation was found and in one only deconditioning was evident. In 2 cases the limitation was exclusive of gas exchange and ventilatory and in another the reason was articular pain. Six pre transplant CPTE were done (age 58±14 years) with VO2 of 11.2±2.4 mL/kg/min. Only 3 patients had VO2<10 mL /kg /min, with indication for heart transplantation. In two theraphy was optimize and subsequently repeated the test maintaining VO2<10 mL/kg/min. The VO2 peak was very variable in the different classes of NYHA functional capacity. Thus, patients in class I had VO2 between 10.5 and 28 mL

/Kg/min; class II VO2 between 6.4 and 21.4 mL/kg/min and Class III VO2 between 6 and 16 mL/kg/min.
Conclusion: CPET provided a comprehensive and safe assessment for patients with cardiovascular disease, even when severe. In some cases pointed out other causes of exercise limitation, being advantageous the cooperation between Cardiology and Pulmonology. It also enabled an accurate determination of objective functional capacity, better than NYHA subjective classification and identified patients in need of optimizing therapy or pointed towards referral to cardiac transplantation.

Keywords: *Cardiopulmonary exercise test. Functional capacity. Cardiovascular failure.*

ASTHMA CONTROL WITH OMALIZUMAB, OUR CLINICAL EXPERIENCE

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Introduction: It is estimated that approximately 5% of asthma patients have severe asthma. The GOAL study demonstrated that 38-53% of patients using "optimal therapy" with high doses of inhaled corticosteroid (ICS), long-acting B-agonist (LABA) and leukotriene antagonists remain poorly controlled. These patients suffer frequently from asthma symptoms with a major impact on their daily activities and often experience exacerbations with multiple hospital visits. The Global Initiative for Asthma (GINA) 2011 guidelines recommend omalizumab as an add-on step 5 treatment to ICS and LABA therapy. In INNOVATE Omalizumab reduced asthma exacerbation rate by 26% and halved the severe exacerbation rate in patients. Emergency visits were reduced by 44%. Omalizumab has also been found to significantly improve asthma-related QoL, asthma symptom scores, and lung function. In EXCELS Omalizumab treatment decreased doses of ICS, short acting B-agonists (SABA) and Leukotriene receptor antagonists. Studies have also shown a steroid sparing effect of Omalizumab.
Aim: Evaluate the efficacy of Omalizumab on asthma control as an add-on therapy on patients from the "Severe Asthma Outpatient Unit" of Hospital Pulido Valente, Lisbon.
Methods: This is an observational study, conducted since the introduction of omalizumab as an add on therapy in selected patients of the Severe Asthma outpatient unit. Patients eligible for treatment with omalizumab were adult patients with severe asthma not completely controlled with optimized standard therapy, with serum IgE 30-700 and with a positive skin prick test for at least one perennial aeroallergen. Information was collected from patient clinical records dating from the year prior to initiation of omalizumab and at 6- 12- and 24 months of treatment. Asthma control was determined from medical records by applying the ACT score and according to GINA guidelines. The rate (number per year) and severity of exacerbations (moderate or severe according to ATS recommendations for asthma clinical trials) were recorded at each time interval as was ongoing asthma maintenance medication. FEV_1 results were collected from available lung function tests. Data was analyzed using the SPSS 15 version and was tested for significant differences at each time interval.
Results: A total of 26 patients were under treatment with omalizumab, all of which classified by GINA with uncontrolled asthma prior treatment. Mean ACT score was 11,5. All patients had treatment with fixed-dose ICS and LABA. 42.3% needed treatment with oral glycocorticosteroids for control. Mean IgE before omalizumab was 160.7 (±121.1) and mean FEV_1 was 57.07%. Patients reported an average of 1.8 moderate and 3.1 severe exacerbations/year. Statistical differences in asthma control were found at 6 months follow-up in most end-points: GINA score improved: 60.9% of patients had partially controlled asthma and only 39.1% with uncontrolled asthma (Wilcoxon 0.00); ACT score improved to 19.52 (Wilcoxon 0.00); mean FEV_1 improved to 76.7% (Wilcoxon 0.025); the proportion of patients requiring oral glycocorticosteroid therapy reduced to 17.4% (Wilcoxon 0.014); and the number of moderate and severe exacerbations also decreased to 1.04 and 1.83 respectively (Wilcoxon 0.007; Wilcoxon 0.002). This study corroborates published evidence on improvement in asthma control therapy with omalizumab.

Keywords: *Severe asthma. Omalizumab. Anti-IgE. Asthma control. Asthma treatment.*

RATE OF VACCINATION AND IMPACT OF IMMUNIZATION AGAINST SEASONAL FLU IN A GROUP OF ASTHMATIC PATIENTS

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Introduction: Influenza is a highly contagious viral disease, usually benign and self-limiting in healthy individuals but a cause of considerable morbidity in susceptible individuals and patients with asthma and other chronic diseases. The best treatment is prevention being asthmatics advised to make annual vaccination. There is no known vaccination rate of patients with asthma in Portugal. In 2010/2011 (latest year for which they are known vaccination rates in Portugal) the overall vaccination rate was 17.5% and 28.8% in patients with at least one chronic disease. The recommendation by the physician is the factor that most influences adherence to vaccination (Table 1).

Table 1

	Vaccinated	Non vaccinated	Total	
Total	130 (86.1%)	21 (13.9%)	151	
Mean age	48.2	39.8	47	*P*=.03
Female sex (%)	73.1	90.5	75.5	*P*=.1
Vaccine reaction (%)	2.1			
Flu syndrome (%)	9.3	16.7	10.2	*P*=.4
Exacerbations-% (0/1/2/3/4)	79.8/11.6/6.2/1.6/0.8	77.8/11.1/0/5.6/5.6	79.6/11.6/5.4/2.0/ 1.4	
Change in usual therapeutic	10.1	16.7	10.9	*P*=.4
System corticosteroids	7.0	11.1	7.5	*P*=.6
Visit to Primary care center	7.0	0	6.1	*P*=.6
Visit to ER	2.3	11.1	3.4	*P*=.1
Inpatient treatment	0.8	5.6	1.4	*P*=.2

Objectives: To evaluate the rate of vaccination in patients with asthma and adherence to the proposed vaccination; assess the impact on exacerbations, changes in usual therapeutic and non-scheduled visits to health services.
Methods: Prospective study in 151 consecutive patients with asthma and older than 12 years seen in the pre-season (15/09 to 30/10/2011). We proposed the flu vaccine to all patients regardless of asthma severity and age. Those who refused were provided information on WHO recommendations and counseling and were subsequently asked if they change their mind. The remaining parameters were evaluated in the post season (April-May 2012).
Results: Of the studied patients 113 (74.8%) were interested in having the flu vaccine. Of the 38 (25.3%) who were not initially interested 17 (44.7%) changed their minds which were translated to a vaccination rate of 86.1%. We observed a low rate of vaccine reaction (pain at the injection site). Table 1.
Comments: The vaccination rate in this group of patients with asthma and with a mean age below 50 years is much higher than the overall rate of vaccination in patients with other chronic diseases. The mean age of patients vaccinated is statistically higher than in patients who refuse the flu vaccine. Although not statistically significant, non-vaccinated patients have a higher rate of flu syndrome, changes in chronic therapy, need for systemic corticosteroids, visit to the emergency department and inpatient admittance and lower rate of use of primary health care services, revealing a worse disease control and greater resource consumption. Informed patients will adhere to vaccination regardless of asthma severity and can achieve better asthma control.

***Keywords**: Asthma. Seasonal flu vaccination.*

PARADOXICAL RESPONSE TO SALBUTAMOL — CASE STUDY

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One of the integrated tests in lung function assessment is the bronchodilator reversibility test. This test evaluates airway reversibility and the response to therapeutics. Short acting beta-agonist bronchodilators (BD) are used in the relief of symptoms and bronchospasm in obstructive airway diseases and their side effects include tachycardia, shaking and headache, and very rarely hypersensitive reactions, cardiac arrhythmia and paradoxical bronchospasm.
Case report: A patient, female, 28 years old, was observed in pulmonology consultation for worsening of asthma. At the time of consultation the patient presented with daily nocturnal wheezing, wheezing with exercise, in contact with dust and temperature changes. It was prescribed lung function tests (LFT) and formoterol, fluticasone propionate and montelucaste. During the LFT, the results indicated an airway obstruction of mild severity with pulmonary hyperinflation. During the pause after the BD, the patient presented with signs of respiratory distress such as increasing wheezing and cough. The LFT results after BD (salbutamol 4×100 μg) indicated and airway obstruction of moderate severity with a proportional increase in pulmonary hyperinflation and airway resistance.
Discussion: With the purpose of evaluating the worsening of asthma symptoms, LFT were prescribed. During the test, a few minutes after BD the patient presented signs of respiratory distress such as cough, wheezing and mild dyspnea. The LFT after BD confirmed the signs, presenting a worsening of airway obstruction regarding the initial assessment. After the BD the expected results are the dilation of the bronchial smooth muscle. In this specific case there was a worsening of the obstructive pattern, therefor it was designated as paradoxical bronchoconstriction. The paradoxical response seems to be triggered by a worsening of inflammation and induction of airway hyperreactivity. The mechanisms that trigger this response seem multifactorial, and currently not very well understood. Several bronchospasm mechanisms have been proposed, besides the reaction to the active agent, such as: reaction to the excipient, mediated by immunoglobulin E; indirect irritation to propellants, such as oleic acid and chlorofluorcarbons (CFC), and preservantes; irritation because of turbulent flow due to inappropriate inhalation technique and bronchial irritation caused by acidity and hyperosmolality of the solution (nebulization). Fatigue may occur during LFT and cause bronchospasm, but this is not a paradoxical bronchoconstriction.
Conclusion: Paradoxical bronchoconstriction, although rare, has been known to happen. Several mechanisms have been admitted to interfere with this airway modulation of bronchial reactivity and consequent paradoxical response. It is of the utmost importance to be aware of this phenomena, and the correct way to approach it, besides the lack of published references.

***Keywords**: Bronchodilation test. Salbutamol. Paradoxical bronchospasm.*

IMPULSE OSCILLOMETRY IN THE CLINICAL PRACTICE-UTILITY IN ASTHMATIC PATIENTS

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Introduction: Impulse oscillometry (IOS) is a non-effort-dependent pulmonary examination that provides information about airway resistance and reactance of the bronchial tree. It has been performed alongside with other methods in the study of pulmonary function, and it has special interest as a complement of spirometry in the evaluation of obstructive ventilatory disorders.
Objective: The aim of the study is to evaluate the importance of IOS as a complementary method to spirometry in the evaluation of pulmonary function of asthmatic patients.
Methods: Asthmatic patients followed in the Respiratory Allergy consultation in HG-CHUC were evaluated, from January to August 2012. These patients were divided into three groups, according to the spirometric results obtained during periods of clinical control of the disease: Group A-no spirometric changes; Group B- large airways obstruction ($FEV_1/FVC<70$ and $FEV_1<80\%$); and Group C-isolated small airways obstruction ($FEV_1/FVC>70$; $FEV_1>80\%$, and FEF 50%<80%). For each one of these groups, clinical and demographic data, smoking habits, atopy status and coexistence of rhinitis, as well as oscillometric data (peripheral and central resistances-PR and CR) were obtained.
Results: One hundred and six patients were included (Group A-42; Group B-41 and Group C-23), most of them females (A-81%; B-88% e C-90%). The average ages in years were 29.4±14.4; 54.1±15.1 and 45.9±16.4 in A, B and C groups, respectively. In group A, 14.28% of patients had no increase in airway resistance evaluated by oscillometry. From the 85.72% which had oscillometric changes, 58.3% had elevated PR and CR (6.8±2.20 and 4.02±0.68 cmH2O/(L/s), respectively); 30.5% had isolated PR elevation (3.86±0.70 cmH2O/[L/s]), and 11.1% isolated CR elevation (4.21±0.72 cmH2O/[L/s]). In group B, 14.63% asthmatics had no elevated airway resistances. Among those who had oscillometric changes, 68.5% had elevated PR and CR (7.43±2.33 and 3.92±0.64 cmH2O/[L/s], respectively); 28.5% had isolated PR elevation (5.50±2.68 cmH2O/[L/s]), and 2.8% isolated CR elevation (3.89 cmH2O/[L/s]). In group C, the evaluated resistances were normal in 30.4% of the cases. Among the remaining, 62.5% had elevated PR and CR (7.08±2.52 and 3.78±0.71 cmH2O/(L/s), respectively), 25% had isolated PR elevation (4.08±0.41 cmH2O/[L/s]), and 12.5% isolated CR elevation (3.34±0.01 cmH2O/[L/s]).

Conclusion: Despite the relatively small cohort of patients, this study showed that in asthmatics whose spirometries were normal, IOS was a useful test, since it revealed the presence of obstructed airways (not detected by conventional study) in a significant percentage of patients. In asthmatics whose spirometries showed an obstructive ventilatory pattern, the IOS results were concordant in most cases, not having provided additional information. In these cases, the execution of IOS should be carefully weighted.

Keywords: *Asthma. Impulse oscillometry.*

MECHANICAL VENTILATION FOR THE TREATMENT OF EXCESSIVE DYNAMIC AIRWAY COLLAPSE IN A PATIENT DIAGNOSED WITH ASTHMA

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A 54 years-old Caucasian female, diagnosed with intrinsic asthma for about 20 years, and gastroesophageal reflux disease, without history of smoking habits or allergies, presents with dysphonia, progressive dyspnea and wheezing exacerbated with exercise and at night. Physical examination showed tachypnea, stridulous breathing and only slight expiratory wheezing at pulmonary auscultation. There was no significant respiratory functional repercussion. Thoracic CT did not show relevant findings. She performed flexible bronchoscopy, revealing pronouncedexpiratory airway collapse(>50%) of the posterior membranous wall of the trachea and bronchial tree bilaterally. Excessive Dynamic Airway Collapse (EDAC) was diagnosed. In the following years, the patient had several exacerbation needing invasive mechanical ventilation support. We decided to start nocturnal Continuous Positive Airway Pressure (CPAP) applied through an oronasal mask. After a period of relative stability, it was introduced Non-Invasive Ventilation (NIV) delivered via bi-level positive airway pressure (BiPAP) following a new hospitalization in intensive care unit because of difficult weaning from mechanical ventilation. Afterwards, she presented progressive dependency to home NIV. Since she was on continuous NIV trough full-face mask, we proposed tracheostomy. The patient was well adapted to bi-level airway pressure ventilation through a tracheostomy cannula, without needing supplementary O_2 and obtaining a gasometric improvement. During additional assessment, she was diagnosed with scleroderma, without criteria of pulmonary involvement. EDAC is a rare entity, which treatment is not consensual. The authors discuss the therapeutic options and the uncertain clinical significance of association to asthma or COPD.

Keywords: *Mechanical ventilation. Excessive dynamic airway collapse. Asthma.*

ENDOBRONCHIAL HAMARTOMA

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Introduction: Hamartoma is one of the most common benign tumors of the lung. Most hamartomas are located in the lung parenchyma, and only rarely they occur within the bronchi. Most endobronchial tumors produced symptoms, such as cough and fever, due to bronchial obstruction.
Aim/methods: The authors report the case of a hamartoma of the right superior bronchus diagnosed and removed by bronchoscopy.
Results: A 50-year-old male patient was admitted to the hospital complaining of cough and progressive shortness of breath. He was no smoking and had diabetes. In physical examination there were reduced lung sounds in the superior half of the right hemithorax. Chest-X-ray revealed right upper lobe atelectasis. Chest CT demonstrated endobronchial nodular image located in the right upper lobal bronchus. On flexible bronchoscopy, an endobronchial polypoidal mass with well limited surface was observed nearly totally obstructing the right upper lobal bronchus. A biopsy specimen was taken and the patient was diagnosed with chondroid hamartoma. The patient underwent a rigid bronchoscopy with argon application and debulking. No complications were noted. At 3 month follow-up, a bronchoscopic examination showed no residual mass lesion and no tumor recurrence.
Conclusion: Endobronchial hamartoma is a special and rare form of the usual intrapulmonary hamartoma, with originates from a large bronchus and grows into the lumen. The management of endobronquial hamartoma must be individualized according to the characteristics of each patient and each hamartoma. Endoscopic treatment (argon, laser, cryoablation, electrocautery) is less invasive modality than thoracotomy and is safe and effective therapeutic option.

Keywords: *Endobronchial hamartoma. Bronchoscopy.*

TRACHEAL DIVERTICULUM: A RARE ENTITY

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Introduction: Tracheal diverticulum is a rare clinical entity defined as an air filled paratracheal lesion which results from a congenital or acquired weakness of the tracheal wall. Although mostly assymptomatic they can also be the cause of chronic cough, repeated respiratory infections or even unsuccessful intubations. Diagnosis is based on radiological findings, namely by CT scan.
Case report: The authors present a case of a 52-year old man referred to their pneumology consult by general doctor because of chronic cough that the patient did not value. He denied any other symptom. He was a heavy smoker (68 pack-year) with no other personal history of patological conditions. He denied otorhinolaryngological surgeries and/or endotracheal intubation. The physical examination was normal. Pulmonary function tests revealed a mild obstructive pattern, with slight post-bronchodilator change. Chest radiography was normal. The chest CT showed four traqueal diverticula emerging from the right postero-lateral tracheal wall with diameters between 3mm and 22mm. No other relevant lesions were reported. Fiberoptic bronchoscopy revealed three small orifices in the right postero-lateral tracheal wall (same localization referred in CT scan). Because of the small dimensions it was impossible entering the orifices. The rest of the bronchial tree presented many mucoceles. No other significant lesions were found. The patient was submitted to conservative treatment with bronchodilator and mucoytic agents and encouraged smoking cessation.
Discussion: Tracheal diverticula are frequently incidental radiographic or CT findings because of being usually asymptomatic. However in some cases they can be clinically relevant causing compression-of-trachea symptoms or acting as a reservoir for secretions, with secondary chronic infections. This may be manifested by chronic cough, dyspnea, stridor, and repeated episodes of tracheobronchitis. As in the case described, tracheal diverticula are often found posterior to and slightly to the right of the trachea. This is probably due to the supportive effect of the esophagus on the left side. The diagnosis is based on CT scan of the trachea. Bronchoscopy can also be very useful to identify the orifices of the diverticula, however many have a narrow opening or just a fibrous connection with the trachea and are bronchoscopically

missed. In the case described identification of orifices of the diverticula was possible. COPD is frequently associated with this entity and should be diagnosed and treated. In general, as in the case presented, conservative medical treatment with bronchodilators, mucoytic agents antibiotics is proposed, leaving surgical treatment for larger-sized or symptomatic diverticula.

***Keywords**: Tracheal diverticulum. Rare entity. Bronchoscopy.*

UTILITY OF BRONCHOALVEOLAR LAVAGE GALACTOMANNAN: THREE CASE REPORTS

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Galactomannan is a heat-stable polysaccharide found in the fungal wall of most *Aspergillus* species. It´s detection in bronchoalveolar lavage (BAL) has the potential to provide evidence of invasive aspergillosis, and has proven to be more sensitive than serum testing. However, uncertainties about what may cause false-positive results compromise the utility of this method. We report 3 cases in which positive BAL galactomannan was found. Two patients had severe immunosuppression caused by HIV infection, while the third had mild immunodepression due to diabetes. Although the cases differ in terms of clinical and radiologic findings, in all tree pulmonary aspergillosis was considered as a possible diagnosis. In all patients serum galactomannan was negative and no fungal species were found in BAL or other biological samples. Positive galactomannan antigenium in BAL performed by enzyme imunoessay (cut-off >0.5) lead to the diagnosis of pulmonary aspergilosis and initiation of antifungal therapy. In one case the further isolation of *Rhodococcus equi* in culture of the BAL, led us to questioner the diagnosis, and consider that the galactomannan was a false-positive result. These reports are an example of the clinical utility of the detection of galactomannan in BAL. It´s use should be considered whenever there is clinical suspicion of invasive pulmonary aspergillosis, especially in immunocompromised patients. Nonetheless, test´s specifity remains a problem. The increase of the cut-off from >0.5 to >1.0 to increase performance has been suggested in the literature, but further investigations in this area is needed.

***Keywords**: Galactomannan. Bronchoalveolar lavage. Aspergillosis.*

CLINICAL CHALLENGE OF PERSISTENT PULMONARY AIR-LEAKS

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Introduction: Air leaks are a common problem after pulmonary resection and can be a source of significant morbidity and mortality.
Case report: We describe a clinical case of a 68-years old male patient with pulmonary adenocarcinoma that underwent pulmonary resection. After the procedure he presented with very difficult to manage persistent air-leak. Diverse approaches failed to resolve, including pleurodesis, implantation of endobronchial one-way valves on the bronchial segments identified using systematically occlusion of the bronchial segments, decortication and thoracomyoplasty. During that time the patient remained in the hospital with a chest tube. At this point another endobrochial approch was attempted for the implantation of endobrochial valves. The bronchial segments involved in the air-leak were identified by instillation of methylene blue trough the chest tube allowing the identification of two bronchial segments. One of which non identified by the previously used balloon method technique. A sufficient air-leak's magnitude reduction was achieved allowing the patient's chest tube to be removed.
Comment: Nonsurgical approaches appear promising and, for some patients may be the only treatment option after all conventional treatments have failed or are associated to high risk.

***Keywords**: Pulmonary air-leaks. Pulmonary resection. Endobrochial valves.*

MEDIASTINAL BRONCHOGENIC CYST-MULTIDISCIPLINARY APPROACH TO DIAGNOSIS AND TREATMENT

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Introduction: The bronchogenic cyst (BC), although rare, is the most frequently cystic lesion in the mediastinum. The diagnosis in adulthood is rare and most symptoms are dependent on compression of adjacent structures.
Aim: To describe a patient with mediastinal bronchogenic cyst, with relevant diagnosis and treatment.
Results: A 48 years old patient, female gender, non-smoker, with history of depression, presented with insidious onset of irritative cough and dyspnea, both worsened with supine position, occasional uncharacteristic chest pain and wheezing. A chest-CT showed a pseudonodular pretracheal image, measuring 3.1cm across. Endobronchial ultrasound (EBUS) was performed, showing a decrease in distal trachea's caliber due to extrinsic compression; transbronchial needle aspiration of a non-pure hypoechoic pretracheal lesion was performed, without producing a definite pathological result. Given the suspicion of BC, mediastinoscopy was held, with identification and removal of a well-demarcated cystic lesion, complicated by tracheal collapse caused by invasion and destruction of cartilaginous rings. The intraoperative bronchoscopy showed collapse of the trachea's anterior wall and right main bronchus. Conversion to thoracotomy allowed complete removal of the BC and reconstruction of the tracheobroncheal tree. Follow-up fibroscopies confirmed good anastomosis integrity and tracheobroncheal patency.
Conclusion: The variability in symptoms, localization, the challenging differential, the various diagnostic and therapeutic options involved, the management of possible complications and the follow-up, favor a multidisciplinary approach in bronchogenic cyst.

***Keywords**: Bronchogenic cyst. Bronchoscopy. Mediastinoscopy. Thoracotomy.*

BRONCHIAL ASPIRATE CYTOLOGY STUDY AND BRONCHIAL BIOPSY IN THE DIAGNOSIS OF LUNG CANCER-A RETROSPECTIVE STUDY

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Introduction: Fiberoptic bronchoscopy is a minimally invasive procedure with diagnostic applications in various thoracic pathologies. It acquires extreme importance in the diagnosis of lung cancer, particularly in the differential diagnosis of imagiological changes.
Objectives: To evaluate the diagnostic yield of the cytological analysis of bronchial aspirate and bronchial biopsies performed by fiberoptic bronchoscopy, in patients with radiological images suggestive of lung cancer.

Methods: Retrospective file analysis of patients submitted to fiberoptic bronchoscopy in a two year period, and assessment of the changes detected, demographic data, diagnostic hypotheses and cytological and histological results.
Results: 186 fiberoptic bronchoscopies were perfomed during the two year period, with a mean age of 65.5 years and a standard deviation of 13.44. There was a male predominance, with 139 patients (74.7%). In 152 patients (81.7%) bronchial biopsies were performed, which allowed us to obtain a histological diagnosis in 94 (61.8%) cases. The initial suspicion of lung cancer was confirmed in 88 (58%). The patients with a confirmed diagnosis of cancer had a mean age of 65.5 years and a standard deviation of 13.19, maintaining the predominance of men (79.5%). Within the histological diagnoses obtained, there was a predominance of Adenocarcinoma (33%), Epidermoid carcinoma (17%), Small cell carcinoma (14.8%) and Carcinoid Tumor (14.8%). In 41 patients (22%) it was performed another technique to obtain histological diagnosis, which confirmed lung cancer in 20 of these patients. The analysis of the endoscopic abnormalities identified the presence of indirect and direct signs of infiltration of the mucosa in 86% of patients with confirmed diagnosis of neoplasia, with particular emphasis on mucosal edema, convergence of mucosal folds, stenosis associated with partial or total obstruction of the bronchial lumen. As a consequence, a statistically significant relationship between the presence of endoscopic changes and the histologic diagnosis of neoplasia by bronchial biopsy was confirmed ($P<.000$). The cytological analysis of bronchial aspirate identified neoplasic cells in only 24 patients (13%), which proved a low profitability. However, despite the endoscopic changes, in eight patients who had insufficient or negative bronchial biopsy, cytological analysis led to a diagnosis without the need for more invasive diagnostic techniques.
Conclusions: This study supports the role of bronchial biopsies in patients with suspected lung cancer. The cytological analysis of bronchial aspirate proved to be insufficient, leading to a diagnosis only in 13% of cases, value below the ones described in recent literature. However, the combination of cytological and histological techniques may significantly increase the yield of bronchoscopy in the diagnosis of lung cancer.

Keywords: *Bronchial biopsies. Cytology of bronchial aspirate.*

A CASE OF ENDOBRONCHIAL HAMARTOMA TREATED WITH ENDOSCOPIC RESECTION

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Benign tumors represent less than 1% of lung neoplasms. Hamartomas, despite being the most common benign tumor of the lung, are usually located in the lung parenchyma, being the endobronchial presentation a rare form. Early diagnosis and treatment is important since they can cause irreversible pulmonary destruction due to bronchial obstruction. In recent years, endoscopic resection is becoming the treatment of choice for these type of lesions. We report a case of a 59-year-old men, who presented with a history of persistent cough in the past two years, and dyspnoea when lying in his left side. Flexible fiber-optic bronchoscopy revealed a polypoid mass obstructing the orifice of the left main bronchus. Biopsy specimens were consistent with chondroid hamartoma. Rigid bronchoscopy was performed, and a combination of argon plasma coagulation, laser YAG and mechanical debulking was used to completely excise the tumor. Endoscopic revaluation showed no signs of tumor. This case is an example that endoscopic resection is a safe and effective method for treating selected cases of benign tumors of the airway.

Keywords: *Hamartoma. Bronchoscopy. Endoscopic resection.*

ENDOBRONCHIAL LESIONS EVALUATION

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Introduction: Fiberoptic bronchoscopy is the ideal modality to evaluate the bronchial tree and its associated techniques are essential for the diagnosis of endobronchial lesions.
Objective: to determine the frequency of endobronchial lesions, etiology and the diagnostic yield of the different endoscopic techniques in malignancy.
Methods: Retrospective study, by review of clinical files of patients with endobronchial lesions who performed bronchoscopy between January 2009 and August 2012.
Results: A total of 76 patients was enrolled, 76.3% male, with a mean age of 66.1 years. Sixty five patients (85.5%) underwent bronchoscopy for initial diagnosis and 11 (14.5%) had already known endobronchial involvement and underwent bronchoscopy due to new symptoms. In the first group of patients the most common etiologies were primary lung cancer (n=46; 70.8%), secondary cancer (n=4; 6.2%) and foreign bodies (n=4; 6.2%); in 10 patients (15.4%) the diagnosis remained undetermined. The most prevalent malignancies were squamous cell carcinoma (47.8%; n=22) in primary lung cancer and colon adenocarcinoma in secondary cancer (n=3; 75%). The lesions were predominantly located in the right inferior and left superior lobar bronchus (both with n=12; 18.5%). Sixty seven per cent (n=33) of the lesions were seen on thoracic CT. In malignancy bronchoscopy had a diagnostic yield of 86.0% (n=43). Bronchial biopsies had the best diagnostic yield-86.0% (n=43); cytology of bronchial aspiration and brushing had inferior diagnostic yield with 22.0% (n=11) and 32% (n=8) respectively.
Conclusion: The most common etiology of endobronchial lesions was primary lung cancer, squamous cell carcinoma being the most prevalent. Bronchial biopsies had the best diagnostic yield. The diagnostic yield of citology of bronchial aspiration and brushing was inferior and did not improved the overall diagnostic yield of bronchoscopy.

Keywords: *Endobronchial lesion. Bronchial biopsies. Diagnostic yield.*

VALUE OF A NEGATIVE RESULT IN CT-GUIDED TRANSTHORACIC LUNG BIOPSIES

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Introduction: CT-guided transthoracic lung biopsies (TTLB) have a high specificity for the diagnosis of malignant tumors. However, with a negative result we cannot reliably exclude a malignant lesion.
Objective: To evaluate the final diagnosis of patients with lung lesions whose initial result of CT-guided TTLB was not diagnostic.
Methods: Retrospective study of results of CT-guided TTLB in the study of lung lesions, in a 4-year period (2007 a 2010). Results were classified as: positive for malignancy, benign specific, and negative for malignancy and insufficient material for diagnosis. Non-diagnostic results were reviewed and related to a definitive diagnosis obtained by surgery or clinical and imagiological follow-up.
Results: We performed 162 TTLB, in patients with an average age of 62.5 years, 73.5% male, 42% non-smokers, 29% with prior respiratory disease. Eighty-three biopsies (51.2%) were positive for malignancy, 13 (8%) benign specific, 9 (5.6%) negative for malignancy and 57 (35.2%) insufficient for diagnosis. Of the non-diagnostic biopsies (negative for malignancy and insufficient material for diagnosis),

59 (89.4%) obtained subsequent definitive diagnosis. Malignancy was confirmed in 43/66 cases (65.2%) and benign specific in 16/66 cases (24.2%). The malignant tumors confirmed were: lung adenocarcinoma (20), squamous cell lung carcinoma (5), NSCLC (6), pulmonary metastasis from other organs (7), other tumors (5). Benign diseases diagnosed were: pulmonary tuberculosis (7), bacterial pneumonia (3), amyloidosis (2), organizing pneumonia (1), other diagnosis (3). The negative predictive value for malignancy, in this sample, was 34.8%.
Conclusion: In this sample, there was a high rate of malignancy in patients with lung lesions without initial diagnosis by CT-guided TTLB, so the performance of additional testing in these patients is fundamental.

Keywords: *Transthoracic lung biopsy. Diagnosis. Malignancy.*

TRANSTHORACIC BIOPSY - A DEPARTMENT'S STATISTIC

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Introduction: The percutaneous biopsy guided by computed tomography (CT) has been widely used as an effective and safe procedure for obtaining a histologic diagnosis in many clinical situations and in different organs. In the lung, percutaneous biopsy has become a major diagnostic option for nodules and masses. Allows access to lesions in various locations of the lung, and can be used for peripheral or more central lesions of various dimensions.
Objectives: To statistically characterize CT-guided transthoracic biopsies (BTT) performed in Imagiology Department during the year 2011.
Methods: Retrospective analysis of the clinical files of all patients undergoing this exam during the period specified.
Results: We performed a total of 58 examinations in 55 patients with a mean age of 63.5 years (38-81), mostly men (75%). 55% of the lesions were masses, 38% nodules and 7% areas of diffuse interstitial lesions. Were carried out 9 "core biopsy", 40 aspiration biopsies and 9 exams with both techniques. To perform the aspiration technique we used 22G or 25G needles. The exams performed with the 22G needles: 58% were inconclusive and 42% allowed the diagnosis; 25G needle: 20% were inconclusive and 80% allowed diagnosis. 72% of the exams didn't have immediate complications. Of the 28% with immediate complications there were 2 cases of hemoptysis, 2 of local hemorrhage and 12 small volume pneumothorax. Note that 37.5% of the complications were registered with 25G and 25% with the 22G needle. There were no 24 hours complications. The most frequent diagnostic was lung adenocarcinoma (36%), followed by squamous cell carcinoma, metastasis to the lung and pulmonary TB (7% each diagnosis), interstitial disease and non-small cell carcinoma (3.5% each) and other diagnoses (4%). 32% of the results were inconclusive and the final diagnosis was obtained by a new transthoracic biopsy or surgical biopsy.
Conclusions: The results obtained are generally coincident with the international bibliography, differing in the diagnostic accuracy. In this case the registered accuracy was lower than the international references (68% vs 80%). The accuracy varies depending on the location of the lesion, its size, the type of needle used, the number of samples performed and experience of the operator. The number and type of complications is coincident with the international benchmarks being the small volume pneumothorax the most frequent (21% vs 20%). Transthoracic biopsies are an indispensable procedure. Improvements in imaging techniques, the needles and the operators' experience made this procedure more versatile, safe and effective, progressively expanding its indications.

Keywords: *Percutaneous biopsy. Transthoracic lung biopsy.*

CATAMENIAL PNEUMOTHORAX. 5-YEAR REVIEW

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Introduction: Catamenial Pneumothorax (CP) is a relatively rare entity and the pathophysiology is poorly understood, although some theories have been proposed. It is defined as spontaneous pneumothorax occurring within 72 hours before or after onset of menstruation. Therapeutic options consist of surgical exploration with resection of endometrial deposits within the pleural space and/ or repair of diaphragmatic defects (if found) with or without pleurodesis. Non-surgical options include hormonal suppression.
Aim/methods: The aim of this study was to determine the prevalence of CP in our unity. We analyzed records of all female patients operated for spontaneous pneumothorax at our unity in 5 years.
Results: Fifty seven women with spontaneous pneumothorax were selected for surgical management. The mean age was 32.1±10 years. Most of these have been secondary to emphysema (61.4%). In 6 patients (10.5%) pneumothorax was cathamenial. All 6 patients were pre-menopausal, with a mean of 32.7±7.7 years. These 6 pneumothoraces were unilateral and right-sided. Five women had thoracic endometriosis and 1 has diaphragmatic lesions. All 6 patients underwent thoracoscopy and surgical pleurodesis was performed in all patients. There was no surgical complication.
Conclusion: CP should be suspected in any menstruating woman presenting with recurrent spontaneous pneumothorax, especially if it is on the right side. The present study demonstrates an incidence of CP of only 10.5% in women suffering from spontaneous pneumothoraces. Other studies suggest that the incidence is much higher. The treatment for CP is directed towards the management of the pneumothorax and the prevention of its recurrence.

Keywords: *Catamenial pneumothorax. Thoracoscopy.*

CATAMENIAL PNEUMOTHORAX

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Catamenial pneumothorax is a clinical entity consisting of recurrent menses associated pneumothoraces. It is usually seen as a rare pathology but some studies report it may account for up to a third of all spontaneous pneumothoraces in women referred for surgery. The etiology is not clearly defined but it is usually associated with pulmonary endometriosis or diaphragmatic defects with transabdominal-transdiaphragmatic passage of air during menstruation. We describe two cases. The first patient was a 43-year-old woman with previous history of asthma and allergic rhinitis and with two previous uneventful pregnancies. The patient had an episode of sudden pain on her right upper hemithorax on the first day of her menses with slight dyspnea so she went to the emergency department and the X-ray confirmed a moderate right pneumothorax, which was treated conservatively. Further investigation showed that she had had a similar episode on the first day of her previous menstruation. Four months later another episode of pneumothorax was confirmed during her menses. She was then referred for surgery. She was submitted to a right, video assisted thoracoscopy (VATS) that showed multiple small perforations in the diaphragm that seemed to be the origin of her pneumothoraces. A talc pleurodesis was performed, especially on the diaphragmatic pleural surface. No endometrial nodules were found. Her last re-evaluation was four months later and she was

clinically well with no signs or symptoms of recurrence. The second patient was a 29 year-old, ex-smoker (2 packs-year), that had a significant right pneumothorax diagnosed, coinciding with the beginning of her menses that required drainage. She then had a similar episode, requiring drainage, one year later. The patient later admitted having right-sided thoracic pain almost every month during her menses, but never complained to her family doctor. The chest CT showed diaphragmatic herniation on the right side. She was submitted to a right VATS, which showed a circular diaphragmatic defect with liver herniation. Diaphragmatic suture and talc-pleurodesis was performed. She was last evaluated five months later with no signs of herniation or pneumothorax. This phenomenon is many times under-diagnosed and might explain recurrence of pneumothoraces in many women. Referring for Thoracic Surgery evaluation is the appropriate approach even though the optimal treatment has not been defined.

Keywords: *Catamenial pneumothorax. Diaphragmatic defects. VATS.*

SPONTANEOUS HYPERTENSIVE HEMOPNEUMOTHORAX

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Spontaneous pneumothorax is a common pathology, especially affecting young males, often smokers, requiring prompt evaluation and treatment. In certain cases it may evolve to a hypertensive pneumothorax, which is a medical emergency. Spontaneous hemopneumothorax is a rare disorder. We describe a case of a 23-year-old young man that went to the emergency department with complaints of abdominal pain, nausea and shortness of breath, which had been present for at least four days but had worsened in the previous few hours. A hypertensive hidropneumothorax with mediastinal shift and significant compression of the diaphragm was identified. Drainage of 3300mL of blood was confirmed after tube thoracostomy, always without hemodynamic instability. After this initial period the haematic drainage stopped, there was a relief of the complaints, no change in haemoglobin level was detected and he had a good imagiological evolution. It was decided not to have an emergent surgery and to do the surgery electively. The patient was operated 3 days later for cloth removal. A zone of adherences and an organized blood clot were identified in the apical region. No other obvious cause for the bleeding could be identified. The post-operatory period had no complications and the patient was discharged seven days later. Spontaneous pneumothorax is a pathology that many times can be treated only with medical vigilance but might also lead to life threatening consequences.

Keywords: *Spontaneous pneumothorax. Hypertensive hemopneumothorax. VATS.*

CYSTIC ADENOID CARCINOMA OF THE TRACHEA - A MULTIDISCIPLINARY SURGICAL APPROACH

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Primitive tumors of the trachea are extremely rare but amongst them squamous cell carcinoma and cystic adenoid carcinoma are the most frequent. Cystic adenoid carcinoma is a tumor typically originary from the salivary glands and cervical region, but also found in other anatomical regions, and usually has an indolent growth, but it's often locally invasive and may metastize. We describe the case of a 54-year old woman that had wheezing complaints in the previous five years and more recently progressively worsening dyspnea She went to the emergency room with an episode of acute dyspneia and the otorhinolaryngology team diagnosed a mass immediately under the vocal cords that almost occluded the tracheal lumen. The bronchoscopy showed a large tumor, inserted in the posterior wall of the trachea, one centimeter under the vocal cords. An emergency rigid bronchoscopy was then performed, the tumor was biopsied and a Dumon® stent placed to assure airway permeability. The histologi cal analysis identified a cystic adenoid carcinoma. A multidisciplinary surgical team with thoracic surgeons and an otorhinolaryngologist underwent a segmentar resection of 6 centimeters of trachea, followed by the anastomosis of the trachea to the cricoid cartilage. The post-operatory period was uneventful and the patient was discharged from the hospital on the 14th day, after a control bronchoscopy. The bronchoscopy and CT 6 months later showed no signs of stenosis of the trachea. And the patient was symptom free. Nevertheless, that CT showed two micronodules in the left lower lobe (LLL). Follow up with Chest CT and volumetric evaluation did not show any signs of growth in the following year. In the CT scan 18 months after surgery there was a slight growth of one of the lesions and a left video assisted thoracoscopy was performed, with wedge resection of the LLL involving the lesions. The histological analysis confirmed that the lesions were in fact metastasis of cystic adenoid carcinoma. The patient is now asymptomatic and without signs of recurrence of disease.

Keywords: *Primary tracheal tumors. Cystic adenoid carcinoma. Tracheal surgery.*

DISSEMINATED ACTINOMYCOSIS IN A PATIENT WITH TUBERCULOSIS

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Actinomyces are normal constituents of the oral flora, fastidious therefore difficult to obtain in culture. Immunosuppression, malnutrition and oral infections are risk factors. Male patient, 25 years, black, non-smoker was admitted due to fever with maximum temperature of 39ºC, weight loss of about 8 Kg and cervical adenopathy with 9 months of evolution. He was submitted to cervical adenopathy biopsy which revealed alcohol-acid resistant bacilli. Mycobacterium tuberculosis sensitive to Isoniazid and Rifampicin was identified in genotipic test. He started Isoniazid, Rifampicin, Pyrazinamide and Ethambutol with good compliance. Six months after starting therapy he noted a swelling on the right anterior chest wall. A chest CT-scan was performed to characterize this lesion, and revealed multiple thoracic and abdominal infectious loci, corresponding to "cold abscesses" and a large abscess in the right pectoral delimited by the anterior costal pleural lining, mediastinal, hilar and abdominal adenopathies with necrotic center. No parenchymal lesions were observed. To clarify this situation he was admitted to our unit. He denied any previous diseases. On examination he had good general condition, a well-defined mass, with lower adherence on the right anterior chest wall. No changes on laboratory findings. X-Ray revealed a right perihilar oval hypotransparency. A new chest CT-scan was performed and showed small areas of peribronchic consolidation on the right, with central cavitation, multiple thoracic and abdominal lymphadenopathies and an abscess at the base of the neck on the right and over the costal grid. To obtain a diagnostic was performed a bronchoscopy that revealed a diminished luminal area at the intermediate bronchus due to a prolapse of an adenopathy. A transbronchic needle aspiration was performed on this site, and histological results were granulomas and Actinomyces. No agent was obtained in culture. The chest wall mass was drained with complete resolution. All microbiological tests were negative. Since

the patient had completed 6 months of therapy for tuberculosis, and with these results, we decided to start therapy with I.V. Penicillin. Patient was asymptomatic, and chest CT-scan 4 weeks after treatment revealed a decrease of some lesions. Since the patient had a good response, he was discharged and had to continue therapy with oral Amoxicillin for 6 months.

***Keywords**: Actinomycosis. Tuberculosis. Thoracic lesion.*

RISK OF TUBERCULOSIS IN RHEUMATIC PATIENTS TREATED WITH IMMUNOSUPPRESSANTS

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Background: Immunosuppressants, including new biological therapies, enhance the risk of developing active tuberculosis (TB). An effective screening program and eventual treatment for latent TB is essential.
Aim: To investigate the impact of a TB screening program on the development of active TB in rheumatic patients treated with immunosuppressants, including biological therapies.
Material and methods: Retrospective analysis of patients screened for latent or active TB in the Pulmonology Diagnostic Center, since February 2005. The evaluation included clinical history, tuberculin skin test, chest x-ray, interferon gamma release assay and chest CT according to protocol. If any of these was positive, 9 months isoniazid (INH) and pyridoxine was prescribed. If all were negative, patients remained under biannual monitoring. All cases of latent and active TB and side effects of therapy were recorded.
Results: The study had 837 patients with 588 females and mean age 52±14 years. The average length of follow-up was 2.8±1.4 years. Main rheumatological diagnoses were rheumatoid arthritis (n=451), psoriatic arthritis (n=148) and ankylosing spondylitis (n=130). 187 were candidates to biological therapy and 650 were proposed or already treated with classic immunosuppressants. 704 patients were prescribed INH, of whom 18 refused and 16 suspended for intolerance (9 hepatotoxicity, 2 neurotoxicity, 1 medullar aplasia and 1 toxidermia). One patient was treated with quadruple antituberculosis drugs owing to suspected ocular tuberculosis. The remaining 132 were proposed only for clinical monitoring. One patient developed pleural TB after 3 years of INH chemoprophylaxis.
Conclusion: In this population, 84.1% required prophylaxis with INH and only 2.3% developed intolerance. The TB screening program used was effective, with a unique reported case of active TB, in others words, 0.15% of all patients who received chemoprophylaxis.

***Keywords**: Tuberculosis. Screening. Immunosuppressant.*

EFFECTS OF IMMUNOSUPPRESSANT THERAPY IN THE DETERMINATION OF INTERFERON GAMMA RELEASE ASSAY AND TUBERCULIN TEST IN RHEUMATOLOGIC PATIENTS

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Background: The tuberculin test (TT) and *Interferon Gamma Release Assay* (IGRA) are used in the screening of latent tuberculosis. One of the high risk groups include rheumatic disease patients, proposed or under immunossuppressants. Since these tests depend on the subjects immune cell-mediated response, questions remain about these tests reliability in patients under immunosuppressant therapy.
Aim: To determine if TT and IGRA are influenced by immunosuppressants.
Materials and methods: Retrospective study of patients followed at the Portuguese Institute of Rheumatology, subjected to screening for latent tuberculosis at Pulmonology Diagnostic Center, between 2004 and 2011. The effect of immunossuppressants was assessed by a univariate logistics analysis.
Results: 642 patients (471 females) with an average age of 52.2±13.4 were assessed. Main rheumatological diagnoses were rheumatoid arthritis (n=390), ankylosing spondylitis (n=104) and psoriatic arthritis (n=70). The most used immunosuppressants were corticosteroids (n=447), methotrexate (n=453), salazopirine (n=203) and hydroxychloroquine (n=102); 48 patients were under therapy with biological agent. A PT was performed on all patients, with 322 positive cases. The proportion of negative PT was higher with azatioprin, hydroxycloroquine and etanercept, however only hydroxycloroquine had a significant statistic association ($P<.05$). IGRA was performed on 304 patients of which, 20 were positive. Apart from the proportion of negative IGRA had been higher with azatioprine, cyclosporine and etanercept, these relationships did not present statistical significance.
Conclusion: In this study the IGRA does not seem to have been influenced by the immunosuppressant therapy. PT, on the other hand, presented a larger variation of results with numerous immunosuppressant agents, even if just significant for hydroxychloroquine.

***Keywords**: Tuberculosis. IGRA. Tuberculin test. Immunosuppressants.*

TUBERCULOUS PERICARDITIS-CAN THE IDENTIFICATION OF *MYCOBACTERIUM TUBERCULOSIS* BE AVOID?

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Introduction: Tuberculous pericarditis is a rare disease and difficult to diagnose. The diagnosis is based on the demonstration of *Mycobacterium tuberculosis* in the pericardial fluid or in pericardial biopsy samples. However, the profitability of bacteriological examinations is low and a definitive diagnosis is not always possible.
Case report: The authors present the case of a 73-year old patient with a history of diabetes mellitus non-insulin-treated, congestive heart failure, atrial fibrillation and stroke, transferred from the Infectious Diseases Department where he had been previously admitted for Febrile Syndrome of indeterminate cause, non-responsive to empirical antibiotic therapy. A chest CT scan showed a pericardial effusion, pleural effusion and a nodular lesion on the lower left lobe suggesting lung cancer. Pericardiocentesis had already been undertaken with drainage of hematic fluid; biochemical analysis showed it to be an exudate. Thoracentesis showed a citrus pleural fluid with biochemical characteristics of a transudate. Due to recurrence of pericardial effusion in the absence of an etiological diagnosis, a surgical thoracoscopic pleuro-pericardial window was performed. Pleural and pericardial samples were obtained and histologic diagnosis was compatible with acute pleuritis and chronic pericarditis. Intra-dermal tuberculin reaction was negative and further study showed no connective tissue disease or confirmation of primary or secondary neoplasia. The patient maintained fever and bacteriological cultures were negative but IGRA (*Interferon-Gamma Release Assays*) test was positive which lead to an antituberculous therapy trial. On the 10th day of therapy the patient became apyretic with progressive clinical and radiological improvement. He was discharged from

hospital with the diagnosis of probable tuberculous pericarditis and followed up in the Pneumology Diagnostic Center in Coimbra, where he was found clinically stable.
Conclusion: The high mortality rate associated with untreated tuberculous pericarditis and the low profitability of diagnostic methods which allow a definitive diagnosis to be made often mean that it is necessary to initiate an antituberculous therapeutic trial. The latter may, indeed, be a diagnostic criterion in countries with a high prevalence of tuberculosis.

Keywords: *Tuberculous pericarditis. IGRA. Antituberculosis therapy trial.*

TUBERCULOSIS-A CLINICAL CASE

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Introduction: Tuberculosis is an infectious disease caused by the Bacillus of Koch, and the contagion done person to person. Ten years ago the World Health Organization declared Tuberculosis as a global emergency, having since then been implemented prevention and control measures of the disease. In the European Union, Portugal is one of the countries with the highest incidence of reported cases and with greater expression of the aspects that give it the character of emerging infection. At present the incidence of this pathology is declining, with an annual average decrease of 6.4%, being the country classified as incidence intermediate. The family doctor is the first contact with the population and thus occupies a privileged place for the early detection.
Description of event: Male, Caucasian, 48 years, unmarried, unemployed, born and resident in Ponta Delgada, living with a sister and 6 nephews. Personal History: Cognitive deficit and COPD. Chronic Medication: Brisomax® 500 inhaler. In 20/01/12 appointement he complained of cough and sputum with aggravation in the previous weeks. He was not taking his medication. Denies dyspnea, rhinorrhea, fatigue, fever, pain, anorexia, weight loss, night sweats. The objective examination: just to emphasize overall decrease of vesicular murmur (vm). Chest radiography (x-ray), analytical tests were asked and enhanced importance of compliance with the therapy. He was instructed to bring the inhalers in the following appointment. In 21 /03/12 appointment there was a worsening of the cough and he complained of right chest pain. The objective examination: Afebrile, hemithorax pressure not triggering pain and overall decrease in vm. Analytically slight leukocytosis with neutrophilia. Chest x-ray showed fibrotic lesions that make the suspicions of ancient or recent tuberculosis, requiring clarification by computed tomography (CT). Completed request for CT chest and BK research in expectoration, verified technique of use of inhaler and reinforcement of the importance of compliance with the same. In 14/05/12 appointment there was a persistence of symptoms. Physical examination was like the previous appointment. The CT showed lesions with cavitation in favor of tuberculosis. BK research negative. The Department of Pulmonology of the Hospital do Divino Espírito Santo de Ponta Delgada, EPE was contacted. The patient repeated the BK research which was positive. He started therapeutics and was send to Centro de Tratamento de Doenças Respiratórias de Ponta Delgada (STDR).
Conclusion: The Family Doctor has a holistic vision and it's the population's first contact with Health Care. That's why Family Doctor has a important role in diagnosis, identifying cohabitants and other contacts that might be in danger, working in partnership with Public Health on prevention. An early diagnosis and treatment of tuberculosis avoids not only the complication of the disease itself but also its spread.

Keywords: *Cough. Chest pain. Tuberculosis.*

MILIARY TUBERCULOSIS: AN UNCOMMON CAUSE OF SEPSIS

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Introduction: Miliary tuberculosis constitutes a potentially lethal form of the disease resulting from massive lymphohaematogeneous dissemination of *Mycobacterium tuberculosis*. The HIV|AIDS pandemic and the widespread use of immunosuppressive drugs changed the epidemiology of miliary tuberculosis. However, severe tuberculosis sepsis is a rare presentation in non-immunocompromised individuals.
Case report: A 51-year-old Caucasian male with no prior pathologic history presented to the Emergency Department with complaints of insidious dyspnoea, non-productive cough, night sweating and pleuritic chest pain with two months of evolution. He also described asthenia, malaise, anorexia and weight loss (>10% of body mass), denying fever. On admission, the patient was emaciated, pale and sweaty; febrile (39.0ºC), tachycardic (104 bpm), hypotensive (89|63 mmHg), tachypneic (28 cpm) and with SpO2 96% (FiO2 24%). Cervical and supraclavicular adenomegalies were identified. Chest radiograph showed a reticulonodular pattern. Chest CT scan revealed a micronodular (miliary) pattern with mediastinal and axillary adenophaties. The patient was promptly started, empirically, on antituberculous therapy. The identification of *Mycobacterium tuberculosis* was made, by direct analysis and nucleic acid amplification, in sputum and urine samples. The complementary study excluded any kind of primary or acquired immunodeficiency. The patient was discharged asymptomatic and without respiratory failure after eight weeks of treatment, being oriented for the Pneumologic Diagnostic Center.
Discussion and conclusion: Tuberculosis sepsis is a rare complication of disseminated disease, being mostly reported in severely immunosuppressed individuals and extremely uncommon in non-immunocompromised individuals, demanding a high grade of clinical suspicion for a proper and early therapeutic approach.

Keywords: *Sepsis. Miliary tuberculosis.*

HYDROPNEUMOTHORAX: CATASTROPHIC PRESENTATION OF A TUBERCULOSIS PRIMARY INFECTION

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Introduction: Primary tuberculosis refers to the development of disease following an initial exposure to Mycobacterium tuberculosis, being mainly a disease of childhood. Many studies have shown a growing frequency of tuberculosis primary infection in adolescents and adults.
Case report: The authors present a clinical case of an 18 year-old Caucasian female, with no prior pathologic history or evidence of trauma, referred to the Emergency Department with a right hydropneumothorax in a chest X-ray. The patient described a clinical scenario, with one month of evolution, of asthenia, anorexia and weight loss, accompanied by productive cough and night sweating. 24 hours prior to the admission, she developed, suddenly, right pleuritic pain associated with fever. On admission, the patient was pale and emaciated; febrile (38.0 ºC), tachycardic (150 bpm), normotensive (120|70 mmHg), tachypneic (30 cpm) and with SpO2 91% (FiO_2 24%). A diagnostic thoracentesis was made, which demonstrated an empyema with ADA levels of 101.8 U/L (N<40 U/L). A thoracic tube was inserted. After the identification, in a sputum sample, of Mycobacterium tuberculosis, antituberculous therapy was initiated. The complementary study excluded any kind of primary or acquired immunodeficiency. The patient was discharge asymptomatic and without respiratory failure, after six weeks of treatment, being oriented to the Pneumologic Diagnostic Center.

Discussion and conclusion: Tuberculosis remains an important cause of pulmonary infection. Large volume hydropneumothorax constitutes a severe and rare presentation of pulmonary involvement for Mycobacterium tuberculosis, demanding its diagnosis a high grade of clinical suspicion.

Keywords: *Tuberculosis. Primary infection.*

VENOUS THROMBOEMBOLIC EVENTS IN TUBERCULOSIS

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Introduction: Respiratory infections are known to increase the risk of venous thromboembolic events (VTE). These complications are considered rare in TB.
Objectives: To assess the frequency of VTE in patients hospitalized for TB, its demographic and clinical characteristics, therapeutic and mortality.
Material and methods: adult patients hospitalized for TB in a tertiary hospital without HIV infection between 2007 and 2011 were evaluated. VTE were diagnosed by chest angio-TC (pulmonary embolism-PE) or ecoDoppler (deep vein thrombosis-DVT) and prophylactic therapy of VTE, prior coagulation studies and VTE treatment was evaluated.
Results: of the 208 TB patients (male: 76%, mean age 54 years) 15 patients (7.2%) were diagnosed with VTE (males: 67%, mean age: 56 years, pulmonary TB: 12 cases, disseminated TB:2, lymph node TB:1), including 6 PE, 7 limb DVT, 2 cervical vessels DVT and 1 PE with jugular VT. VTE occurred less than 1 month after initiation of antibacillary agents, except in 1 patient (3 months). Most patients (73%) didn't receive thromboprophylaxis; 73% were treated with low molecular weight heparin. Two patients died (13%), with the remaining being discharged, most (85%) medicated with anticoagulants.
Conclusions: The frequency of VTE in this study was higher than in previous studies with hospitalized patients for TB (0.6 to 3%). The authors conclude that VTE is an important complication in this population, especially in the initial phase of antituberculosis treatment, justifying the use of thromboprophylaxis in patients with known risk factors for VTE.

Keywords: *Tuberculosis. Venous thromboembolic events.*

GENDER DIFFERENCES IN TUBERCULOSIS

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Background: Although it is not clear understood, tuberculosis (TB) notification is twice as high in men as in women. Among the reasons pointed to this sex inequality are socioeconomic and cultural factors as well biological mechanisms. Therefore, investigating the difference between genders may lead us to understand better the causes of lower TB notification rates among women. Our study aimed to identify the clinical, socioeconomic and behavioural differences between female and male patients with active TB.
Methods: During June and July 2012, adult patients (>18 years) with active TB under treatment in CDP-VNG were prospectively enrolled. Data was obtained from a patient questionnaire and missing information was collected from medical records.
Results: Among a total of 54 patients included, 37 (68.5%) patients were male and 17 (31.5%) were female, with an overall male female ratio of 2.18:1. Almost 1/3 of the patients aged 45-55 years. The mean ages of male and female patients were 49.1±12.1 and 47.1±18.3 years, respectively. There was a female predominance in the age groups lower than 34 and higher than 65. No differences were found in the educational level or employment status of the two subgroups. However, women had lower income compare to man. Similar social habits were present in male and female groups, but the average number of contacts screened was higher in women. Concerning comorbidities and habits, women showed less HIV infection (P=.026) and lower levels of smoke and alcohol abuse (P<.001 and P=.012, respectively). Fewer women than men presented cough and sputum production (P=.003 and P=.001). The median diagnosis delay was 25 days longer in women, although without statistical significance (W 78.5 vs M. 53.5 days).
Conclusion: The peaks ages for the respective sex vary between the different countries. In your study female predominance was observed at the extremes of age (<34 and >65 years). Among the differences between sex, men with active TB show higher levels HIV infection, smoking and alcohol abuse. These aspects are also seen in the general population. As describe in literature, fewer women reported symptoms of cough and sputum production. This difference may complicate the diagnosis of tuberculosis in women and explain a delay in diagnosis higher among women compare to men.

Keywords: *Tuberculosis. Gender. Differences.*

TUBERCULOSIS IN A PATIENT UNDER IMMUNOSSUPRESSIVE TREATMENT

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Patient 19 years old, female, Caucasian, student, with persistent fever for 15 days. The patient, with rheumatoid arthritis diagnosed at 16 years of age, had previously been observed in CDP Santarém to exclude latent tuberculosis infection, when biological treatment has been proposed. With the diagnosis of pulmonary tuberculosis, presenting diffuse infiltrates in lower lobes, began treatment with four drugs (HRZE), improving. Two weeks later she had recurrence of fever and worse general status. The diagnosis of TB was confirmed with isolation of *M. tuberculosis,* without resistances. With the maintenance of the same drugs, associated with oral corticotherapy, she had progressive improvement, clinical and radiological. As said, the patient had been previously (1-2 years before) screened for TB. She as a posterior contact with a patient with bacillary pulmonary TB, residing in her neighbourhood. Of that patient our service had screened 11 contacts, even the mother of this patient. The authors present this case, discussing the difficulties of the treatment of TB in the immunossupressed patient, difficulties in the screen of latent TB and insufficiencies in contact tracing.

Keywords: *Tuberculosis. Latent tuberculosis infection. Rheumatoid arthritis. Contact tracing.*

TUBERCULOSIS AND DIABETES: EXPERIENCE OF 10 YEARS IN CENTRO DE DIAGNÓSTICO PNEUMOLÓGICO DE SANTARÉM

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Introduction: Diabetes mellitus (DM) is a condition that increases the risk of tuberculosis (TB). The convergence between the two diseases may be responsible for increased morbidity and mortality.

Objective: to delineate the demographic, clinical and evolutive profile of TB associated with DM in a chest clinic, Centro de Diagnóstico Pneumológico (CDP) of Santarém.
Material and methods: a retrospective study, in which 56 were reviewed clinical processes of patients with TB and DM, of a universe of 671 cases of TB in Santarém CDP between 2002 and 2011, comparing the two groups.
Results: the mean age was 61 years. There was a higher incidence in males (76.2%); the age groups most affected were the ≥75 and 45-54 (25% and 21.4% respectively). In 7.1% of cases found himself associated with addiction, at 5.36% HIV infection and 3.6% to IRC in hemodialysis. Alcoholism was the main risk group found with 17.9%. The location of pulmonary TB was prevalent with 37(66.1%); in these cases there were 3 concomitant extrapulmonary location. The extra-pulmonary forms accounted for 22 cases (39.3%): 9 pleural, 5 ganglion, 2 pericardial, 2 spinal, 1 meningitis, 1 genito-urinary, 1 disseminated, 1 cutaneous. In pulmonary forms radiographically 51.4% showed cavitary lesions. As the result of treatment was 84% success rate, with 8 deaths occurred, of which 50% of the age group ≥75. One patient, of 2011, (1.7%) is still under treatment.

Keywords: *Tuberculosis. Diabetes.*

PULMONARY TUBERCULOSIS AND DIABETES: RADIOLOGICAL ASPECTS

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Introduction: There are different radiological presentations of pulmonary tuberculosis (TB) and the atypical presentations are frequently associated to other comorbidities such as diabetes and HIV/AIDS.
Objective: Describe the radiological aspects of pulmonary TB in patients with diabetes mellitus (DM) followed in a chest clinic, Centro de Diagnóstico Pneumológico (CDP) de Santarém.
Material and methods: Were reviewed 38 cases of patients with DM in a universe of 423 cases of pulmonary TB followed at CDP Santarém in a 10 years period, 2002 to 2011.
Results: The mean age of the patients was 60 years. Males were 30 (78.9%) and females 18 (21%). The right lung was more affected, in 15 cases (39.5%), and there were bilateral lesions in 14 cases (38.6%). In 23 cases (60.5%) were present cavitary lesions. In many cases we found multilobar lesions: in 76.3% in the upper third, in 65.8% in the middle and in 47.4% in the inferior. Using ATS classification of extension, there were 18 cases (47.4%) of moderate extended, 11 (28.9%) of very extended and 9 (23.7%) minimal. The evolution after treatment showed 16 cases with minimal scars and 7 with extensive scars and 7 with no apparent lesions. The main group risk was alcoholism, 113.2%. The simultaneous pathologies were hematological disease 4 cases (10.5%), neoplasm 3 (7.9%) and chronic renal failure 3 (7.9%)

Keywords: *Pulmonary tuberculosis. Diabetes mellitus. Radiological aspects.*

MILIARY TUBERCULOSIS: A CASE REPORT

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We describe a case of a 54 year old, male patient, who was unemployed. He was an alcoholic and a former smoker. He was admitted to our hospital with dyspnea, productive cough with purulent sputum and weight loss with 2 months of evolution. He also referred a subcutaneous nodule with a soft consistency on the right lumbar region. Physical examination highlighted severe cachexia. Arterial blood gas test showed hypoxemia (PaO_2 66.8). The PA chest radiograph showed a bilateral heterogeneous opacity, more pronounced in the upper half of the right lung. The sputum cultures were positive for M. tuberculosis complex. An ultrasound of the right lumbar region was performed and showed a superficial collection with 13.6x6.9 cm, non-pure, in relation to the palpable swelling. The abdominal-pelvic CT scan showed multiple abscesses in perihepatic location, in subcostal location involving the right abdominal wall and in right psoas and right-vertebral muscle. A lumbosacral CT scan was also performed and showed extensive retrovertebral and right lumbar paravertebral abscesses, without active spondylodiscitis. The blood tests (including HIV 1+2) were negative. A percutaneous drainage of the subcutaneous and right psoas abscesses was performed, with aspiration of purulent liquid. The cultures were positive for M. tuberculosis complex. There was clinical improvement after the introduction of antimycobacterial therapy.

Keywords: *Abscess. Tuberculosis. Disseminated.*

ACTIVE TB CASES ON COIMBRA'S PENITENTIARY INMATE POPULATION, OVER 2000-2012

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Background: The implementation of the TB control programs in prison environment has been slow and heterogeneous. The prison population presents a higher risk of infectious diseases, majorated by prison's lifestyle, high turnover, and the inevitable prioritizing of security over health.
Methods: Retrospective study regarding Coimbra Penitentiary inmates diagnosed/treated for active TB by Coimbra's "CDP" over the last 12 years.
Results: We found 11 cases of TB. Mean age was 38.2 years, with 9.1%being immigrants. About 63.6% and 27.3% were CHV e HIV positive. The average CD4 count was 175.000/mm^3. Two thirds of them were under ARVT. Alcoholism, smoking and drug-abuse were present in 36.4%, 90.9% e 63.6%, respectively. About 18.2% suffered from neurosis and 9.1%from psychosis. Previous pulmonary disease was present in 9.1% and 36.4% have had prior TB. 72.7% presented with pulmonary TB, while ganglionar, pleural and peritoneal TB had minor expressions. There was 75% smear-positive pulmonary TB. There was only one case of multirresistence. Average treatment length was 9.9 months. Culture negativization time was achieved at 4 months. DOT was assured in 54.5%. Among the rest, 80% presented good compliance. Treatment was completed with cure in 81.8% of cases. Mortality rate was 9.1%.
Conclusions: Compared to the indicators for the general population, the group of inmates showed inferior age, higher HCV/HIV infection rates, and higher prevalence of drug-abuse and psychiatric comorbidities. Average treatment length was longer, so as the culture negativization time. The majority of cases resulted from passive detection. DOT is a valious instrument to achieve good prisional performance indicators.

Keywords: *Tuberculosis. Prison system.*

TUBERCULOSIS AND ONCOLY-WHEN THE FRONTIER VANISHES

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The Tuberculosis epidemiology have shown a global variation by the appearance of new immunosupression state conditions, defined by HIV epidemics, as well as by the use of newly effective and potent chemotherapeutic agents to oncologic diseases. Disseminated

tuberculosis, acquired by primary or latent infection, raises new challenges in the treatment of oncologic patients. The actual clinical case is about disseminated tuberculosis in a patient with previous stomach cancer diagnosis (T3N3M0) and treatment-surgery plus adjuvant chemotherapy (ended 6 months before). He has been admitted with a constitutional syndrome, intermittent fever and thoracic pain within the last 2 months, without other symptoms/deficits. Disseminated tuberculosis was diagnosed: imagiologic milliary pattern (with positive direct test in bronchial lavage), urinary tuberculosis (with positive direct test in urine), bony tuberculosis (spondylodiscitis and paravertebral abscesses), central nervous system tuberculosis (multiple nodular lesions in both hemispheres and in the cerebellum). The contemporaneity of this issue as well as the exuberant imagiologic manifestations in a patient with subtle symptoms reinforce the importance of this clinical case and raise some questions: should the latent tuberculosis screening be generalized and mandatory in the initial evaluation of an oncologic patient and pre-chemotherapy treatment, even without known exposure? Should we search for another organs involvement in an asymptomatic patient with disseminated tuberculosis and 2 known affected organs?

Keywords: *Tuberculosis. Oncology. Immunosuppression.*

PULMONARY ACTINOMYCOSIS: CASE REPORT

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Introduction: Pulmonary actinomycosis is a rare condition caused by *actinomyces* infection. The form of presentation ranges from subacute to chronic mimicking, sometimes, a chronic fungal infection, tuberculosis or even malignancy. The infection has an excellent prognosis if recognized early and with proper treatment.
Case report: A 63 year-old woman was admitted in July 2011 with complaints of anorexia, weight loss and a productive cough. Additionally she reported episodic right sided thoracic discomfort for the last 5 month. A chest CT with endovenous contrast was performed in the emergency room witch highlighted middle lobe opacity with extension to the hilar, paravertebral e paramediastinic regions, with necrotic areas, suggestive of neoformation. Fiberoptic bronchoscopy revealed partial obstruction of intermediate bronchus by infiltration. Culture of the bacteria in bronchial aspirate and histological exam of transbronchial biopsies were negative. Transthoracic needle aspiration allowed the identification of *actinomyces* in the sample. The patient started a 6-month treatment with amoxcillin plus clavulanic acid with radiological and clinical monitorization.
Conclusion: Pulmonary actinomycosis is a rare and difficult to diagnose condition with excellent prognosis if recognized early and with proper treatment.

Keywords: *Actinomyces. Pulmonary actinomycosis.*

HYDATIDOSIS-RARE PULMONARY INVOLVEMENT

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Introduction: Pleural effusion may be associated with a variety of pathologies and to determine the etiology is essential for the treatment. As a consequence, it is important to consider the rarer causes of pleural effusion, as sometimes there can be difficulties in the differential diagnosis.
Development: The authors describe the case of a 75-year-old man with complaints of dry cough, dyspnea on exertion, paroxysmal nocturnal dyspnea and edema of the lower limbs with about one month of evolution. The diagnostic procedures revealed elevated infection parameters and a diffuse heterogeneous opacity in the right lower lobe, consistent with pleural effusion. The patient had a past history of chronic alcohol abuse. The patient was subjected to diagnostic thoracentesis, allowing drainage of pleural fluid which was confirmed to be exudative. The microbiological study of pleural fluid was negative, showing high cellularity and the presence of high level of leukocytes with 79% of neutrophils. Bronchoscopy was also performed and revealed a friable mucosa with inflammatory signs. CT scan confirmed the presence of organized pleural effusion, with partial collapse of the left lower lobe, and in the abdominal extent of the exam, a bulky hypodense formation with calcified wall was observed on the right lobe of the liver. Due to the liver abnormalities detected, abdominal ultrasound was performed, demonstrating the presence of a complex formation of probable parasitic origin. Sera Western Blot analysis showed anti-hydatic antibodies, confirming the diagnosis of hydatidosis with pulmonary involvement. The patient initiated treatment with albendazole and subsequently was proposed to surgical excision of the hydatid cyst.
Conclusion: Hydatidosis is a parasitic disease caused by various species of helminths of the genus *Echinococcus*. Portugal is among the countries considered by the WHO as endemic, with an estimated national incidence of 2.2/100 000 inhabitants. Hydatid cysts are usually localized in the hepatic parenchyma (50 to 70%) and in the lung (5 to 30%), and the infection may be silent or develop various complications, being the most frequent the cystic rupture. In the case described, the pleural effusion is a result of the rupture of a hepatic hydatid cyst into the pleural cavity, showing good clinical improvement with the pharmacological treatment. In a country where this disease is endemic, we should consider hydatidosis in the approach of the patient presenting with pleural effusion.

Keywords: *Pleural effusion. Hydatidosis.*

CHARACTERIZATION OF THE PNEUMONIAS OF A PULMONOLOGY WARD

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Introduction: Pneumonia is still a relevant entity of a pulmonologist daily practice. In 2005 a new category of pneumonia was introduced by the ATS/IDSA guidelines: health care associated pneumonia (HCAP). Therefore, it is demanding to better characterize and validate this entity.
Aims: Characterize all pneumonias admitted to a pulmonology ward concerning epidemiologic factors, etiology, severity, length of hospital stay, comorbidities, mortality, prognosis and empiric therapy.
Methods: The authors analyzed retrospectively all patients with pneumonia admitted to the pulmonology ward of a district hospital in a year period. All data were treated using SPSS 18.0.
Results: The authors analyzed a total of 221 patients: 89 met the criteria of HCAP and 132 of community acquired pneumonia-CAP. The mean age was 69 years old (HCAP 75.6±15.3 and CAP 64.5±19.1). The mean length of hospital stay was 13.45±8.8 days (HCAP 14.58 and CAP 12.69) and the mortality rate was 17.2% (HCAP 11.8% and CAP 5.4%). A total of 44.8% of patients (HCAP 29.9% and CAP 14.9%) had poor functional status. The mean number of comorbidities per patient was 2.5, highlighting cardiovascular disease (41.2% of patients), Diabetes Mellitus (25.8%), obstructive lung diseases (23.1%), cerebrovascular disease (21.7%) and structural lung diseases (11.3%). Concerning the pneumonia severity the authors found that 163 patients presented with PSI ≥IV (73.8%) (HCAP 85.4% and CAP 66%) and 125 patients with CURB65 ≥2 (56.6%)(HCAP 67.4% and CAP 49.3%). In 74.7% of patients the empiric antibiotic therapy was according to the CAP guidelines. The

etiologic agents more frequently isolated were enterobacteriaceae (6.8% of all patients and 38.5% of all microbiological cultures) and, among these, *E. coli* followed by MRSA and *Acinetobacter baumannii*. *Streptococcus pneumoniae* was in 4th place (2.3% of all patients). All of these microorganisms, except pneumococcus were more frequently isolated in HCAP patients.
Conclusion: Pneumonia is still an entity with significant mortality and well represented within a pulmonology ward. It is noteworthy the significant number of comorbidities per patient, which can justify the admission of patients with non-severe disease. Despite the relevant prevalence of HCAP and its concordant etiology, ¾ of patients were treated empirically according to CAP guidelines, which may express some resistance by the physicians to apply the HCAP guidelines. There is still need for validation of the HCAP concept and better characterization of this entity.

Keywords: *Community acquired pneumonia. Health care associated pneumonia.*

GOOD'S SYNDROME - A CAUSE OF REPEATED RESPIRATORY INFECTIONS

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Introduction: Good's syndrome (thymoma associated with hypogammaglobulinemia) is a rare entity (less than 60 case reports in the literature) with unknown physiopathology and high mortality rate (30% and 60% at 5 and 10 years, respectively). This disease is characterized by hypogammaglobulinemia, low B cell count in the peripheral blood and low T CD4 cell count. Therefore, there are repeated infections especially in the respiratory and gastrointestinal tracts (chronic diarrhea in 50% of patients), caused by encapsulated bacteria and opportunistic agents. The first line treatment is intravenous immunoglobulin G.
Case report: The authors describe a case report of a 46 year-old male patient, non-smoker, with personal history of thymoma (surgery 12 years before), pulmonary tuberculosis 11 years before and repeated respiratory tract infections since 8 years ago, who presents with acute respiratory disease. The etiologic investigation included: 1) Thoracic CT-bilateral central and peripheral bronchiectasis with probable ongoing infection; 2) Bronchoscopy-generalized inflammatory sings, negative bacteriology and negative mycobacteriology; 3) Pulmonary function tests-severe obstructive ventilatory defect, with no response to bronchodilators and resting hypoxemia (65mmHg); and 4) Blood work-lymphopenia, low CD4/CD8 ratio, hypogammaglubulinemia (2%), IgG 213 mg/L (RV: 723-1685) and IgA 8 mg/L (RV: 60-382). The diagnosis of Good's syndrome was made based on the clinical information and the results above described and the patient was started on immunoglobulin G every 3 weeks. After starting the treatment the patient maintained some respiratory tract infections. He died 7 years after the diagnosis after an episode of hypercapnic respiratory failure with need of mechanical ventilation.
Conclusion: The Good's syndrome prognosis is bad so the diagnosis should be achieved as briefly as possible in order to start the treatment so the infectious complications, frequently associated to bronchiectasis, can be avoided. The clinical acumen and cunning should be combined with the clinician diagnostic suspicion when confronted with the typical clinical and immunologic profile of the Good's syndrome. This disease should always be taken into account concerning the differential diagnosis of the adult hypogammaglobulinemia. More studies are needed to understand this entity.

Keywords: *Repeated respiratory infections. Hypogammaglobulinemia. Thymoma.*

RARE ASSOCIATION BETWEEN ACTINOMYCOSIS AND PULMONARY MALT LYMPHOMA

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Hospital Professor Doutor Fernando Fonseca.

Actinomycosis is a rare infeccious disease caused by Gram-positive, facultative anaerobic filamentous bacteria. It may affect the lung in less than 15% of cases and it presents with unspecific respiratory and radiologic features, thus creating great diagnostic difficulties. MALT lymphoma is a haematological malignancy with rare pulmonary involvement, corresponding to less than 1% on non-Hodgkin lymphomas in general. The authors describe a case of an 83 year-old caucasian female, with a history of hypertension, diabetes, cardiac failure and chronic bronchitis, presenting in the outpatient clinic with persistent productive cough and hypoxemia. She has been previously treated with several antibiotics without improvement. Radiological examination revealed bilateral infiltrates and a CT scan confirmed numerous nodular bilateral lesions, the bigger one with 26 mm. Bronchoscopy revealed diffuse inflammatory signs, and bronchoalveolar lavage was unremarkable, with negative microbiological cultures. Due to worsening of purulent sputum she repeated sputum examination, and cytological examination revealed organisms compatible with Actinomyces. She was admitted as an inpatient and treated with benzatinic penicillin, with clinical improvement. However, in the following months there was worsening of nodular lesions and a transthoracic lung biopsy was ordered. Histological exam revealed type B lymphoma CD20+, CD35, CD23, CD5, CD10, bcl2, bcl6 and ciclidine negative. The study was completed with body CT and bone marrow biopsy and cytogenetic studies, confirming the diagnosis of non-Hodgkin type B pulmonary MALT lymphoma. She underwent immunochemotherapy (R-CVP in 8 cycles) and maintenance therapy with Rituximab for two years and had frank clinical improvement and reduction of number and size of the pulmonary nodules. Three years after diagnosis, the patient remains stable and is still followed in Pulmonology and Oncology. There are some cases describing the association between Actinomycosis and pulmonary malignancies, and only one case in a patient with a previously diagnosed non-Hodgkin lymphoma, already treated. Both entities carry problems with differential diagnosis and Actinomycosis can mimic malignant lesions. In this case there was no culture confirmation for Actinomyces, but the lack of image response to correct therapy led to the successful diagnosis of the second disease.

Keywords: *Actinomycosis. MALT lymphoma. Lung.*

LUNG ABSCESS AND NECROTIZING PNEUMONIA: A HOSPITAL'S EXPERIENCE

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Introduction: A lung abscess is defined as the necrosis and cavitation of pulmonary parenchyma secondary to a microbial infection. Some authors utilize the term necrotizing pneumonia to distinguish pulmonary necrosis with multiple small abscesses from a larger cavitary lesion, although these 2 terms represent a continuum of the same process.
Objectives: To analyze the clinical, epidemiological, radiologic and microbiologic characteristics of a lung abscess or necrotizing pneumonia in a given hospital, as well as the treatment and prognosis of these patients.
Methods: A retrospective study of medical records of all patients hospitalized between January 2006 and June 2012, with the diagnosis of either lung abscess or necrotizing pneumonia, was conducted.

Results: A total of 51 patients were included (42 men/9 women), averaging 60 years of age. The diagnosis of lung abscess was established in 46 patients, while 5 patients were diagnosed with necrotizing pneumonia. The patients most frequently complained of fever (84%), cough (69%), which was productive in 51% of cases, constitutional symptoms (47%), dyspnea (43%) and chest pain (31%). Comorbidities/risk factors were identified in 84% of patients, the most frequent being arterial hypertension (28%), smoking (26%), alcoholism (19%) and pulmonary neoplasm (12%). The principal motives for hospitalization of these patients consisted of lung abscess (49%), pneumonia (33%), and chest trauma (10%). All patients did a chest x-ray, and in 55% of cases an image suggestive of a lung abscess was visualized; 90% of these patients had a thoracic CT scan performed. The lesions were predominantly localized in the lower right lobe (35%), upper right lobe (26%) and lower left lobe (22%). The cultures were positive in 45% of cases and the principal agents isolated were *Klebsiella pneumoniae* (12%), *Pseudomonas aeruginosa* (8%), *Streptococcus pneumoniae* (6%) and *Staphylococcus aureus* (5.9%). All patients underwent antibiotic therapy, initially empirical and later adjusted according to the antibiotic sensitivity test, with a 30 day average of treatment (from admission to outpatient). Only 10% of patients underwent surgical treatment. During hospitalization, 9 patients (18%) died, with an age average of 76.7 years, which was significantly superior to that of those who survived (56.3 years).
Discussion: A lung abscess is a pathology which manifests itself with nonspecific respiratory complaints and that affects primarily men with comorbidities, as was the case in this series. The main diagnostic tests performed were chest x-ray and thoracic CT scan. The right lung was the preferential location of the lesions, which corresponds to that described in literature. Worth noting is the fact that in the majority of cases, a cavitated lesion was already visible by chest X-ray. In this patients group, the number of cultures which presented microbiological growth was superior to that published in previously studies and bronchial secretions were the main site of isolation. The mortality rate increases directly with patient age (P=.005) as well as with the presence of risk factors/comorbidities, although in the latter case the relationship is not statistically significant.

***Keywords**: Lung abscess. Necrotizing pneumonia.*

HUMAN PAPILLOMA VIRUS IN THE BRONCHIAL TREE

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Background: Solitary endobronchial papillomas (SEP) are rare, noninvasive tumors considered until now to have low malignant potential. However, Human papilloma virus (HPV) infection, strongly linked to the pathogenesis of SEP, has been associated with dysplasia and a high risk of squamous cell carcinoma, hence the need to carefully diagnose and treat these lesions.
Case report: A 52-year old single male, heavy smoker and drinker, was referred to a follow-up appointment two months after being admitted for pneumonia at our pulmonology ward. He had recent complaints of productive cough of bloody sputum. Physical examination was unremarkable and routine blood sample analysis was normal. A videobronchoscopy revealed a cauliflower lesion of the upper lobe right bronchus posterior segment. CT scan showed features compatible with fibrotic lesions and thickened bronchial walls and PET/CT reported mild hypercaptation in the right hilar region. Biopsies disclosed a bronchial papilloma virus lesion.
Conclusions: Data on solitary bronchial papilloma are sparse and published mainly as case reports. Until further studies arise, the authors contribute with this rare case study to enhance the understanding of the disease.

***Keywords**: Human papilloma virus (HPV). Solitary endobronchial papilloma. Tumor.*

AN UNUSUAL CAUSE OF PLEURAL EFFUSION

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Introduction: Pleural effusions are often unilateral exudates and the most common causes are infections, malignancies or pulmonary thromboembolism. Pleural effusion and lymphadenopathy are rare findings in pulmonary infection by *Mycobacterium xenopi,* and the typical radiographic appearance is an apical cavitation. In certain areas of Canada, the United Kingdom and Europe, M. xenopi is second to *Mycobacterium avium* complex (MAC) as a cause of lung infection caused by non-tuberculous mycobacteria (NTM). In this regard the authors present the following case report: Male patient, 81 years, ex-smoker (60 pack-years), retired (former railway machinist, having worked as a stoker machinist for over 10 years), with previous moderate alcohol consumption and a diagnosis of adenoma with high-grade dysplasia in the ascending colon in 2007. In April 2012, he went to the CHVNGaia/Espinho Emergency Department (ED) motivated by a progressively worsening dyspnea with moderate exertion for two months. Two months before coming to the ED, the patient had cough with mucopurulent sputum and general symptoms (fever, asthenia, anorexia and 2-3 kg weight loss) which improved with the antibiotics prescribed by his general practitioner. The digital radiography allowed the diagnosis of a large pleural effusion. It was performed diagnostic and therapeutic thoracentesis. Macroscopically, it was a yellowish, not turbid fluid. Analytically, it had biochemical characteristics compatible with an exudate, glucose 43, adenosine deaminase 60, polymorphonuclear/mononuclear-7.8/92.2%; negative cytology for malignant cells. The histological evaluation of pleural biopsies was consistent with nonspecific chronic pleuritis. The chest computed tomography scan performed after thoracentesis showed atelectasis of the lingula and of the left lower lobe with a small pleural effusion, 4 nodes with 3 to 5 mm diameter distributed by the superior, middle and inferior right lobes and a calcified granuloma with 8 mm in middle lobe. He underwent bronchoscopy and a *M. xenopi* species was identified in the bronchial lavage culture. The patient started treatment with isoniazid, rifampicin, ethambutol and clarithromycin with clinical and radiological improvement. Currently there is no consensus concerning the optimal treatment regimen, neither concerning its duration. Nevertheless, the ATS/IDSA 2007 statement on diagnosis, treatment and prevention of non-tuberculous mycobacterial diseases supports the chosen treatment regimen and states that it should be maintained for 12 months after negativation of mycobacterial culture of sputum.
Conclusion: Pleural effusions caused by infection with *M. xenopi* are rare in immunocompetent patients and isolation of these microorganisms often corresponds to contamination or colonization rather than true infection. In the reported case all criteria defined in ATS/IDSA statement of 2007 were present.

***Keywords**: Mycobacterium xenopi. Pleural effusion.*

CHRONIC NECROTIZING ASPERGILLOSIS - CLINICAL CASE

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Chronic necrotizing aspergillosis or semi-invasive, is an indolent, destructive process of the lung due to invasion by *Aspergillus* species; different from aspergilloma and invasive aspergillosis. Male patient, 42 years, black, born in Togo, independent for everyday

life activities, construction worker, admitted due to progressive worsening of fatigue with 6 months of evolution, dyspnea, cough and mucopurulent sputum with some weeks of evolution. On the admission day he referred moderate haemoptysis, non-measured fever, weight loss and dyspnea at rest. He denied any previous illness, including tuberculosis; non-smoker, with moderate alcohol consumption. On examination he was under-weight, polipneic, with peripheral oxygen saturation <88%; laboratory findings CRP-5.5mg/dL, LDH-383 U/L, hypoxemia with a PaO_2-60 mmHg, HIV negative, no mycobacterias were identified in the sputum. X-Rays revealed multiple cavities. Thorax CT-scan showed bronchiectasis and bilateral cavitations, the largest, on the left, with 8cm and, on the right with 4.2cm, some with dense contents; many segments of the lung were collapsed, mediastinal lymphadenopathy. To exclude tuberculosis he performed additional investigation with the following results: no agent was isolated from sputum; bronchoscopy with subsidiary techniques revealed *Aspergillus niger,* positive Galactomannan, serum IgG antibodies to *A. niger* positive (0.88). We admitted the hypothesis of chronic necrotizing aspergillosis, so the patient started therapy with voriconazole. There was significant clinical improvement, with resolution of haemoptysis, dyspnea and hypoxemia with no need to oxygen therapy. Imagiologic findings showed no regression compared with the initial study. He was discharged after 50 days of therapy, with dyspnea on moderate exertion, referred to our rehabilitation unit. He kept antifungal therapy with itraconazole.

Keywords: *Aspergillosis. Lung cavity.*

ENDOBRONCHIAL ACTINOMYCOSIS: A RARE INFECTION

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Introduction: Actinomycosis is a chronic infection caused by anaerobic gram-positive bacteria which colonize the oropharynx and the gastrointestinal tract. In the majority of cases, these bacteria are located in the cervicofacial (50%) and abdominopelvic (20%) regions, rarely presenting in the endobronchial region, with merely a few documented cases of such occurrence. Endobronchial actinomycosis is characterized by bronchial obstruction secondary to irregular granular bronchial thickening or colonization of a foreign body, which may appear in the bronchofibroscopic exam as a whitish exophytic mass. It does not present with specific clinical or radiologic characteristics, signs of infection are often absent and the cultures are frequently negative, making it difficult to establish an early diagnosis. The recommended treatment consists of antibiotic therapy for a 6-12 month period and the prognosis is generally favourable.
Case report: 67-year-old male, previous smoker (45 pack-years) more than 17 years ago, with a history of severe obstructive sleep apnea syndrome, mild chronic obstructive pulmonary disease, hypertension, type 2 diabetes mellitus, atherosclerotic disease involving the 3 main territories (acute myocardial infarction, stroke with resulting left brachial hemiparesis, chronic lower limb ischemia) and an abdominal aortic aneurysm (5.2 cm). Referred to Pulmonology due to a chronic cough with hemoptoic expectoration and wheezing for the last 2 years. Physical examination revealed wheezing localized to the left lung field. Analytically, there was no elevation of inflammatory parameters, screening with Phadiatop inhalant was negative and IgE levels were normal. There were no specific findings on the patient's chest x-ray. The chest CT scan revealed a nodular endobronchial mass measuring 5mm, located in the left upper lobe bronchus (LULL), with peripheral ring enhancement after injection of contrast agent. The patient underwent bronchofibroscopic exam, during which a whitish lesion was visualized obstructing the entry into the LULL and lingula, which was biopsied. *Haemophilus influenzae* was isolated in the bronchial aspirate and the culture for Acid-Alcohol Resistant Bacilli (A.A.R.B.) was negative. Histopathologic examination revealed an intense lymphoplasmacytic infiltrate in the lamina propria, along with bacterial colonies and actinomyces, and the diagnosis of Endobronchial Actinomycosis was established. The patient began antibiotic therapy with amoxicillin and clavulanic acid, and is currently in his fifth month of treatment. He maintains follow up in Pulmonology.
Discussion: The authors present the case of an endobronchial infection with a rare etiology, in a patient with risk factors for microaspiration of oropharyngeal contents. As described in literature, emphasis is placed on the difficulty in identifying the agent, isolated only histologically, and in establishing a differential diagnosis with other pathologies which cause bronchial obstruction, like lung neoplasm or the presence of a foreign body.

Keywords: *Actinomycosis. Endobronchial mass.*

NECROTIZING PULMONARY ASPERGILLOSIS: A PECULIAR CASE

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Necrotizing pulmonary or semi-invasive aspergillosis (NPA) is a little know pathology and difficult to diagnose; corresponds to an indolent infectious process, with consequent destruction of lung parenchyma, caused by inhalation of *Aspergillus*, usually *Aspergillus fumigatus*. The authors present a clinical case which for its clinical severity and coexistence of prior severe lung disease without known immunosuppression, illustrates the particularities of this pathology. This is a 42 year old woman, non-smoker, with a medical history of Pulmonary Tuberculosis in 2001, treated for nine months, COPD GOLD C (FEV_1 0.96L-27%) with bilateral bullous emphysema (two determinations of α_1 antitrypsin within the normality), status post left thoracotomy with surgical recession of bubbles in the left upper lobe and pleurodesis after a spontaneous pneumothorax hypertensive in 2005 during the puerperium and depressive syndrome. Clinically stable until March 2008, date when she begins recurrent small-volume hemoptysis and exertional dyspnea. Chest Computed Tomography (CT) scan showed multiple bullae of emphysema bilaterally, several areas of bilateral cylindrical and traction bronchiectasis, heterogeneous condensation focus of the right lower lobe with air bronchogram and a cavitation area with dense contents without air-fluid level. Bronchoscopy showed signs of bleeding from the right upper lobe bronchus. In bronchial secretions was isolated *Aspergillus fumigatus* and cytology showed an acute inflammatory exudate. Direct and culture mycobacteriological examination was negative. Our patient began therapy with voriconazole (200mg twice a day) which lasted 3 months, with clinical and radiological improvement. Control chest CT scan showed known structural pathology and suggestive image of aspergilloma in the right upper lobe. She had recurrent respiratory infections with the isolation of multiple bacterial without new radiographic changes. Mycobacteriological exams successively negative. Until March 2011, our patient showed clinical and radiological stability, date when she begins having severe hemoptysis, with hemodynamic consequences, and was documented an extensive multilobar pneumonia with severe respiratory failure and elevation of inflammatory markers. It was diagnosed recurrence of aspergillosis, with new growth of *Aspergillus* on respiratory samples. She restarted therapy with voriconazole, which lasted 6 months, with imagiological resolution of pneumonia and blood gas normalization, persisting suggestive image of aspergilloma. The diagnosis of NPA is a real clinical challenge. This diagnosis requires a histological demonstration of tissue invasion by *Aspergillus* and its growth on culture, but are

defined in the literature, clinical, radiological and laboratory findings that when are present in patients with predisposing factors allow consideration of this disease. In this context it is crucial having a high diagnostic suspicion to ensure the rapid and accurate identification of this pathology, vital for reducing its morbidity and mortality.

Keywords: *Necrotizing pulmonary aspergillosis (NPA). Aspergillus fumigatus. Aspergilloma.*

THE LUNG IN PRIMARY IMMUNODEFICIENCIES — APROPOS OF A CLINICAL CASE

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Primary immunodeficiency diseases (PIDs) are inherited disorders of the immune system, resulting in increased susceptibility to unusual infections and predisposition to autoimmunity and malignancies. Usually manifested in the postnatal period and during childhood, although some cases are detected clinically in adulthood. The symptoms are associated with the degree of immunodeficiency and immune system affected. The authors describe a case of a 36 years-old male patient, smoker, with mild mental delay, short stature (5^{th} percentile), facial dysmorphia and total alopecia since the 3 years of age, of unexplained etiology. He presented a history of recurrent respiratory infections since childhood and a hospital stay, four years before, for pneumococcal pneumonia with pleural reaction, treated uneventfully. Once again hospitalized with pneumonia complicated with multilolated empyema and *Streptococcus pneumoniae*(SP) bacteraemia. A chest tube was placed and broad spectrum antibiotherapy was instituted. Chest computed tomography scan, in addition to pleural effusion empyemic, showed multiple cylindrical and cystic bronchiectasis, thick-walled, covering all lobes.HIV negative. Due to suspicion of PIDs, an immunological study was performed, demonstrating severe panhipoglobulinemia: immunoglobulin (Ig) G 19.6 (normal: 680-1450), IgM<17 (34-214), IgA<25.5 (83-407), with severe G2 and G4 deficit and normal complement. IgG intravenous therapy was initiated, with slow but favorable evolution. Having regard to the phenotype associated with mental retardation and primary immunodeficiency, a genetic study has been requested to characterize the syndrome. Early diagnosis of IPDs allows adequate treatment, with unequivocal benefit in patients mortality and life quality, making diagnoses investigation mandatory in the face of suspect. In IPDs with predominantly humoral deficit, pyogenic infections, due to SP for example, are common, and the institution of replacement therapy with intravenous IgG allows reducing infections frequency and severity, preventing complications and sequelae. The European Society for Immunodeficiencies established warning signs which must raise the threshold for suspicion of IPD in adulthood. Because respiratory system infections are very common, pulmonology emerges as a specialty of choice in the diagnosis of this entity.

Keywords: *Primary immunodeficiency. Pneumococcal pneumonia.*

ATYPICAL PRESENTATION OF CASE OF H1N1

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Female, 21 years old, manicure, born in Minas Gerais, living in Portugal for 2 years, healthy, with no pneumological history, non-smoker and no other risk factors. Appealed to the ER (SU) for dyspnea and chest pain radiating to the right anterior shoulder homolateral. Two days earlier, would have been treated with anti-inflammatory drugs and muscle relaxants without relief. Said the viral symptoms with slight 1 week before that yielded to symptomatic therapy, without medical resource hospital. The day I used the SU found himself prostrate, with no signs of difficulty breathing, eupneic and peripheral oxygen saturation 97% on room air. Hypotension and tachycardia (AT: 77/34 mmHg and HR 145 beats per minute) with decreased breath sounds in the right lung auscultation, percussion and palpation in presenting symptomatology of stroke/pneumonia. The remaining physical examination was negative. Performed imaging studies (chest radiography and chest CT) confirmed that images suggestive of pneumonia with condensation and air bronchogram of the right lower lobe atelectasis passive underlying lung parenchyma, extensive empyema right and right anterior pneumothorax. Performed chest drain output with purulent fluid and isolation of Streptococcus pyogenes bacterial strain Lancefield group A, metilcilina sensitive. The research of legionella and pneumococcal antigens in urine was negative. The cultures of the bacillus of Koch, Legionella and HIV from blood and pleural fluid were negative. The result of the urine was negative. There was isolated MRSA. It was identified in the exudate pharyngeal swab for H1N1 virus by RT-PCR. By presenting septic shock and severe hypoxemia with respiratory acidosis was admitted to the Intensive Care Unit Pneumológicos (PICU). Doctored up with Oseltamivir, Amoxicillin/Clavulanate and Azithromycin respiratory physiotherapy with supplement. There has been a slow but progressive respiratory and hemodynamic recovery, without the need for mechanical ventilation. However, it was necessary to resort to pleural decortication (VATS) for drainage and cleaning of empyema given their persistence and flocculation with cisural component and slight passive collapse of the right lower lobe. After 27 days of hospitalization, was discharged asymptomatic and hemodynamically stable eupneic going to be monitored in consultation specialty. CONCLUSIONS: The interest of this case is that a healthy patient without previous pathology and immunocompetent but probably colonized with *Streptocos pyogenes* infection with influenza virus A, H1N1, predispose it to a fibrinous pneumonia with empyema. The *S. pyogenes* is an agent less common pulmonology infection with severe septic shock, infecting only 2 to 3% of adults. This mode of presentation is not common with this bacterial strain (where the predominant pharyngo-tonsillitis and skin infections) being the rarity and severity of this type of agent in a patient without predisposing factors, as it detects an infection concomitantly The flu virus the reason that justified the disclosure.

Keywords: *Pneumonia and H1N1.*

RESPIRATORY DISEASES IN INFANCY, CHILDHOOD STRESS AND PARENTAL STRESS

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This research, centered in infant respiratory diseases, aimes at studying the relationship between parental stress and marital satisfaction with: a) parental self and sociodemographic variables and (b) the child 24 hours urinary cortisol as a very strong indicator of stress. This is a longitudinal study, which the dimensions in analysis were mesured in two different moments: Time 1 (summer) and Time 2 (winter) and compared between them. The participants (ages between 6 months and 5 years old) and their parents are distributed in two groups, taking into account the number of respiratory episodes of the child being high or low. The respiratory symptoms

of the child were identified by the information obtained in the ENVIRH study (financed by FCT and were this research is anchored, which aims to study the air quality in day-care kindergarden and their relationship with the health and well-being of the children that attend them). In Time 1 we collected 475 questionnaires with information related to parental stress, marital satisfaction and parental sociodemographic variables and 102 urine samples of the children aged 4-5 (to determined the 24hours urinary cortisol, a very strong indicator of stress) and this information was associated with the information gathered in the 475 ENVIRH questionnaires of the same children, more precisely with the information regarding the respiratory diseases of these children. By doing this research we intend to verify if children that have a higher number of respiratory infections also have a higher level of urinary cortisol and, on the other hand, also have parents with a higher level of parental stress and a lower marital satisfaction.

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Keywords: *Infant respiratory diseases. Parental stress. Urinary cortisol.*