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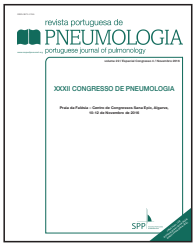

POSTERS PRCC

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PRCC-001. *ROS1* MUTATION HAS MALE PREDOMINANCE IN PULMONARY ADENOCARCINOMAS

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Introduction: *ROS* proto-oncogene 1 (ROS1) rearrangements in pulmonary adenocarcinomas confer sensitivity to *ROS* kinase inhibitors, including crizotinib. The presence of *ROS1* rearrangements are typically identified by FISH. FISH patterns of *ROS1* gene and pulmonary adenocarcinoma subtyping were evaluated in this work.
Methods: A total of 94 biopsies of pulmonary adenocarcinoma (59 men and 35 women), collected between January 2015 and December 2015, were tested for *ROS1* rearrangements by FISH with ZytoLight® SPEC *ROS1* Dual Color Break Apart Probe. FISH-positive cases were defined as over 15% split signals in tumor cells. Associations between FISH results and clinicopathological features were assessed.
Results: *ROS1* rearrangements were identified in four (4.3%) of the 94 biopsies with patients varying between 40 and 84 years old. Compared with *ROS1*-negative patients, the positive rate of *ROS1* in male 3 (75%) patients tumours was significantly higher than in female patients - 1 (25%) case.
Conclusions: *ROS1* gene rearrangement defines a rare subset of pulmonary adenocarcinoma. In 94 wild-type EGFR/ALK patients biopsies screened in our Labs, 4.3% showed *to be ROS1*-positive adenocarcinomas. In our positive cases no specific association was found with adenocarcinoma histological subtyping. Nevertheless, this genetic translocation appeared to be associated with male gender.

Key words: *ROS1 gene. FISH. Male gender. Pulmonary adenocarcinoma.*

PRCC-002. *ALK* GENE REARRANGEMENT CORRELATES WITH MUCINOUS DIFFERENTIATION IN FEMALE PULMONARY ADENOCARCINOMA

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Introduction: There has been efforts to correlate the histomorphology with molecular alterations in lung adenocarcinomas. The detection of specific mutations for targeted treatment and preview of prognosis. *ALK* gene mutation defines a tumor group of pulmonary adenocarcinomas responsive to targeted crizotinib anti-*ALK* treatment. The FISH patterns of *ALK* gene and histomorphology were evaluated in this work.
Methods: Histological classification/WHO 2015 was applied to 166 biopsies of bronchial-pulmonary carcinomas - 94 men and 72 women with ages varying between 28 and 77. *ALK* rearrangements were screened by FISH, with Zyto*Light*® SPEC ALK/EML4 TriCheck™ Probe. The mutational analyses of EGFR (exons 18-21) were performed by sanger sequencing techniques. Associations between molecular results, histological and clinicopathological features were assessed.
Results: *ALK* rearrangements were identified in 12 cases (7%): 8 (67%) women and 4 (33%) men. Ages below 50 represent 40% of these cases. This genetic translocation appeared to be associated with younger age, and female gender. Mucinous adenocarcinoma was the predominant histological type - 33%, followed by solid adenocarcinoma (12%); cases of micropapillary adenocarcinomas, adenosquamous and squamous carcinoma exhibited also *ALK* mutations. In this study, there was not coexistence of *ALK* rearrangements and *EGFR* mutations.
Conclusions: The clinical and pathological features of the *ALK*-positive cases in this series of patients were similar to those previously reported in literature. *ALK* rearrangements were significantly increased in solid pattern with predominance of mucin production subtype and in mucinous signet ring cells and

micropapillary pattern. Histologically, extra/intracytoplasmic mucin, cribriform, tubulopapillary, and solid signet-ring cell patterns were the most well-known indicators of *ALK* rearrangement that also persists in squamous and adenosquamous carcinomas.

Key words: *ALK gene. FISH. Histomorfology. Female gender. Pulmonary adenocarcinoma.*

PRCC-003. CELL COUNT IN INDUCED SPUTUM SAMPLES: PROCESSING PROTOCOL OPTIMIZATION

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Introduction: The analysis of induced sputum samples is a method used for phenotyping asthma patients. There are different protocols for processing and the analysis of these samples, with variable efficacy. We used the European Respiratory Guidelines Society[2] as the basis for starting the protocol optimization. The aim of this study is to optimize a protocol for processing of induced sputum samples that allows the total cellular and differential count.
Methods: For protocol optimization we processed seven samples of induced sputum, using the protocol-type for the preparation of these samples with the following steps: I. Macroscopic evaluation of the sample; II. Adding mucolytic; III. Filtering with nylon mesh: IV. Cell count; V. Cytocentrifugation forwards; VI. sample coloring with May-Grünwald Giemsa (MGG); VII. cell differential count. Changes have been introduced in several stages of this protocol: in step III (evaluation of the samples with and without filter) and VI (introduction of the Papanicolaou staining). We perform a quantitative and qualitative evaluation of the samples obtained.
Results: We found that the use of the sample filter with nylon mesh led to cell loss (total cellular loss in two samples tested) and that the omission of this step did not increased cell aggregation by the presence of mucus. We also checked by observation and cell count that lysis of the mucus was effective, at the end of stage II, not requiring to do step III. On qualitative evaluation of stained samples (step IV) with Papanicolaou and MGG we observed that samples stained with Papanicolaou had less background, cell overlap, and less obfuscation by bacteria and best cell preservation.
Conclusions: The optimized protocol in our laboratory allows the processing of sputum samples and their preparation under appropriate conditions to perform a total cellular count, to carry out effectively mucolysis (and reduces costs inherent to the use of filters). Papanicolaou staining improves cell preservation for excellent differential count.

Key words: *Induced sputum. Total cell count. Differential cell count.*

PRCC-004. THE BROAD SPECTRUM OF EOSINOPHILIC LUNG DISEASES

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Introduction: Eosinophilic lung diseases are a heterogeneous group of disorders with multiple etiologies. They evolve with varying degrees of eosinophilic infiltration of the interstitium and alveolar spaces and are often associated to eosinophilia (80% of cases). Three cases are described, two of which are representative of secondary eosinophilic pneumonias and one of unknown cause.
Case reports: 1) 38-year-old woman, smoker, with gastroesophageal reflux (GERD) and depression treated with duloxetine, presented with dry cough, dyspnea and digital clubbing with 4 months of evolution. Chest-CT: extensive areas with ground-glass opacification and honeycombing. She had restrictive pattern (FVC = 59.3% and DLCOsb = 22.7%) and eosinophilic alveolitis (51.2%). Eosinophilic pneumonia secondary to duloxetine was assumed, and the drug was suspended and prednisolone 1 mg/kg/day initiated. Clinical worsening was verified in the following four months, with stool ova and parasites exam positive for Ascaris lumbricoides, confirming the diagnosis of Löeflller syndrome. She was treated with albendazole, but had clinical, radiological and functional deterioration. An open lung biopsy demonstrated Usual Interstitial Pneumonia. The patient is currently without specific treatment, but uses oxygen therapy at ambulation and has been accepted for lung transplantation. 2) 65-year-old woman, non-smoker, with hypertension, depression, deep vein thrombosis, GERD surgery, referred by dry cough, fatigue and dyspnea for 9 months. Usual medications: bupropion, sertraline, alprazolam, irbesartan/hydrochlorothiazide, atorvastatin, rivaroxaban. Two admissions for bronchospasm and severe respiratory failure, slightly improved with bronchodilators, inhaled and systemic corticosteroids. At the follow-up appointment, prednisolone dose was reduced and gradually tapered off. Two months later was readmitted with similar complaints. From the investigation: Chest-CT: diffuse opacities; bronchoalveolar lavage (BAL): eosinophilic alveolitis (30%); negative stool ova and parasites exam. Diagnosis of eosinophilic pneumonia secondary to sertraline was made, and this drug was discontinued, resulting in clinical and radiological improvement. 3) 55-year-old man, non-smoker, with dyslipidemia, GERD and rhinitis, with a 2 years history of dry cough, shortness of breath and wheezing. Regularly medicated with montelukast, bronchodilators, prednisolone 20 mg/day and pravastatin. The methacholine challenge test was positive, serum tests showed peripheral eosinophilia (11.2%) and increased total IgE (475 IU/ml). At the appointment, an inhaled corticosteroid was initiated and prednisolone was suspended. Few weeks later was readmitted with severe worsening and respiratory failure. Chest-CT: diffuse bronchial wall thickening and centrilobular nodules; blood analysis: peripheral eosinophilia (28%), increased total IgE and sedimentation rate; BAL: eosinophilia (85.2%). Further investigation ruled out secondary causes. Treatment with systemic corticosteroids end up in rapid clinical and radiological improvement, associated with normalization of eosinophil counts in peripheral blood. A chronic eosinophilic pneumonia diagnosis was assumed, associated with late-onset asthma.
Discussion: In eosinophilic lung diseases, clinical suspicion is essential, highlighting the importance of the BAL in the diagnosis. The etiological investigation should be conducted in order to distinguish secondary from idiopathic forms.

Key words: *Eosinophilia. Eosinophilic pneumonia. Broncho-alveolar lavage.*

PRCC-005. PULMONARY TOXICITY TO BLEOMYCIN. CLINICAL CASE

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Introduction: The therapeutic scheme adriamycin-bleomycin-vinblastine-dacarbazine (ABVD) is considered the standard in the treatment of Hodgkin's lymphoma. Bleomycin is a recognized agent associated with the pulmonar toxicity - fibrosis, hypersensitivity pneumonitis or knuckles - whose incidence increases with age, with

the existence of structural chronic pulmonary disease, with use of high doses or concomitant scheme with radiotherapy, etc.
Case report: Male gender patient, 64 years old, smoker with smoking charge of 40 pack units/year, with syndrome antecedents of obstructive sleep apnea in noninvasive ventilation. Hodgkin lymphoma diagnosed in November 2015 on chemotherapy with ABVD since January 2016. In March 2016 initiates clinical condition characterized by easy fatigability and dyspnea rapidly progressive effort accompanied by severe partial respiratory failure with 1 month of evolution. The imagiologic evaluation identified a reticular pattern of diffuse distribution with comb areas - pulmonary fibrosis. Initiated systemic corticosteroid therapy, long-term oxygentherapy and changing ventilation mode with slow and progressive symptomatic improvement, subsequently restarting a new chemotherapy regimen without bleomycin.
Discussion: Bleomycin can cause pulmonary fibrosis in 8-10% of the cases, with no functional or clinical criteria to predicting the pulmonary toxicity of the drug. Clinical provides most often a subacute onset characterized by dyspnea, dry cough and thoracic pain and is common presented as reticular infiltrates or bilateral micronodular evolving in more serious cases to honeycomb. Treatment with corticosteroid therapy and suspension of the drug presents a response to a treatment around 50-70%, may however, be a more severe evolution as fibrosis, respiratory failure and, in rare cases, lead to death.

Key words: *Bleomycin. Toxicity. Lung. Fibrosis.*

PRCC-006. A RARE CAUSE OF FATIGUE

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Introduction: The neuromuscular diseases (NMD) includes a great range of diseases with different prognosis. The congenital myopathies represent 10% of all NMD, mostly diagnosed in childhood due to neurodevelopmental disorders. However some variants may be detected only in adulthood.
Case report: We describe a 62-years-old Caucasian male, nonsmoker, with previous history of asthma and osteoporosis. His mother had been hospitalized for a long period, with invasive mechanical ventilation and subsequently underwent tracheostomy and she had been discharged with long-term oxygen therapy, but without an accurate diagnosis. He complains of fatigue with several years of evolution and orthopnea since about a month ago, without previous hospital admissions. On physical examination, the patient was eupneic, acyanotic, without thoracic deformities. His neurological evaluation was normal. He had previously performed an echocardiographic exam which revealed no alterations and pulmonary thromboembolism was excluded by angio-CT scan of the chest. The pulmonary function tests showed a severe restrictive pattern and decreased maximal inspiratory and expiratory pressures. He was referred to neurology consultation and underwent muscle biopsy and genetic study. The results were compatible with multi-minicore disease (MmD). Currently, 5 years after the diagnosis, he is clinically stable, under nocturnal noninvasive ventilation and maintains autonomy in daily living activities.
Discussion: MmD is a rare NMD, which was first described in the 1970s. It usually presents with hypotonia and proximal weakness in childhood, with a few cases described in adulthood. Muscle biopsy reveals multifocal, well-circumscribed areas with reduction of oxidative staining along the longitudinal axis of the muscle fiber ("minicore"). MmD is related to mutations in both the selenoprotein N (SEPN1) and the skeletal muscle ryanodine receptor (RYR1) gene. Classic phenotype is characterized by spinal rigidity, early scoliosis and respiratory impairment. The weakness involves mainly the axial muscles. The respiratory impairment is the main prognostic factor. It is often grossly out of proportion to the overall degree of weakness and to the mobility impairment, therefore it is crucial to look for nocturnal hypoventilation. In this case, the authors intend to draw attention to nonspecific complains such as fatigue and to the possibility of late presentation of a NMD.

Key words: *Neuromuscular disease. Myopathy. Minicore.*

PRCC-007. ALPHA-1 ANTITRYPSIN DEFICIENCY – CHARACTERIZATION OF A CENTRAL HOSPITAL

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Introduction: Alpha-1 antitrypsin deficiency (AAT) is one of the most common inherited disease, associated with an increased risk of pulmonary and extrapulmonary disease being, however, largely under-diagnosed.
Objectives: Evaluation and characterization of individuals with AAT deficiency in a central hospital.
Methods: Clinical records from patients with AAT, whose phenotype/genotype was determined between January 2006 and April 2016 at Centro Hospitalar São João, were retrospectively reviewed and analyzed.
Results: Of the 268 patients analyzed, 67.2% were men, mean age 38.9 years (min 0, max 79 years old). At the time of diagnosis 22.4% were under 18. The most frequent genotype was MZ (31.3%), followed by the SZ (19.8%), MS (19%), ZZ (13.8%) and SS (7.8%). Different types of rare alleles were detected: PI*Mmalton (n = 10), PI*Plowell (n = 2), PI*I (n = 4), PI*Mwürzburg (n = 1), PI*Mheerlen (n = 2), PI*T (n = 2) and PI*Q0G (n = 1). Mean AAT plasma levels was 65.8 mg/dL considering all patients, 28 mg/dL among ZZ, 63.2 mg/dL among SZ, 79 mg/dL among MZ, 65.8 mg/dL among SS and 104.9 mg/dL among MS. Respiratory symptoms were the reason for the diagnosis in 51.9% and liver disease in 24.6%, followed by family screening (13.1%). The respiratory function tests, at diagnosis, revealed obstructive ventilatory disease in 27.4%, 59.4% of them with severe/very severe obstruction. Between the 144 of the patients who had thoracic computed tomography, the most frequent radiological findings were emphysema (47.9%) and bronchiectasis (39.6%). 38% were smokers or former smokers. Liver disease was found in 34.1% of all patients and in 73% of the patients with ZZ genotype. Eleven patients started augmentation therapy, 10 of which with ZZ genotype and one with ZMheerlen. Four patients underwent lung transplantation (3 ZZ genotype and one SZ) and one had liver transplantation (SZ genotype).
Conclusions: In this sample, the majority of patients were referred for respiratory symptoms, with predominance of the MZ genotype. Several rare and null alleles were identified. At the time of diagnosis, a significant proportion of patients had abnormal lung function and emphysema or bronchiectasis. Liver disease proved to be very prevalent, specially within the ZZ genotype group. These findings support the need for guidelines and a national registry in order to improve the rate of diagnosis and provide earlier intervention in the natural history of the disease.

Key words: *Alpha-1 antitrypsin deficiency. Phenotypes. Genotypes.*

PRCC-008. ALPHA-1 ANTITRYPSIN DEFICIENCY: A POTENTIALLY FATAL DISEASE

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Introduction: Alpha-1 antitrypsin deficiency (AAT) is a very common codominant autosomal inherited disease, associated with an increased risk of pulmonary and extrapulmonary disease being, however, largely under-diagnosed. It is the most common fatal genetic disease in adulthood.
Objectives: To evaluate and characterize mortality among individuals with AAT deficiency in a central hospital.
Methods: Data from adult patients, carrying a Z or rare/null allele, whose phenotype/genotype was determined between January 2006 and April 2016 at Centro Hospitalar São João, were consulted and retrospectively analyzed. The number, causes of deaths, demographic and clinical characteristics of dead patients were analyzed.
Results: Of the 143 patients analyzed, most were men (59.7%), with age at diagnosis between 18 and 79 years (mean 48.3 years). Until July 2016 19 patients died (13.2%), mostly men (63.2%), with age of death between 45 and 84 years (mean 59.9 years). 42.1% of them were ZZ, 36.8% MZ, 10.5% SZ and 10.5% Mmalton. Mean AAT plasma levels was 62.7mg/dL considering all patients and 57mg/dL among dead patients. The most frequent causes of death were hepatic events (43.8%), followed by respiratory events (31.3%) and others (24%). Considering the ZZ group 83.3% died from respiratory causes. On MZ group 57.1% died from hepatic causes and 42.9% from others causes. Liver disease was found in 78.9% of patients. Obstructive ventilatory disease was detected in 42.1%, 77.8% of which with severe/very severe obstruction. 42.1% of dead patients were smokers or former smokers. Thoracic computed tomography was performed in 14 of the patients who died and 57.1% presented emphysema at time of diagnosis. Tree dead patients, with Z phenotype, were under augmentation therapy.
Conclusions: In this sample, 13.2% patients died, 43.8% attributed to hepatic causes. Among ZZ group more than 80% died from respiratory causes while in MZ group the most frequent cause of death were hepatic events. These findings support that severe AAT deficiency is a disease with high mortality at middle age, not only by respiratory causes, and reinforce the need for early diagnosis and multidisciplinary intervention.

Key words: *Alpha-1 antitrypsin deficiency. Mortality.*

PRCC-009. NEW ERA IN PRIMARY CILIARY DYSKINESIA DIAGNOSIS

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Introduction: Primary ciliary dyskinesia (PCD) is a rare genetic disorder characterized by motile cilia dysfunction and/or defective ciliary ultrastructure. Patients present with recurrent and chronic respiratory infections, recurrent otitis/otorrhea and sinus disease, but also non-pulmonary manifestations such as *situs inversus* and infertility. The diagnostic process is complex. According to recent national and international guidelines, diagnosis can be established if there is a clinical phenotype consistent with PCD and changes in at least two of the following tests: (1) nasal nitric oxide (nNO) levels, (2) transmission electron microscopy (TEM), (3) high speed videomicroscopy analysis (HVMA) and (4) biallellic disease-causing mutations identified by gene testing.
Objectives: Characterize the clinical presentation and diagnostic pathway of a group of paediatric patients with PCD.
Methods: Descriptive analysis of patients with PCD diagnosis made in 2015-2016, followed in a Paediatric Pulmonology outpatient clinic from a university affiliated hospital. Retrospective chart review with collection of demographic and clinical data, as well as the results from diagnostic tests. nNO was measured with a portable electrochemical analyser NIOX MINO® Nasal (Aerocrine AB, Solna, Sweden). For each patient two nasal brushing samples were obtained for analysis by the two methods: HVMA [Nikon Eclipse Ti-U inverted microscope FASTCAM MC2 camera (Photron)] and TEM (Hitachi H-7650).
Results: In these eighteen months 10 patients were diagnosed: 5 boys, median age 7.8 years (min 3; max 16.5 years). The most frequent clinical manifestations were: chronic productive cough (8), bronchiectasis (5), neonatal respiratory distress (6), chronic/recurrent otitis media (6), *situs inversus* (1) and hydrocephalus (1). Concerning nNO results, measurements were performed in nine patients, seven had abnormally low levels suggestive of PCD (median 26 ppb, min 9 ppb, max 159 ppb) and two patients had normal levels. With the HVMA, changes were detected in nine patients (main finding was uncoordinated and low frequency beating pattern) and one result was inconclusive. With TEM, ultrastructural defects were identified in eight patients (main finding involved defects in the dynein arms) and two results were inconclusive. Patients who presented with one normal/inconclusive diagnostic test had alterations in other two tests, allowing the PCD diagnosis confirmation. Genetic testing is still ongoing.
Conclusions: PCD is a rare disease that is currently still underdiagnosed or diagnosed late, since diagnosis requires complex execution and interpretation procedures. In our group, the occurrence of normal or inconclusive results imposed a frequent interdisciplinary discussion for the jointly interpretation of results. Given the complexity of these methods, international guidelines recommend that diagnosis be made in referral centres. The increase in diagnosis suspicion and availability of these methods will allow better patient identification and their timely multidisciplinary follow-up.

Key words: *Primary cliliary dyskinesia. Paediatrics. Nasal nitric oxide. Transmission electron microscopy. High speed videomicroscopy analysis.*

PRCC-010. PULMONARY ATRESIA VS POST TRAUMATIC LUNG COLLAPSE? A CLINICAL CASE

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Case report: YZ, male, 40 years old, trader, Chinese nationality, with a history of pulmonary trauma in childhood of which resulted sequelae pulmonary lesions that he can not specify, refers to the Emergency Service (SU) by vomiting, epigastric pain and asthenia with 4 days of evolution and progressive worsening, worsening after meals and no relief factors. On the physical exam performed on the ED it was noted discrete tachycardia, with cardiac heart rate of 100 bpm, absence of vesicular murmur on the left hemithorax and pain on the epigastric palpation without peritoneal reaction. From the analytical point of view we can demark leucocitosis without neutrofilia – Leuc 12,400 N 68% - discrete elevation of CRP (1.91). Ambiente air gasimetry: pH 7.42, pCO2 34.1, pO2 70.5, HCO3. 21.7, SatO2 94.7%, Lac < 1. Chest radiography showed mediastinal shift to the left and full opacification of the left hemithorax. The patient failed to clarify what kind of changes there were in radiographs taken previously. Given the gravity of the findings of the chest radiography and objective examination, and being unable to clarify the moment in which they arose, we opted for the hospitalization and started the diagnostic evaluation. On the first day of

hospitalization the patient underwent a chest tomography. The changes identified are: "mediastinal deviation to the left conditioned by the total collapse of the left lung which is reduced to thin area of calcified atelectasis, conditioning hiperexpansibilidade the right lung. A tubular image is noted relate to left bronchus completely filled by hypodense content, relatively homogeneous and can translate impaction of secretions/mucocele and that could be the cause of the atelectasis of the entire left lung cited above, but its nature is not clear. No pleural or pericardial effusion (...)". To determine whether the left bronchial obstruction was amenable to endoscopic correction, a rigid bronchoscopy performed during hospitalization: "(...) total occlusion/agenesis/ atresia of the Main Left Bronchus, observing discreet infundibulum at the site of its anatomical origin with normal mucosa at the level of carina (...). An acute gastroenteritis was diagnosed as the cause of symptoms and signs that initially motivated the dislocation to the ED and the patient was discharged with surveillance indicated by the treating physician. Morphological changes identified at the level of left bronchial tree with occlusion of the left main bronchus may have been the result of chest injury suffered by the patient in childhood, or even be congenital, and there was no possibility for endoscopic therapeutic intervention.

Discussion: Lung collapse is a rare condition that can result from blunt chest trauma. In cases of pulmonary atresia this clinical entity is usually detected and diagnosed early in childhood. The presentation of this case is relevant for the rarity of this clinical entity, especially when it comes to the adult patient, and for the peculiar images on CT and rigid bronchoscopy at the level of the carina.

***Key words**: Pulmonary atresia. Collapsed lung. Rigid bronchoscopy. Thorax CT.*

PRCC-011. ONE CASE OF DIFFUSE ALVEOLAR HEMORRHAGE WITH MULTIFACTORIAL ETIOLOGY

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Introduction: Diffuse alveolar hemorrhage (DAH) is a clinicopathologic syndrome featured by bleeding into the interior of the alveolar spaces resulting from disruption of the alveolar-capillary basement membrane. Recognition of DAH often requires bronchoscopy with bronchoalveolar lavage, since their clinical presentation is nonspecific and may include hemoptysis, anemia, diffuse pulmonary infiltrates and respiratory failure. Histopathological pattern may reflect the underlying etiology, which should be determined once diagnosis is established. The DAH is a medical emergency with high morbidity and mortality when treatment is not readily established.

Case report: The authors present the case of a 78 year old man with atrial fibrillation treated with apixaban 5 mg bid and amiodarone 100 mg three times a week. He was admitted to the emergency room for dyspnea, productive cough with purulent secretions and fever with five days of evolution. On admission, he presented respiratory failure type 1 and started coughing hemoptysis and bleeding at the insertion site of peripheral venous catheter that has not resolved with local measures, having received transfusion of two plasma concentrate units, with resolution of the blood loss. Analytically, showed fall in hemoglobin level of 0.6 g/dL, platelet count 174,000/uL, leukocytosis (12,840/uL) with neutrophilia (87.7%) and C-reactive protein of 2.62 mg/dL. Chest X-rays showed a hypotransparency in the lower field of the right lung and computed tomography of the chest revealed aspects suggestive of alveolar hemorrhage. The antigenuria to search for *Streptococcus pneumoniae* was positive. The patient was hospitalized for pneumococcal pneumonia associated with likely alveolar hemorrhage. Anticoagulation and amiodarone were suspended and he completed two weeks of amoxicillin/clavulanic acid and three days of azithromycin, without obvious clinical improvement of infectious frame. Bronchoscopy (10th day of antibiotics) documented abundant purulent secretions and mucous congestive tracheobronchial tree, friable, with easily bleeding controlled with saline. The study of the bronchoalveolar lavage was compatible with neutrophilic alveolitis and hemosiderin research in the cytoplasm of macrophages by coloring Pearls was positive, with Golde score 84.5, translating slight bleeding. Resumed anticoagulation to 20th day of hospitalization, initially with enoxaparin at prophylactic dose and later in therapeutic dose and after 27 days of hospitalization, re-started apixaban 2.5 mg bid with no new hemorrhagic complications. Bacteriological examination of bronchoalveolar lavage fluid allowed the isolation of *Pseudomonas aeruginosa* multiresponsive so it was administered one second antibiotic treatment cycle with piperacillin/tazobactam and gentamicin for two weeks with clinical, analytical and imaging improvement.

Discussion: The authors present a case of DAH with a probable multifactorial etiology and three potential triggers: anticoagulation with a new oral anticoagulant; amiodarone - an antiarrhythmic associated with multiple forms of pulmonary toxicity that, in rare cases, can manifest itself in the form of DAH; and, finally, the infection of lung parenchyma. The peculiarity of the present case also follows from the fact that it portrays a possible complication of the therapeutic approach of atrial fibrillation, the cardiac arrhythmia most frequently encountered in clinical practice.

***Key words**: Diffuse alveolar hemorrhage. Atrial fibrillation. Pneumococcal pneumonia.*

PRCC-012. RARE CAUSE OF LIFE-THREATENING HEMOPTYSIS

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Case report: The clinical case refers to a male patient, 31 years old, Caucasian, administrative, former smoker (average pack-years:15), with a history of Churg-Strauss vasculitis/ hypereosinophilic syndrome with multisystemic involvement (including lung) medicated with prednisolone and azathioprine; paroxysmal atrial fibrillation already subjected to 3 ablations; antifosfolipid syndrome, anticoagulated with warfarin; lacunar stroke without sequelae in 2012 and Type 2 diabetes. One week prior to admission is subjected to cardiac electrophysiological study with abnormal pathway ablation. After the procedure develops severe stenosis (greater than 95%) of the ostia of the left pulmonary veins, so that balloon angioplasty is performed with positive result by angiography and slight right pulmonary hypertension (25-30 mmHg) at right catheterization. The patient is hospitalized for sudden onset of left pleuritic pain and live blood hemoptysis (estimated amount of 150 ml) with a few hours of evolution. Denies other complaints. On examination: hemodynamically stable, eupneic, with bilateral and symmetrical vesicular murmur without adventitious sounds. Arterial blood gas without oxygen does not show respiratory insufficiency and chest teleradiography reveals slight interstitial markings in the left base. The blood tests shows no lowering of hemoglobin (Hg: 15.1 g/dl), without thrombocytopenia, INR 3.9, normal renal function and PCR: 0.31 mg/dL. It is administered 4 plasma units and phytomenadione for INR correction, maintaining, however, small amount of hemoptysis. Performs CT angiography that excludes pulmonary thromboembolism and shows no changes in pulmonary veins permeability. There are areas of focal hypotransparency in the left upper lobe and lingula (already present in a CT of February). Performs, too, a transthoracic echocardiogram that estimates a PSAP of 30 mmHg. In this context, performs emergency rigid bronchoscopy that shows active bleeding

from the posterior segment of the left lower lobe that is controlled with aminocaproic acid and topical epinephrine. The exam shows a diffuse hypervascularization throughout the bronchial tree, particularly at left. After the procedure the patient doesn't have new episodes of hemoptysis.
Discussion: One of the complications of abnormal pathway ablation is pulmonary veins stenosis with subsequent pulmonary hypertension, in this particular case aggravated by the existing vasculitis substrate, making the benefit of angioplasty possibly transient. It is assumed, therefore, that the changes found in bronchoscopy are in the context of acute pulmonary venous hypertension, probably by restenosis of the pulmonary veins. It is suggested to carry out a pulmonary venography and cardiac catheterization and consider the placement of stents in the pulmonary veins if stenosis is confirmed. After one week of hospitalization, by maintaining hemodynamic stability and not presenting new episodes of hemoptysis the patient is discharged with indication for hemodynamic reevaluation at the Cardiology Department. Since then he didn't have new episodes of hemoptysis.

Key words: *Hemoptysis. Atrial fibrillation. Abnormal pathway ablation. Pulmonary veins stenosis.*

PRCC-013. AN UNDER RECOGNIZED CAUSE OF BRONCHIECTASIS

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Introduction: Non cystic fibrosis bronchiectasis (NCFBQ) is an "orphan disease", underdiagnosed and with great variability in the pursuit of its etiology in clinical practice. Systemic diseases, including autoimmune and immunodeficiency diseases are the leading cause in developed countries, contrasting with the infectious etiology in developing countries.
Case report: We describe a 58-year-old female secretary. Non smoker. The patient had medical history of breast cancer in 2004, treated with right total mastectomy, chemotherapy and radiotherapy; total hysterectomy with salpingo-oophorectomy in 2006 due to uterine miomas. She was referred to pulmonary consultation following recurrent complaints of cough with purulent expectoration. Pulmonary lung function tests revealed mild obstructive ventilatory pattern and chest computed tomography showed cylindrical bronchiectasis in the lower lobes, middle lobe and lingula. The etiologic investigation excluded immunodeficiency and alpha 1-antitrypsin deficiency. Bronchoscopy revealed nonspecific inflammatory changes and bronchial secretions bilaterally and microbiological cultures were negative. During follow-up she presented with chronic diarrhea, and the diagnosis of ulcerative colitis was made by colonoscopy with biopsy a year after the first pulmonology consultation. Currently she has improved clinically, and is being treated with long-acting bronchodilators, systemic corticosteroids, azathioprine, mesalazine and isoniazid. She has also done pneumococcal and anti-influenza vaccinations and respiratory rehabilitation program.
Discussion: The authors intend to draw attention to the association between bronchiectasis and inflammatory bowel disease (IBD), in which the airway involvement can take many different manifestations. The first descriptions of the association between IBD and NCFBQ date from the 60s and the common embryologic origin of the respiratory and gastrointestinal systems suggest involvement in the same pathological process. Unlike most of the cases described, in this case, the respiratory symptoms preceded the diagnosis of IBD, highlighting the need to consider this diagnosis in patients with bronchiectasis.

Key words: *Bronchiectasis. Inflamatory bowel disease.*

PRCC-014. CAVITARY LUNG LESION: THE IMPORTANCE OF A TIMELY DIAGNOSIS

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Introduction: Pulmonary cavitations may have various aetiologies, including infectious and neoplastic causes. Cavitary lung cancer can be masked initially by an infectious process, leading to a late diagnosis and therefore advanced or metastatic presentation of disease.
Case report: A 68-year-old male, was referred to the Emergency Department with a productive purulent cough and hemoptotic sputum, anorexia and weight loss > 10%, with approximately 3 months progression. He had already completed two courses of antibiotherapy with amoxicillin/clavulate and was on the 7th day of levofloxacin with no improvement. He was a retired stonemason, ex-smoker (40-pack year), with moderate alcohol consumption (46 g/day), he had insulin-dependent diabetes mellitus, hypertension, dyslipidemia and duodenal ulcer. Physical examination showed white patches on the oropharynx with no other changes. Laboratory testing showed elevation of inflammatory parameters (leukocytes 20.0 G/L, CRP 22.0 mg/dL), hypochromic normocytic anemia, thrombocytosis, increased LDH (487 U/L) and normal arterial blood gas values. BAAR and urinary *Legionella* and *Strept. pneumoniae* antigen were negative. Chest radiograph showed a large cavitation with irregular borders in the upper third of the right lung. The patient was admitted and started on empirical antibiotherapy with piperacilina/tazobactam and topical oral Nystatin. On the 3rd admission day, due to evening rise of temperature, metronidazole was initiated. On day 7 flexible bronchoscopy was performed which showed diffuse inflammatory signs of the bronchial mucosa and thick secretions in the inlet orifice of the right upper lobar bronchi leading to a large cavitation with loss of the normal lung architecture and thick pus adherent to the wall. Biopsies for histopathological and microbiological analysis and BAL for mycobacteria CRP were performed. All were negative, except for bronchial biopsies, which showed an adenosquamous carcinoma over-infected by filamentous fungi. Thoracoabdominal CT showed a massive cavitated lesion (11.8 × 8.5 cm), well-circumscribed by solid and irregular borders in the apical and posterior segments of the right upper lobe (RUL) extending to the apex of the right lower lobe (RLL). Other cavitated lesions with thin borders were observed in the anterior segment of the RUL, in the middle lobe and the basal segments of the RLL, also two nodules in the posterior basal segment of the LID and various irregular nodular infra-centimetric opacities, some of them cavitated, throughout the left lung, suggesting metastasis. Subcarinal adenopathy and lytic lesions in the dorsal vertebrae and left shoulder blade were also present. There were no suspicious abdominal lesions. Bone scintigraphy confirmed bone metastasis. Head CT showed no alterations.
Discussion: Most cavitated lung tumours are squamous cell carcinomas. The cavitated lung carcinoma is associated with a poor prognosis due to diagnostic difficulties. The possibility of malignancy should always be considered in the differential diagnosis of slowly-resolving pneumonias in smokers. The absence of clinical signs and symptoms of infection and the presence of hemoptysis may be suggestions for the correct diagnosis.

Key words: *Cavitation. Lung cancer. Adenosquamous carcinoma.*

PRCC-015. BODY COMPOSITION AND PULMONARY FUNCTION IN PATIENTS WITH TYPE 2 DIABETES

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Introduction: Obesity is a chronic disease characterized by excessive body fat mass and is associated with the development of many other diseases such as type 2 diabetes. High levels of body fat appears also to cause respiratory changes with serious consequences on the functionality and quality of life of individuals. This study aimed to analyze the relationship between body fat and respiratory function in patients with type 2 diabetes.
Methods: There were analyzed 46 patients with type 2 diabetes candidates for *Diabetes em Movimento*® Vila Real (NCT02631902) – a community-based exercise program – in an analytical cross-sectional study (29 women and 17 men, 63.61 ± 7.06 years of age; HbA1c 6.57 ± 0.92%; 5.63 ± 4.26 years of diabetes; IMC 30.00 ± 4.94 kg/m^2). Body fat (%) was assessed by bioelectrical impedance analysis (Tanita, BC-418 MA) and respiratory function was assessed by spirometry (MiroLab 3500 UK v6.00), with evaluations of peak expiratory flow (PEF), forced expiratory volume in one second (FEV_1), forced vital capacity (FVC), ratio FEV_1/FVC and maximum expiratory flow (MEF 25/75), according with ATS/ERS criteria.
Results: Participants presented average fat mass levels of 38.90 ± 9.11%; PEF of 288.67 ± 85.69 mL (77.59 ± 19.73%); FEV_1 of 2.20 ± 0.51 mL (100.54 ± 23.75%); FCV of 2.95 ± 0.68 mL (109.93 ± 22.30%); FEV_1/FVC of 76.22 ± 8.93%; and MEF 25/75 of 1.73 ± 0.81 mL (60.00 ± 25.12%). The statistical analysis identified negative and significant correlations between fat mass and the values of PEF ($r = -0.374$, $p = 0.010$), FEV_1 ($r = -0.329$, $p = 0.025$), FVC ($r = -0.516$, $p < 0.001$) and FEV_1/FVC ($r = -0.378$, $p = 0.010$). It was not found a significant correlation with the values of MEF ($r = 0.104$, $p = 0.490$). The linear regression models revealed that fat mass is capable of predicting 14% of PEF ($R^2 = 0.140$, $\beta = -0.374$), 11% of FEV_1 ($R^2 = 0.108$, $\beta = -0.329$), 27% of FVC ($R^2 = 0.266$, $\beta = -0.516$) and 14% of FEV_1/FVC ($R^2 = 0.143$, $\beta = -0.378$).
Conclusions: The results of this study suggest an important relationship between body fat and respiratory function, with higher body fat levels predicting a worse respiratory function, especially a lower forced vital capacity. Intervention strategies to reduce overweight and obesity levels may contribute to the improvement of the respiratory function of these patients and a better quality of life, which could include exercise training and pulmonary rehabilitation.

Key words: *Pulmonary function. Type 2 diabetes. Spirometry.*

PRCC-016. CAMPAIGN "CARING OF WHO TAKES CARE OF US" OR THE PROFILE OF THE RESPIRATORY HEALTH OF PORTUGUESE FIREFIGHTERS: EVALUATION OF THE RESPIRATORY IMPACT IN FIRST RESPONDERS (FR)

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Introduction: Around 3 million and 600 acers of forest have burned in the last 25 years, the majority of first responders (FR) are firefighters from which 2/3 of them are volunteers and their health is not being monitored. Recent studies indicate that the repeated exposure to smoke are the origin of inflammation and the increasing bronchial reactivity and obstruction. The Portuguese firefighters (PF) who make part of the FR are exposed to a wide variety of chemicals and pollutants.
Methods: In 2007 we initiated medical plan for health monitoring – HM – campaign "Caring of who takes care of us" to evaluate the PF state of health and respiratory impact in the FR in order to build a health profile and estimate the lesion risk, establish the prevalence of respiratory pathologies and elaborate norms/measures. Simultaneously we involved the media (TV, press, blogs, social media and web) in order to create awareness among society as well as the government towards this issue. In addition we created leaflets about "Recommendations in case of fire" and "Avoid the carbon monoxide". We organized the first conference about firefighter's health and collaborated in numerous others. Launched a population oriented campaign "Where there's smoke there is more than fire" and implemented partnerships to ensure HM to the PF. Population selection: PF from corporations from the most fire fustigated areas (from 5 to 10 PF of the FR); evaluated outside of period Charlie. HM: Supervised auto fill of a questionnaire, spirometry (SPR) tests, medical advisement towards smoking habits and referral if pathology was found. 42 spirometrys excluded.
Results: Complete HM 582, PF from FR of 57 corporations, mean age of 34 (17-68); 13.4% female and 86.6% male; smoking habits (SH) (smokers 43.6%, ex-smokers 22.6%, non-smokers 33.6%. Physical exercise 47.9%, physical training 30.2%. Use of protection equipment: individual (IRPE) 73.5%, respiratory 50.1%. IMC > 30 21.5%. Years of service (YS) of PF (until 5YS-25.4%, 6 to 10YS – 27.2%, 11 to 15YS – 19.3%, 16 to 20YS – 10.6%) History of respiratory diseases 17.3%; current respiratory symptoms "Do you have any of these symptoms?":dyspnoea (loss of breath) 9.9%, Respiratory difficulty: when walking fast 14.7%, accompany 3.9%, hygiene 1.9%, cough and sputum (S) 21.3%, S 2 months/year 4.5%. hemoptysis S 0.8%, morning cough 14.7%, lying down 5.5%, pieira 8% (working 6.6%). Obstructive syndrome SPR 5.4%, small pathways obstruction 12.2%. Correlation $p < 0.05$ linking YS and changes in SPR and independence between SH and spirometry changes.
Conclusions: Changes in SPR related to YS and symptoms in a young population impose the necessity of a yearly respiratory supervision and, in present setting, have a direct connection to firefighting activity meaning that there is a professional pathology, which can give place to incapacity indemnity even in volunteers. To the PF we propose a change in habits (smoking, use of IRPE, weight loss and an increase in physical exercise). As a result from our campaign, in 2015 Ministério da Administração Interna/Liga dos Bombeiros Portugueses introduced systematic screening in PF not including SPR.

Key words: *Portuguese firefighters (PF). Forest fires. Respiratory symptoms. Spirometry. "Caring of who takes care of us".*

PRCC-017. THE ELDERLY IMPORTANCE IN A DEPARTMENT OF PULMONOLOGY – 2010-2015: NEW ATTITUDES TO TAKE?

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Nacher wrote, in 1909, for the first time the word Geriatrics. The first unit of Geriatrics (Specialized Geriatric Assessment Unit) was born in Great Britain in 1948, with Marjory Warren, considered the mother of Geriatrics. In Portugal, the life expectancy at birth is 79.78 years (2010-2012 data). The aging index of the population (number of elderly people per 100 young people) increased from 102 in 2001 to 128 in 2011 (Census from 2011). Aging changes the absorption, distribution, metabolism and excretion of drugs, affecting the pharmacodynamics and pharmacokinetics. The authors conducted a retrospective study in Pneumology B Service (HG-CHUC) to study the impact of the elderly population (≥ 65 years) in pulmonology appointments (general and subspecialties), and hospitalization during 2010-2015 (6 years). During this period it was found that the impact of the elderly population in the Pulmonology internment, corresponding to 69.3% of a total of 3674 patients, of which 42.3% (n = 1,553) were between 65-80 years old. The Pneumology Department has 12 subspecialties differentiated by respiratory disease. During the study period they were carried

out 56,407 appointments of which 12,454 were first ones and 43,963 subsequent ones. The impact of appointments in elderly patients corresponds to 45.2% of which 37.5% corresponded to the age group from 65 to 80 years. The appointments with the highest percentage of elderly were the Functional Rehabilitation Respiratory Consultant (FRRC) Respiratory Rehabilitation Program Consultation, the Oncology Pulmonology Consultation (OPC) and the General Pulmonology Consultation (GPC) with 71.6%, 62 2%, 53.3% and 50.7%, respectively. In FRRC, the group of 65 to 80 years represented 53.7% and 81 to 90 years 17.9%. The OPC had 53.3% elderly, in a total of 13,266 appointments, and 48.3% aged between 65 and 80 years. The GPC had 50.7% elderly, a total of 13,887 appointments. The age range of 65 to 80 years had an impact of 40.9% in the group. As expected, the Allergology Respiratory Consultation had high amount of young adults, and the elderly had an impact of 13.9% (9,304 appointments). When we analyse the Severe Asthma Consultation Record, the elderly represented 12.4% in 957 appointments. We can conclude that the elderly represented more than 2/3 of patients admitted in the Pulmonology Department, and General Pulmonology Consultations begin to be predominantly elderly, and there may be need to rethink the way that it is working and what needs to changes towards this new reality. In subspecialty appointments, the elderly patients had high participation in specific appointments for respiratory failure patients due to COPD/ heart disease..., as well as Oncology. These will tend to be increased by elderly population, because the cancer disease has a higher incidence in the elderly, and COPD has been increasing in the general population. "The future does not belong only to the elderly, but also will not be made without them" Michel Loriaux in 1994.

Key words: *Department of Pulmonology.*

PRCC-018. END-OF-LIFE IN ADVANCED CHRONIC RESPIRATORY DISEASES: THE PRACTICE OF CARING

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Introduction: Chronic respiratory disease is a group of diseases that causes a reduction in quality of life, reduced life expectancy and considerable suffering for patients and families. There has been greater interest in palliative aspect of care in these patients and it is recommended an integrated palliative approach. The provision of quality care at end of life, should be a concern in a Pulmonology Department.
Objectives: Evaluate interventions provided to respiratory terminally ill patients in the last 72 hours of life.
Methods: Descriptive retrospective study of the deceased chronic respiratory patients in our ward in the last two years.
Results: During this period, 54 patients died. Although the most common primary diagnosis were cancer (39%), many patients presented other chronic respiratory diseases: obstructive pulmonary disease (26%), interstitial diseases (17%) and terminal evolution after lung transplant. Despite the most frequent symptom at admission was dyspnoea (67%), there wasn't a methodic evaluation of it with an appropriated scale. Only 37% of patients were referred to the hospital Palliative Care Team; in 9 cases this was done in the last 72 hours of life. In 100% of the patients there was a systematic evaluation of pain with the pain numeric scale and most of them registered some relief: 29 patients were treated with strong opioids, 16 had breakthrough analgesic and in 9 patients was used palliative sedation. In clinical diaries of the majority of patiens there wasn't any note for the recognition of agony, and there were only a few patients with a clear intervention plan and care reorientation in this time. Few patients had clearly stated an advanced care plan treatment, namely concerning decisions not-to intubate, directive of wills of the patient, beliefs or care to the family. The interaction with other professionals to manage the not physics aspects of caring in the end-of-life (spiritual, psychological, social) was residual.
Conclusions: There are still difficulties in recognizing proximity to end-of-life and in the reorientation of the interventions to the symptom control, patient comfort and family support. More than in an oncologic setting, in the chronic respiratory diseases, because of the variable and uncertain natural disease history, it became more difficult to recognize this stage. So it is essential a team work to improve the attention to the signals of imminent dead, creating a sensibility to respect the dignity and the beliefs of the patient, avoiding unnecessary and invasive procedures and creating an efficient plan of sedation. We need to improve our competences in such matters to alleviate the suffering of our patients and their families. The evaluation of the care to our respiratory patients in the end-of-life must be a quality index of our work but also an opportunity of becoming better and constant learning.

Key words: *Palliative care. Comfort. Respiratory patients.*

PRCC-019. IMMUNIZATION OF HEALTH CARE WORKERS AGAINST INFLUENZA REDUCES ABSENTEEISM?

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Introduction: Immunization of health care workers against influenza should be considered beneficial in reducing infection, absenteeism at work and in the protection of patients being recommended by health authorities. The IPO Oporto has an annual vaccination program established by the Occupational Health Service on a voluntary basis, starting in the month of October, involving all professionals (population: 1968 workers; with 347 doctors, 660 nurses, 465 operational assistants (OA) and 496 other professional).
Objectives: The association between influenza vaccination of health care workers and their absenteeism was investigated in three consecutive seasons; 2012/13, 2013/14 and 2014/15.
Methods: Age, sex and professional group were analysed in a total of 642, 615 and 574 vaccinated health care workers in the 3 seasons respectively, and the association with the number of days of absenteeism defined as at least one period of less than 30 days in the months of October to May. It was performed a logistic regression analysis with variable response the occurrence of at least one period of absenteeism, stratified by professional group and by the immunization season.
Results: In 2012/13 season, 89 doctors, 211 nurses, 189 OA and 153 other employees were vaccinated; in 2013/14 season, 81 doctors, 196 nurses, 189 OA and 149 other employees and in 2014/15, 78 doctors, 171 nurses, 183 OA and 142 other employees. There was no gender difference with regarding to immunization adherence. The professional group with greater immunization adherence was the OA (39.9%). The median age of nurses vaccinated was consistently higher than the non-vaccinated ($p < 0.05$). In the OA group this fact was noted just in the last season. The average number of days of absenteeism in the workplace according to professional group was 1.1; 1.8; 3.2 and 1.6 for the vaccinated and 1.8; 1.9; 3.4 and 1.7 for the unvaccinated (respectively in the doctors, nurses, OA and others). After adjusting for sex and age, no increased chances of absenteeism, with statistical significance, was observe in the unvaccinated group. Statistical analysis also showed that in non-vaccinated doctors, the number of days of absenteeism was higher in all the three seasons and in the nurses and OA groups this increased absenteeism rate also occurred in the first two seasons.

Conclusions: We conclude that these data are according to the few existing studies published on this topic. It should be noted that the voluntary basis of the vaccination program may select the most compliant with the rules the rules against infection. Immunization of health care workers against influenza is associated with fewer days of short duration absenteeism during the period from October to May. Other prevention programs can be discussed.

Key words: *Health care workers. Absenteeism. Influenza and immnunization.*

PRCC-020. TELE NURSING IN AN INTERVENTIONAL PNEUMOLOGY UNIT (IPU)

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Introduction: According the International Council of Nurses (2011) tele-nursing is considered as the practice of nursing remotely via telecommunications, aiming to improve attendance, quality and access to multiple health services. What will be the effect of a phone call on the life and health project of the user who performs tests on this unit? In order to address this issue, a project to improve the quality of care was implemented: Nursing in IPU - A phone call makes a difference.
Objectives: The overall objective of this project was to improve the quality of care provided to users on an outpatient basis, subject to examination in IPU. The specific objectives were to reduce the faults to users of scheduled exams and promote the fulfillment of the prerequisites for the exams.
Methods: We proceeded with the elaboration of a semi-structured telephone interview to inform/validate compliance with the prerequisites and answer questions about the procedures, providing an orientation space for users promoting continuity of care. The choice of this form of communication has taken into account the flexibility, the speed of service and its low cost. It was chosen a project method and a retrospective quantitative study, of the descriptive type to evaluate it, using the semi-structured interviews as a data collection method. All users on an outpatient basis with computer scheduling timely and telephone contact were included. Defined as prerequisites: the presence, the fulfillment of fasting (if applicable), timely suspension of antiplatelet/anticoagulant medication and be in possession of diagnostic tests required for the exams.
Results: In the period analyzed (1/7/2015-30/6/2016) of a total of 658 cases, 646 telephone contacts were made (12 cases were excluded). 550 effective telephone interviews were made (85%). 98.72% (543) of users with telephone interview met the prerequisites for the exam. Comparing the number of absences among the users interviewed and not interviewed, we obtained a value of statistical significance ($p < 0.01$). It was found that of the 189 patients under antiplatelet/anticoagulant medication 32.8% had no medical information to discontinue the medication and this situation was solved after the interview in all cases.
Conclusions: It was concluded that the implementation of this project has significantly reduced the number of absences to the examination and contributed to increase the compliance of the prerequisites for the exams in the Unit, providing obtaining health gains. It has also identified the patients under antiplatelet/anticoagulant medications without information and directing them to the attending physician in order to provide compliance with this prerequisite in time. Our analysis also suggests that besides increasing the fulfillment of the prerequisites for the exams, the telephone interview also improves knowledge and decreases anxiety related to the technique, this issue is the object of an ongoing investigation.

Key words: *Nursing care. Health promotion. Telenursing.*

PRCC-021. A RESISTANT HYPOXEMIC FAILURE: A RARE CAUSE

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Introduction: Patent foramen ovale (PFO) is a common condition, present in 25% of general population, due to incomplete fusion of the two embrionary layers, the *septum primum* and the *septum secundum*, leading to a residual interauricular communication. Clinically, PFO relates to cryptogenic stroke, platypnea-orthodeoxia syndrome, decompression illness and paradoxical embolism.
Case report: We present a clinical case of a 75 years-old woman with previous diagnosis of asthma, obstructive sleep apnea (OSA), arterial hypertension, dyslipidemia, previous stroke without sequelae (14 years ago). She was admitted in the context of acute asthma exacerbation due to upper airway respiratory infection and partial respiratory failure. During in-patient treatment the patient developed acute aphasia and was submmited to Head – computed tomography (CT) that revealed an ischemic stroke of middle cerebral artery. The transthoracic echocardiogram was normal and a Holter ECG which revealed periods of paroxistic auricular tachycardia. We admitted the diagnosis of cardioembolic stroke. The patient presented several episodes of oxygen desaturation, confirmed by blood gas analysis (hypoxemia with signs of alveolar hyperventilation). The diagnosis of pulmonary embolism was considered. A CT angiogram was performed and no perfusion defects was detected in that main pulmonary artery. We considered the diagnosis of intracardiac shunt and was confirmed with the transesophageal echocardiography which revealed "a septum interatrial thin, hypermobile, sugesting fenestrated septum. Conclusion: Patent Foramen Ovale". The patient was transferred to the Cardiology Department and heart catheterization was preformed to treat the atrial septum defect. They performed the correction with good results.
Discussion: This clinical case showed the importance of the clinical suspicion and the diagnostic management for PFO. The treatment resolves immediately and effectively the right-left shunt and the severe respiratory insufficiency.

Key words: *Patent foramen ovale. Respiratory failure. Stroke.*

PRCC-022. FROM 100% TO 21% (AT DAY TIME)

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Introduction: Idiopathic pulmonary arterial hypertension is a rare condition leading to heart failure and acute respiratory failure (ARF). When intensive care unit admission and mechanical ventilation is needed it often leads to death. The early start of specific therapy is the key for a better prognosis. High-flow nasal cannula oxygen therapy is a method of oxygenation that alleviates symptoms and improves oxygenation by several mechanisms in a comfortable way to the patient.
Case report: We describe the case of a 41 year-old man, current smoker of 20 pack-year and former user of inhaled opiates. His past medical history includes hepatitis C virus infection without follow up. Transthoracic echocardiography done three months before his admisson showed signs of pulmonary hypertension classified with PSAP 98 mmHg. He was schedule for right heart catheterization but, due to clinical deterioration, was admitted to the hospital. On admission at our intensive care unit, he presented with severe

dyspnoea, orthopnoea, anasarca and acute respiratory failure with PaO_2/FiO_2 = 49. Blood test showed active replication of HCV with viral load of 236,268 U/L (genotype 1a); HIV serology was negative. Right heart catheterization showed mean pulmonary artery pressure (mPAP) of 60 mmHg and mean pulmonary capillary wedge pressure of 12 mmHg. With the confirmation of pulmonary arterial hypertension, he was immediately started on intravenous epoprostenol, ambrisentan, vasopressors and high-flow nasal cannula oxygen therapy (FiO2 = 100%). After clinical improvement it was possible to proceed with etiologic investigation: pulmonary embolism was excluded; abdominal ultrasound showed chronic hepatic disease but without portal hypertension. An Hickman catheter was implanted and epoprostenol progressively increased to 14 ng/kg/min before discharge. The patient final diagnosis was pulmonary arterial hypertension group 1. Six months later he is on 20 ng/Kg/min of epoprostenol, 5 mg/day of ambrisentan and 3 L/min nocturnal oxygen and in NYHA class I. He is waiting for HCV therapy.

Discussion: This case-report describes the successful treatment of a patient with group 1 severe pulmonary hypertension presenting in cardiogenic shock and severe respiratory failure. Prompt combined therapy prostanoids clearly contributed to this good results. The efficacy on delivery of oxygen through high flow nasal cannula avoided mechanical ventilation, proving its usefulness in managing PAH patients.

Key words: *PHT. Acute respiratory failure. Epoprostenol. High-flow nasal cannula oxygen.*

PRCC-023. THE OUTPUT OF A MAZE IN PULMONOLOGY

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Hospital Garcia da Orta.

Introduction: Organizing pneumonia is a diffuse interstitial lung disease that can be classified as cryptogenic or secondary when associated with multiple factors, such as lung infections, connective tissue diseases or drug toxicity.

Case report: We present the case of a 81-year-old male patient, who was admitted to our hospital because of a 3-months history of dyspnea and left pleuritic chest pain, associated with cough with purulent expectoration, anorexia and weight loss for two weeks. His medical history was remarkable for pulmonary tuberculosis at age 25 with right upper lobectomy and passive exposure to tobacco smoke for over 30 years. On admission, he was febrile and diminished breath sounds and crackles were audible in the lower half of the right lung field. He presented leukocytosis with neutrophilia and increased C-reactive protein and the gas analysis revealed a gmoderate hypoxemia. In the thorax CT results, a total collapse of the right upper lobe, and multiple bronchiectasis and consolidations with air bronchogram on the lower lobe were identified. On the left hemitorax we could also see multiple consolidations or ground-glass opacities: two with 16 and 18 mm in the upper lobe, and several in the lower lobe, namely one with irregular borders and 6 cm of diameter. The initial diagnostic hypotheses were community-acquired pneumonia with infectious exacerbation of bronchiectasis, and empiric antibiotic therapy was started. To exclude lung cancer, flexible bronchoscopy was applied to the patient. The cytological examination of bronchial brush in the left lower lobe showed suspicious changes for malignancy. In the absence of response to antibiotic therapy, we decided to repeat bronchoscopy and samples were collected for the detection of *Mycobacterium tuberculosis* by PCR amplification and Galactomannan antigen. All results came negative. At the same time, serum Galactomannan was tested and came positive. Based on these findings, the patient might have a fungal infection of unknown origin and voriconazole was started. Other diagnostic procedures were applied to the patient and it was possible to identify an inflammatory process with an organizational standard by histological examination of transthoracic lung biopsy, and the presence of fungi in the cytological examination of bronchial secretions. The diagnosis of an organizing pneumonia probably secondary to fungal infection was assumed, and prednisolone was added to therapy. A clear clinical, analytical and radiological improvement was observed and the patient was discharged for follow-up in general pulmonology consultation. Currently, he maintains progressive improvement and corticosteroids in decreasing doses.

Discussion: The authors wish to emphasize that, although rare, fungal infection can be a cause of organizing pneumonia, as was presumed in the case of this patient. Furthermore, we want to point out that the quality and quantity of the samples are of utmost importance for the diagnosis of organizing pneumonia, making it a challenge often with the need for more invasive tests.

Key words: *Organizing pneumonia. Fungal infection. Pulmonary consolidations.*

PRCC-024. EPIPHRENIC DIVERTICULUM COMPLICATED BY ASPIRATION PNEUMONIA

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Introduction: Epiphrenic diverticulum is a rare condition (prevalence ≤ 4%). It is usually caused by pulsion mechanism due to an esophageal motility disorder, as achalasia. A wide spectrum of symptoms is associated with this disorder, such as anorexia, weight loss, dysphagia, regurgitation, chest pain, odynophagia and nocturnal cough (a symptom of aspiration). The awareness of differential diagnoses in the presented clinical case allowed the authors its complete resolution.

Case report: A 44 year old ex-drug male addict (abstinence for 10 years) was previously diagnosed with achalasia and a large epiphrenic diverticulum. He was evaluated in the Urgency Room for anorexia, weight loss, asthenia, mucopurulent sputum and cough for one week associated with fever (axillary temperature: 37,9 ºC). He presented with 22,4 mg/dL C protein reactive and he underwent a thoracic CT that was described as a central mass (47 mm) associated with incarceration and bronchial and vascular invasion of the right hilum with air bronchogram involving the right superior lobe and the apical segment of the right inferior lobe. The patient was hospitalized. Because of the clinical presentation and the previous esophageal disorders presented by the patient, the main diagnostic hypothesis was pneumonia (by aspiration or obstruction) instead of pulmonary nodule of other aetiologies. It was started amoxicillin/clavulanic acid and azithromycin. During hospitalization, he was subjected to a bronchoscopy that revealed no abnormalities other than appearing with a hyperaemic mucosa on the right and left bronchial tree compatible with inflammatory signs. The performed transbronchial biopsies were negative for neoplastic cells and presented with interstitial inflammatory infiltrate due to pneumonia. There were no positive bacterial results in the bronchoalveolar lavage. On the 5th day of hospitalization because of the fever reappearance (39.1 ºC) and hypoxia he performed an X-ray that revealed unilateral pleural effusion. He was submitted to a thoracocentesis with drainage of 400 mL of serous fluid that revealed to be an exudate by Light's criteria. Adenosine deaminase, cultural tests and neoplastic cells of the pleural effusion were negative. After changing antibiotherapy to piperacillin/tazobactam there was complete resolution of the clinical and radiological condition.

Discussion: Over the years epiphrenic diverticula have been associated with respiratory complications as laryngitis and aspiration pneumonia. There have been reports of 45% of prevalence for aspiration and 15% of potential life-threatening

pulmonary complications in patients with thoracic esophageal diverticula. We present a case with symptoms explained by aspiration pneumonia due to an epiphrenic diverticulum. The authors pretend to reveal with this case the importance for differential diagnosis with esophageal diseases and its treatment in patients with respiratory symptoms.

Key words: *Epiphrenic diverticulum. Aspiration pneumonia.*

PRCC-025. AN INTERESTING CASE OF BILATERAL PULMONARY CONSOLIDATION

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Introduction: Faced with a case of dragged "pneumonia" that does not resolve with antibiotic treatment, differential diagnosis of bilateral pulmonary consolidation should be considered. Cryptogenic organizing pneumonia (COP) is a rare disorder characterized by cellular granulation tissue, myofibroblasts and connective tissue infiltration on the alveolar wall, which should be considered when other causes, including infectious, connective and neoplastic pathologies or drugs, are excluded. The definitive diagnosis is made histologically, and the recommended treatment is corticosteroids, often with good response but a significant relapse rate.
Case report: We present the case of a patient of 53 years old, female, with unquantified fever and odynophagia, initially self-medicated with symptomatic therapy. By persistence of symptoms she was observed in the Emergency Room. Analytically, elevated inflammatory parameters with maximum CRP 25 mg/dL, VS 79 mm/h. Chest radiograph showed bilateral hypotransparency on the bases. Medicated empirically with moxifloxacin and later with amoxicillin/clavulanate and clarithromycin without clinical benefit or analytical improvement. Computed tomography (CT) of the chest revealed "areas of pulmonary condensation with air bronchogram covering the lower lobe, central region of the right upper lobe, middle lobe and left lower lobe suggestive of infectious process". Flexible bronchoscopy showed no changes. The bronchoalveolar lavage revealed a predominance of lymphocytes (84.1%); negative for cancer cells. Bacteriological and mycobacteriological examination were negative. Bronchial biopsy revealed only mild inflammation. She was admitted to the Pulmonology Department by progressive clinical deterioration with progressive weight loss, dyspnea and hypoxemia (PO2 68). Medicated empirically with piperacillin/tazobactam, without improvement. Blood cultures and bacteriological examination of the sputum were negative. The serology for HIV, CMV, EBV, *Mycoplasma pneumoniae, Chlamydophila pneumoniae, Coxiella burnetti* and respiratory viruses were negative for recent infection. The assay of autoantibodies and tumor markers were negative. Rapidly progressive deterioration of clinical and imaging in one month, with suggestive imaging pattern of lung cancer. Transthoracic lung biopsy was inconclusive (exuberant inflammatory pattern). Surgical lung biopsy via thoracoscopy showed organizing pneumonia of unknown cause (COP). Started corticosteroid therapy with prednisolone 1 mg/kg/d, verifying clinical, radiological and analytical improvement with reverse of the hypoxemia. CT scan after 3 months of treatment showed complete resolution of the lesions and negative inflammatory parameters without signs of relapse of the disease.
Discussion: COP is a rare disease, very rewarding regarding the excellent response to therapy. The authors aim with this clinical case is to illustrate the importance of the differential diagnosis of this identity in patients with bilateral pulmonary consolidation without resolution with antibiotic therapy, requiring extensive diagnostic march to exclude underlying causes and obtaining histological diagnosis. Treatment options and relapses of disease are discussed.

Key words: *Cryptogenic organizing pneumonia. Corticosteroid therapy. Thoracoscopy.*

PRCC-026. NOCTURNAL SOUND BEYOND THE SNORING: CATATHRENIA

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Introduction: Catathrenia, also known as sleep-related groaning, is characterized by a prolonged expiration with monotonous vocalization resembling groaning. It is now considered a rare sleep-related breathing disorder but was previously classified as a parasomnia.
Case report: The authors report the case of a 39 year-old women evaluated in the outpatient clinic for respiratory sleep disorders. The main complaint was the presence of "abnormal sounds during sleep" observed by the husband. The sounds occurred during the expiration and began during the adolescence. There was no presence of excessive daytime sleepiness (Epworth sleepiness scale 9/24P) or any other known sleep disorder. The medical history did not reveal relevant abnormalities. Physical examination was normal with exception of a reduced BMI (17,6 Kg/m^2). Standard polysomnography (PSG) demonstrated decreased sleep efficiency (70.8%) with initial and maintenance insomnia. The apnea-hypopnea index (IAH) was normal (4.2/h). The breathing events occurred almost exclusively during rapid eye movement (REM)-sleep (IAH REM 10.3/h) with a central/mixed apnea/hypopnea pattern. The microphone recording documented a high frequency groaning during a prolonged expiration, which permitted the diagnosis of catathrenia. Catathrenia is considered a rare benign sleep disorder. However, it often presents a morose or sexual connotation causing a social problem. Its true prevalence is unknown, there are a few dozen of case in the literature. The typical PSG is characterized by a deep inspiration followed by a groaning during a prolonged expiration. This is the hallmark that distinguishes catathrenia from obstructive respiratory sounds that occur more likely during inspiration. Due to bradypnea with no relevant respiratory movement, it may be misdiagnosed as a central apnea. Oxygen dessaturations are unlikely but the respiratory events may be associated with bradycardia and are preceded by a deep inspiration. The differential diagnosis is broad and includes snoring, nocturnal wheezing, laryngospasm and stridor.
Discussion: The treatment of catathrenia is not well defined but it has been proposed continuous positive airway pressure by some authors. This interesting clinical case report aims to raise awareness to this rare breathing-sleep disorder that must be included in nocturnal sound differential diagnosis.

Key words: *Sleep disorders. Catathrenia.*

PRCC-027. PULMONARY INFILTRATES WITH EOSINOPHILIA. ONE CLINICAL CASE, TWO DISTINCT SYNDROMES

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Case report: We present the case of a 32 year-old female, born in Timor, living in Portugal for the last 20 years, non-smoker and working as a waitress. Two years previously she was admitted to the department for infectious diseases due to a hypereosinophilic syndrome with pulmonary, cardiac and bone marrow involvement. She had peripheral eosinophilia, eosinophilia in the bronchoalveolar

lavage and bone marrow biopsy, and an MRI showing myocardial involvement. After an extensive evaluation, no etiologic cause was found and she was discharged, clinically better, medicated with prednisolone 7.5 mg per day. However, approximately 5 months after discharge, she abandoned the infectious diseases appointments and suspended the medication. Two years later she was admitted to our pulmonary department with complaints of pleuritic chest pain for 1 week. She also reported gastrointestinal disturbances with an increased frequency of dejections. Analytically she presented peripheral eosinophilia but there were no signs of renal, hepatic or cardiac involvement. The chest X-ray showed bilateral pulmonary infiltrates and she had hypoxemia. The patient denied recent travels abroad and taking medication. The thoracic CT showed peripheral, bilateral, ground glass opacities, without pleural effusion or lymphadenopathy. From the etiologic study performed she presented eosinophilia in the bronchoalveolar lavage with sterile microbiology, however, there was an antigenic positivity for *Entamoeba histolitica* in the faeces. Due to this result she was medicated with metronidazole for 10 days and systemic corticosteroids (prednisolone). There was clinical and laboratorial improvement with eosinophilia resolution, gasometric and imagiological normalization. She continues to be followed up by our pulmonary department and is still medicated with prednisolone 5 mg per day in a reduction scheme, without evidence of relapse from both eosinophilia and radiographic changes. The hypereosinophilic syndrome is characterized by persistent eosinophilia associated with signs and symptoms of multi-organ damage after exclusion of other causes of eosinophilia (parasitic infection, atopy, etc.). It may occur due to a clonal bone marrow expansion however, in the majority of cases (75%), the true etiology remains unknown. Treatment consists in immunosuppressive medication, usually with a good response to systemic corticosteroids which are maintained at the minimal dose necessary to control the eosinophilia and associated symptoms. Pulmonary infiltrates associated with eosinophilia may have numerous causes, one of them the hypereosinophilic syndrome. However, the most frequent causes are parasitic infections and systemic reactions to drugs. It is important to always exclude these possibilities.

Discussion: We present the case of a patient with a personal history of hypereosinophilic syndrome, who subsequently developed an eosinophilic pneumonia secondary to parasitic gastrointestinal infection. This example serves to point out the importance of reevaluating the eosinophilia's etiology even where there is an apparent relapse following the initial illness.

Key words: *Hypereosinophilic syndrome. Secondary eosinophilic pneumonia.*

PRCC-028. PHANTOM PLEURAL METASTASIS

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Introduction: Phantom or vanishing tumor of the lung can be the only manifestation of heart failure. It results from transient loculation of a pleural effusion within an interlobar fissure. The existence of pleuritis with adhesion and obliteration of the pleural space contributes to the development of this entity. Phantom tumors are tipically found on chest radiographs and more commonly in the right hemithorax.

Case report: We report the case of an 82-year-old male with history of type 2 diabetes mellitus, essential hypertension, severe aortic valvular stenosis with implantation of a bioprosthetic aortic valve in 2015, ischemic heart disease, peripheral artery disease and NYHA class II heart failure. The patient was hospitalized with Malory-Weiss syndrome characterized by gastrointestinal bleeding and type 2 myocardial infarction. The chest radiograph performed during hospitalization showed multiple micronodules in the left hemithorax that were not present in the previous exams. For better clarification of the micronodules a chest computed tomography (CT) scan was performed and showed a left pleural effusion as well as a nodular thickening of the pleura in the right oblique fissure. In addition, there were also mediastinal lymphadenopathies. These findings were suggestive of pleural metastasis. Because the patient had symptoms compatible with decompensated heart failure he initiated appropriate medication. After the pharmacological treatment of heart failure, a chest-abdomen-pelvis CT scan was performed to study a presumable cancer of unknown primary origin. The chest CT scan showed complete resolution of the pleural nodular thickening. This form of presentation is characteristic of phantom or vanishing tumors of the lung.

Discussion: Its name is due to the fact that it vanishes after appropriate management. Phantom tumors can recur with decompensated heart failure episodes. Therefore, this diagnosis must be considered in any patient with history of heart failure and an apparent lung mass on imaging studies. The identification of this radiological finding is important to avoid unnecessary diagnostic and therapeutic procedures.

Key words: *Phantom tumor. Heart failure. Pleural effusion. Pleural metastasis.*

PRCC-029. MULTIPLE PULMONARY NODULES APPROACH. CLINICAL CASE

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Centro Hospitalar Lisboa Norte.

Introduction: The presence of multiple pulmonary nodules, especially in patients with high risk of cancer, remains a diagnostic and therapeutic challenge. Multiple diseases in addition to cancer, may manifest themselves in the form of nodules. The initial approach passes by assessment of the risk and radiological characteristics of nodules - size, number, location and morphology - but often the diagnosis is inconclusive implying the use of other complementary means to exclude malignancy or obtain alternative diagnosis.

Case report: The authors describe the case of a 47 years old man, smoker, with no previous known diseases, who appealed to the emergency service with left chest pleuritic pain with four days of evolution and weight loss in recent months. The physical examination showed no significant changes. Analytically with only a slight increase in C-Reactive Protein (CRP). The chest X-ray showed elevation of the left hemi-diaphragm and an opacity in the base of the left lung field. A thoracic computed tomography (CT) revealed a lateral condensation in the left lung base of indeterminate nature, associated atelectasis without air bronchogram; several nodules and micronodules in both lung fields, the largest in the apical segment of the left lower lobe (11 mm), the periphery of the posterior segment of the right lower lobe (15mm) and the internal segment of the right lower lobe (12mm). The patient was hospitalized on suspicion of neoplastic disease. Given the increase in CRP antibiotic therapy was introduced. A bronchoscopy was performed showing no significant changes. The cytological and microbiological examination of bronchoalveolar lavage (BAL) were negative. The serological study was also negative. By inaccessibility of the lung lesions it was not possible to perform transthoracic needle aspiration. As a complementary study conducted a new thoraco-abdominal-pelvic CT a week after admission, which showed size variation of the nodes comparing to the previous examination, decreasing size of the nodules and central cavitation of one of them, more suggestive of an infectious process. In the abdomen there was a millimetre nodular hypodensity located in liver and a nodular heterogeneity of the head of the pancreas with poorly defined limits. To exclude septic embolization a transthoracic echocardiogram was performed but no significant

changes were found, particularly thrombi. In face of the dimensional changes of the pulmonary nodules remained the therapeutic measures were maintained, and the patient discharged with indication to complete the antibiotic treatment cycle for 30 days, assuming the diagnosis of cavitated multifocal pneumonia. CT control one month after discharge with full resolution of all lung nodules.
Discussion: This case highlights the importance of clinical research when in the presence of multiple pulmonary nodules. If, on one hand, there is the need for cancer exclusion, especially in patients at high risk, we cannot exclude other differential diagnoses that mimic or follow this framework. Faced with scattered nodules bilaterally without specific features of malignancy and the presence of other signs consistent with infectious process, an antibiotic cycle followed by imaging control can expose an indolent process and avoid more invasive approaches in the search for a definitive diagnosis.

***Key words**: Pulmonary nodules. Differential diagnosis. Infection.*

PRCC-030. THE ART OF BREATHING IN GOOD ALL THE TIME

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The quality of the breathable air and land planning issues agri-environmental policies decisively influence people's health! In the field of morbidity diseases of the respiratory disorders occupy a prominent place in the epidemiological indicators, including incidence and prevalence. The emphysema asthma and COPD and community Pneumonia concern us greatly. Quality of life is greatly affected by this set of conditions. By their nature they are chronic and tend to co-exist with other conditions. The dependence of clinical services is huge. The Alentejo produced 7% of the oil in ten years and went on to produce two-thirds of the oil. Through spatial planning policies, has changed much the breathing air quality in the Lower Alentejo the intensive exploitation of olive groves (usually has forty olive trees per hectare, this scheme has three hundred) whose pollination is anemophilous, to add to the traditional grass, take the highly allergenic environment for people vulnerable to these allergens, but thought possible prevent and enable the ventilatory condition despite this level of environmental pollution. The emergence of new cases to add to existing in pulmonary disease, acute exacerbations and infections associated with its co morbidities negatively influence the quality of life of the Lower Alentejo. Avoidable admissions and adequate control of conventional face therapeutic to some variables that contribute to the onset of pneumonia in the community, such as age, gender corticosteroid therapy overweight and immobility imposed and the need for effective breathing and a set of good advice, including anti pneumococcal vaccination, oral hygiene smoking cessation, increased exercise tolerance in particular. Based in customized intervention, aimed at healthy individuals and patients with respiratory condition in children of pre-school, in the school and adults to attend the physical mobility programs promoted by the Sports Office of the Municipality. Physical Education teachers were involved in the foundation and activities of this project. We believe that the promotion of a diaphragmatic breathing can be inculcated in order to automate the individuals, even in the male gender, since women have an abdominal breathing pattern. If we improve the diagrammatic tour are influencing the respiratory dynamics, benefiting fisiometabólicos processes. You will also impart skills in bronchial hygiene and respiratory etiquette. We also believe that we do not know all the benefits of an inspiration by the nose and an exhalation through the mouth, but we know that if we apply the Castro Tenth individuals with respiratory disease benefit from a better quality of life for the prevention of bronchospasm, optimization of capacity vital, improvement in lung compliance, enriching the arterial blood. What are the Tenth Castro? There are ten breaths. An active abdominal inspiration in one second by nose, followed by expiration active pursed-lip using the abdomen of the transverse lasting between five and ten seconds. This exercise is done at night in bed and wake up, still in bed.

***Key words**: Breath. Diafragmatic breath. Breathable air.*

PRCC-031. FUNCTIONAL DIAGNOSIS OF CYSTIC FIBROSIS

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Introduction: Cystic fibrosis (CF) is a genetic disease caused by mutations in the CFTR gene. CF classic phenotype is the presence of bronchiectasis, chronic sinus disease and pancreatic insufficiency. In the presence of a consistent phenotype, the diagnostic criteria are based on evidence of dysfunction of the CFTR protein (sweat test) or presence of two CFTR gene mutations. In cases of difficult diagnosis, one can resort to other functional studies of CFTR. We present the case of a patient with no clinical phenotype of CF and two CFTR mutations identified.
Case report: Woman 67 years old, retired hairdresser, smoker (30 pack year). Since the age of 56 in pulmonology consultation after hospital admission for exacerbation of chronic bronchitis. Treated with ipratropium bromide, with symptomatic control without exacerbations requiring antibiotics. Stable lung function (FEV_1/FVC-79%; FEV_1pred.-104%, DLCO decreased but normalized when corrected for alveolar volume), with persistently negative sputum microbiological isolations and normal computational CT scans. The assay of alpha 1 antitrypsin was normal. Mutation screening was performed as a part of a research project in order to analyse the frequency of CFTR mutations in chronic respiratory patients (methodology applied in DNA samples, RNA: ARMS - amplification refractory mutation system). Two CFTR mutations (G542X/G576A) were revealed. Sweat test was negative and the determination of fecal elastase was used to exclude pancreatic insufficiency. Functional study of CFTR was performed by rectal biopsy (Ussing chamber methodology and intestinal organoid) which excluded major dysfunction of ion transport. CF diagnostic was excluded and genetic screening to relatives was offered.
Discussion: Despite the two CFTR mutations identified, the expressed phenotype was not suggestive of CF. This fact was corroborated by the functional tests of the CFTR protein, that lead to the exclusion of this diagnosis. Although about 2,000 CFTR mutations have been identified, most do not present a known pathogenic role. Despite the deleterious role of the mediterranean mutation G542X, the G576A mutation was initially considered neutral but later associated with classic CF and CF-related diseases. This mutation induces changes in the splicing process, leading to the production of both normal and aberrant RNA transcripts, with variable levels between different patients. Nowadays, it is estimated that if only 10% of the ionic transport is maintained, there will be no manifestation of CF. Moreover, the phenotypic heterogeneity of CF disease, the modifying genes and environmental factors, result in expressions, making the interpretation of a genotype challenging. In the case of this patient, it is arguable whether its CFTR genotype may have boosted the pathological effects of tobacco, resulting in an increased susceptibility to the development of respiratory symptoms. The analysis of the CFTR gene can lead to the identification of mutations with unknown significance. This limitation must be apprehended by the doctor in

a therapeutic and psychological context due to the strong consequences resulting from a CF diagnosis. In selected cases the functional study of the CFTR protein can be a great help.

Key words: *Cystic fibrosis. CFTR. G542X. G576A.*

PRCC 032. QUALITY OF LIFE ASSESSMENT IN CYSTIC FIBROSIS

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Introduction: In recent decades there has been an increase of life expectancy in patients with cystic fibrosis (CF). However, these patients remain very symptomatic and submitted to complex drug regimens that cause a decrease in quality of life (QoL).
Objectives: Assess QoL in patients with CF by generic and specific instruments, and, if possible, establish a correlation between the two. Establish a relationship between QoL and prognosis (measured by functional impairment).
Methods: Prospective cross-sectional study of patients with CF followed in the specialized center from CHLN. In order to evaluate the QoL of patients the EQ5D test (generic instrument) and the CFQR14+ (specific instrument) were applied during a stable clinical phase. Demographic and clinical data were obtained by reviewing the patients' medical records. 3 groups were created in accordance with the functional impairment: mild FEV1 > 60%; moderate FEV1 30-60% and severe FEV1 < 30%.
Results: 33 patients (55% women) with median age 28 years [19; 55] and BMI of 21.49 kg/m^2 [15, 35] were included. The mean FEV_1pred. was 62.82% ± 20.62. Functional impairment of degrees (% of patients): severe 6.1%, moderate 39.4% and 54.5% mild. Results from EQ5D (mean ± SD): EQVAS- 77.12 ± 10.62 and EQ5Dindex- 0.864 ±.20, where a higher number of patients reported the anxiety/depression domain. There was no correlation with age, BMI and gender. Patients with severe, moderate and mild functional impairment had progressively higher EQ5Dindex values (but p > 0.05). Patients with chronic bronchial infection by *Pseudomonas aeruginosa* showed lower EQ5Dindex. In CFQR14 + the domains with lower scores were treatment and physical activity, in contrast to the areas food and digestive symptoms that obtained the highest values. In the body weight and image domains, women had higher scores. FEV_1pred. was only significantly associated with body image domain. Finally EQVAS seems to correlate with six domains CFQR14+ (physical activity, emotions, vitality, health perception, daily activities and respiratory symptoms). Conversely, EQ5Dindex not correlate with any domain CFQR14+.
Conclusions: The assessment of QoL by EQ5D in the sample group revealed that patients maintain a reasonable QoL. However, and accordingly to some references, no significative correlation was found between EQ5Dindex and FEV_1, showing the short-sightness of generic methods in predicting FEV_1. The application of a specific, more discriminative questionnaire showed that physical activity and treatment domains had the lowest scores. These domains seem to be the most important areas for patients. In this study, no statistical significance between FEV_1 and the majority of CFQR14+ domains was established, perhaps because of the small sample size. Also the correlation between the two instruments was not consistent. In the future, equivalences between specific and generic instruments should be considered, being the latter useful tools to support health policies. This study proves the consistency of the published evidence on the importance of assessment of QoL in CF, even for guidance of therapeutic interventions. Prospective longitudinal studies are needed with larger samples to confirm these results.

Key words: *Cystic fibrosis. Quality of life. EQ 5D. CFQ R14+.*

PRCC-033. THE LONG ABSENCE...

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Introduction: Unilateral pulmonary artery agenesis (UPAA) is a rare congenital anomaly due to a malformation during embryogenesis of the sixth aortic arch. It was first diagnosed in 1868 and since then, only 420 cases have been reported worldwide. It can occur as a single disorder or may be associated with other congenital malformations. The estimated prevalence of single UPAA is 1 in 200,000 young adults and there is no sex predilection. The diagnosis is usually set at adolescence, however it can occur as a late diagnosis in adulthood. In two thirds of cases described, the right pulmonary artery is the one affected. We report the challenging case of a female adult patient with right pulmonary artery agenesis who was primarily assumed as a right pulmonary embolism.
Case report: Female patient, 49 years old, non-smoker, previously diagnosed with pulmonary tuberculosis with sequelar bronchiectasis, right inferior lobe pneumonia, bronchial asthma and ischemic heart disease. Due to complaints of fatigue and dyspnoea with progression to small efforts, the patient was submitted to complementary study. Pulmonary function test showed small-airway obstruction and diminished carbon monoxide diffusing capacity. Arterial blood gases revealed hypocapnia without partial respiratory failure. Polysomnogram reflected mild obstructive sleep apnea syndrome. Chest computerized tomography (CT) highlighted honeycomb areas with ground glass patches on the right lung, with loss of volume on the right chest area and compensatory hyperinsuflation of the left lung. Ventilation-perfusion scintigraphy showed diffuse hypoperfusion of the right lung area suggestive of pulmonary emboly (PE) of the right pulmonary artery. Chest angio-CT concluded the absence of the right branch of the pulmonary artery, without evidence of PE. Chest echocardiography excluded relevant malformations and pulmonary hypertension (PHT). After multidisciplinary reunion with was assumed as the main diagnosis probable UPAA, with the patient being referenced for consultation with Cardiothoracic Surgery for treatment orientation.
Discussion: UPAA is very rare and both clinicians and radiologists should be well aware of the possibility of undiagnosed UPAA cases in adults, with many atypical characteristics. The combination of chest pain, pleural effusion, and recurrent infections may be present in 37% of patients, while dyspnoea or exercise intolerance in 40% of patients. Massive, life-threatening haemoptysis could also occur. PHT is observed in almost 45% of patients and determines long-term survival. Diagnosing UAPA can be difficult, but the diagnosis can be definitively made by CT, magnetic resonance imaging or transthoracic echocardiogram. Treatment options comprises of surgical, pharmacological and behavioural management. Pneumonectomy and surgical revascularization are considered in cases of recurrent haemoptysis, pulmonary infections and PH. Selective embolization of bronchial or non-bronchial systemic arteries is a valid alternative for patients with massive haemoptysis not eligible for surgery. Pharmacological treatment for PH is strongly recommended for patients unable to undergo surgical revascularization or in cases with no improve after surgery.

Key words: *Unilateral agenesia. Pulmonary artery. Pulmonary embolism.*

PRCC-034. RESPIRATORY PAPILLOMATOSIS. TWO CASE REPORTS

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Introduction: Recurrent respiratory papillomatosis (RRP) usually occurs in infancy, being the most common cause of benign tumor of the larynx in children. The adult form typically occurs between the 2nd and 4th decade, the risk factors being multiple sexual partners and high frequency of oral sex. The chronic nature of the disease, with multiple recurrences, can lead to the spread from the larynx to the trachea and bronchi. Although benign, airway obstruction requires multiple invasive procedures. The papillomatous lesions may have a more aggressive growth and in rare cases malignant transformation occurs. Serotypes 16 and 18 are considered to be oncogenic. The authors present two clinical cases of respiratory papillomatosis.
Case reports: Case 1: female gender, 59 years old, non-smoker, personal history of recurrent laryngotracheal papillomatosis - HPV 6 (tracheostomy for 18 years), accompanied in our department since 2008. A total of 4 rigid bronchoscopy were performed and complete patency of the airway was achieved in all situations after Argon-Plasma Coagulation and debulking. Biopsies were performed in all exams and malignant transformation was excluded. Case 2: male gender, 73 years old, non-smoker, personal history of dyslipidaemia and benign prostatic hypertrophy. Presents with cough and hemoptysis in the last month. Chest CT - exophytic thickening of the endotracheal wall causing decreased airway caliber in a 57 mm extension. Fiberoptic Bronchoscopy - multiple masses in the lower 2/3 of the tracheal wall prolapsing into the lumen, with higher expression in the inferior 1/3; 80% stenosis of the LMB. Rigid Bronchoscopy - argon-plasma coagulation and debulking was performed with excellent endoscopic result. Histopathology - stratified epithelium squamous papilloma with high-grade dysplasia and a corresponding focus of squamous cell cancer. Viral DNA Sequencing - HPV 18. The patient is currently undergoing radiotherapy treatment.
Discussion: Bronchology plays a key role in addressing these patients: bronchoscopy is the preferred method for the observation of lesions and tissue sampling; current treatment of choice for respiratory papillomatosis is laser ablation or argon-plasma coagulation. Airway obstruction and malignant transformation are the main complications.

***Key words**: Human papillomavirus. Recurrent respiratory papillomatosis. Bronchology.*

PRCC-035. PULMONARY HISTIOCYTOSIS OF LANGERHANS CELLS: DIAGNOSTIC CHALLENGE IN TWO CLINICAL CASES

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Introduction: Pulmonary histiocytosis of Langerhans cells (PHLC), also called histiocytosis X, is a rare interstitial lung disease that primarily affects young adults. It is characterized by abnormal proliferation of Langerhans cells, with the formation of nodular lesions of variable evolution. Its prevalence and actual incidence is unknown. The tobacco consumption is so far the only established association with the disease, since it occurs almost exclusively in smokers. We describe 2 clinical cases with similar initial clinical presentation but with particularities of diagnostic orientation and distinct therapy.
Case reports: Female, 35 years of age, non-smoker, diagnosed at 19 years old with pulmonary histiocytosis of Langerhans cells and, since then, followed by pneumology. The diagnosis was made in the context of Pneumology hospitalization due to subacute state of apparent respiratory infection. From the additional diagnostic tests, we emphasize bilateral inflammatory changes, ground glass pattern, and non cavitary nodules of bizarre morphology in CT scan of high resolution (HRCT), lymphocytic alveolitis in bronchoalveolar lavage (BAL) with citobacteriological and mycobacteriology studies negative, therefore lung biopsy by thoracoscopy was done. Histological examination revealed cells with immunoposivity for S100 protein, consistent with the diagnosis of pulmonary Langerhans cell histiocytosis. More or less regular systemic corticosteroid therapy was applied over the years, however due to the worsening of the clinic, imaging and lung function we prescribed methotrexate in 2014 with subsequent achievement of clinical stability. Male of 33 years of age, smoker (8 unit pack year) was admitted in January 2014 with subacute dry cough and dyspnea in moderate exertion. The diagnosis of pulmonary pneumocystosis was made in an immunocompetent patient. He took for 3 weeks cotrimoxazol with good clinical evolution with negative parasitological examination of the LBA control. Initial imaging changes of HRCT showed scattered ground glass pattern that improved in the control CT of December 2015. It was also observed cavitary nodules with some cystic formations that worsened on this exam and is very suggestive of PHLC, to be confirmed by BAL cytology and histology.
Discussion: The clinical presentation of PHLC is varied, with 25% of asymptomatic patients. In symptomatic patients dry cough and exertional dyspnea are the most common symptoms. Disease severity is not influenced by tobacco load. The most widely used therapy are the corticosteroids. In patients with refractory response to corticosteroids or multiorganic disease, other drugs are indicated such as methotrexate, vinblastine, cyclophosphamide, etoposide or cladribine. With regard to diagnosis of this pathology, HRCT is the gold standard test that reveals nodular and cystic characteristic lesions. In the first case, it is described a symptomatic patient in need of immunosuppressive therapy without characteristic lesions in CT, which is why it was of high importance to have histological confirmation of immunopositivity for S100 protein. In the second case we describe an immunocompetent patient with a pneumocystosis. There are few cases described in the literature of this opportunistic infection in immunocompetent patients. Despite not having been histologically confirmed, the imaging changes are typical of PHLC. This patient is currently asymptomatic, with no indication for immunosuppressive therapy, and is under clinical, imagiological and functional surveillance.

***Key words**: Pulmonary histiocytosis of Langerhans cells. Interstitial lung disease. Langerhans cells.*

PRCC-036. SWYER-JAMES-MACLEOD SYNDROME: A RARE ENTITY

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Introduction: Swyer-James-MacLeod syndrome (SJMS) is a rare entity, characterized by unilateral or lobar pulmonary hyperlucency, secondary to a post-infectious obliterating bronchiolitis process that occurs in childhood. It is characterized by hypoplasia and\or agenesis of the pulmonary arteries and alveolar overdistention. The clinical presentation can be varied. The affected children may be asymptomatic, but more often present recurrent respiratory infections and develop bronchiectasis. Despite being rarer the diagnosis might be done in adult life.
Objectives: Clinical and imaging characterization of patients followed in pulmonology consultation in Centro Hospitalar São João diagnosed with SJMS during adulthood.
Methods: Medical records of patients with SJMS were reviewed and analyzed the demographic and clinical data, namely the beginning and presentation of respiratory symptoms. Data information from respiratory function and chest computed tomography were also collected. The data analysis was performed using SPSS version 24.
Results: Thirteen patients were identified with SJMS diagnosed in adulthood, 69.2% were male, with mean age of diagnosis of 42.2 years (min. 18, max. 70). The patients were referred to pulmonology consultation for recurrent respiratory infections in 38.5%, dyspnea

in 30.8%, asthma in 7.7% and chronic obstructive pulmonary disease in 7.7%. 15.4% were referred by findings on chest radiograph. All the patients underwent thoracic computed tomography (CT) during etiological investigation. 53.8% of the patients presented radiological alterations on the right side and 41.7% on the left side. Bronchiectasis were present in all the patients. Functionally all patients presented obstructive ventilatory disease, 46.2% of which severe obstruction. More than 90% of patients presented a clinic with several years of evolution, most since childhood (53.8%). The most common symptoms were dyspnea and recurrent respiratory infection. 85% of the patients had a history of some severe respiratory infection in childhood.
Conclusions: Although less common the diagnosis of SJMS can be done in adulthood. We emphasize the presence of respiratory symptoms with prolonged evolution, the presence of obstructive ventilatory syndrome and the high prevalence of recurrent respiratory infections with the presence of bronchiectasis on CT, which can have an impact on therapeutic approaches.

Key words: *Swyer-James-Macleod syndrome. Unilateral hyperlucency.*

PRCC-037. SWYER-JAMES-MACLEOD SYNDROME: A RARE CAUSE OF PULMONARY HYPERLUCENCY

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Introduction: Swyer-James-MacLeod Syndrome is a rare disease resulting from a post-infectious bronchiolitis obliterans at early age. The pathogenesis of the disease is not fully understood, but seems to focus on an inflammatory response at distal airways, which involves fibrosis and destruction of alveolar-capillary units, resulting in hypoplasia of pulmonary vasculature from the affected side. Imaging hallmarks include one or more areas of unilateral lung hyperlucency, associated with mosaic attenuation by air trapping and hypovascularization. Despite its early onset, the diagnosis may be delayed, particularly in midly symptomatic patients and when there is a previous diagnosis of chronic lung disease from other causes. The authors present a case of Swyer-James-MacLeod Syndrome diagnosed in the 7th decade of life, following a hospitalization for tracheobronchitis and type 2 respiratory failure (RF).
Case report: A 63 year-old non-smoking female, working in plastics industry, had history of mite atopy and was not on chronic medication. Despite ignoring childhood diseases, she reported tiredness and episodes of coughing and wheezing from adolescence. In January 2015 she presented in emergency department with cough, expectoration and asthenia for two weeks. On admission she was normotensive, subfebrile, polypneic and presented with type 2 RF in arterial blood gas (ABG) (pH 7.44, paCO2: 61 mm Hg, paO2: 62 mmHg, $HCO3^-$: 41.4 mmol/L); auscultation revealed decreased breath sounds at right side plus bilateral wheezing, and serum inflammatory markers were elevated. Chest radiograph showed a slight tracheal deviation towards the right side, a diminished right hilar shadow and also a loss of right lung volume with a more hyperlucent area in the middle third of the right lung. The patient was diagnosed with asthma exacerbation and treated with amoxicilin and clavulanic acid with good response. In spite of that, oxygen therapy was required at discharge. Respiratory function tests subsequently performed revealed a severe airflow obstruction with hyperinflation, negative bronchodilator test, significant desaturation during the six-minute walk test and hypoxemia and hypercapnia in ABG (pH: 7:42; pCO2: 52 mmHg, pO2: 61 mmHg). The CT scan showed a decreased diameter of the right pulmonary artery and an area of hypoattenuation in right lower lobe (RLL) associated with tubular bronchiectasis, a pattern suggestive of bronchiolitis obliterans. A pulmonary CT angiography was performed, allowing to state a significant reduction in the caliber of the right pulmonary artery and lower lobar branches without pulmonary thromboembolism (PE). There was also a hypodensity in the RLL, suggesting a bronchiolitis obliterans and also explaining the vascular asymmetry. The ventilation-perfusion scintigraphy showed a matched ventilation-perfusion defect in the RLL, and the ventilation scan was more heterogeneous than the perfusion scan. The findings allowed the diagnosis of Swyer-James-MacLeod Syndrome. The patient maintained surveillance and treatment with inhaled corticosteroids and double bronchodilation.
Discussion: Although rare, Swyer-James-MacLeod syndrome should be considered in the differential diagnosis of unilateral hyperlucent lung, along with common diseases as asthma and PE, and rarer ones like cystic fibrosis, congenital lobar emphysema or vascular malformations. Treatment includes cinesitherapy, bronchodilators and inhaled corticosteroids. Surgical resection is reserved for infections refractory to medical therapy.

Key words: *Swyer-James-MacLeod. Hyperlucency. Bronchiolitis. Mosaic.*

PRCC-038. WHEN SERIOUS INFECTIONS MULTIPLY! ABOUT A COMMON VARIABLE IMMUNODEFICIENCY CASE

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Case report: The authors present a case study of a 37 years old woman, Caucasian, followed in the Primary Immunodeficiency's (PID) Consultation, diagnosed with Common Variable Immunodeficiency (CVID) since the age of 16 years, after an hospital admission for bilateral pneumonia associated with empyema and anemia, concomitant with thrombocytopenia and lymphopenia. Since the diagnosis the patient has made replacement therapy with Polyclonal IgG intravenous at high doses. Along the years has been attended by several clinical situations, namely, multiple respiratory infections, bronchiectasis and chronic inflammatory infiltration. Associated with these clinical conditions she develops hepatomegaly with granulomatous hepatitis with nodular regenerative hyperplasia, and later homogeneous splenomegaly, and recurrent diarrhea with deficit of absorption, osteoporosis, hemolytic anemia and autoimmune thrombocytopenia. The severity of inflammatory/autoimmune complications have justified immunosuppressive therapy for long periods with corticosteroids and cyclosporine. Since 30 years the patient develops, among other problems like severe respiratory infections with uncommon agents of PID humoral predominance, *Mycobacterium gordonae* infection at the age of 30 years treated with antituberculosis therapy for 9 months; *Pseudomonas aeuruginosa* infection at 31 years controlled with inhaled *tobramycin* for 4 months and *colistina* during 12 months; *Aspergillus flavus infection* at 34 years remaining medicated prophylactically with *voriconazole* therapy. At the age of 37, in the year 2016 the respiratory functional study reveals loss of lung function when compared to the previous year: FEV1/FVC 66.1; FEV1 40%; TLC 96%, RV 169% Raw 190%, DLCO 31.1%, DLCO/VA 47% without answer to bronchodilation. In April 2016 comes to the Primary Immunodeficiency Consultation due to cough with purulent abundant expectoration, fatigue, and dyspnea for minimal efforts and fever. The thoracic computed tomography (CT) reveals areas of bronchiectasis with fibrosis in the right upper lobe (RUL), middle lobe and in both lower lobes cavitation areas that communicate with multiple bronchiectasis with the RUL, and on the left lung base and in the right lung base. The cultural examination of sputum revealed *mycobacterium avium*. Begin therapy with *amikacin* (first intramuscular injection and later inhaled), *clarithromycin* and

rifabutin. The bronchoscopy examination revealed *Aspergillus fumigatus* in bronchial secretions by culture. The *galactomannan* was positive in different determinations. After interdisciplinary case discussion it was decided to establish *posoconazol*. Two months later, with antituberculosis therapy and antifungal therapy the patient shows significant clinical improvements, reduction of *galactomannan* and negative culture tests.
Discussion: In conclusion the case study presented is an example of a Common Variable Immunodeficiency, which is the most common symptomatic PID in adults. This patient throughout their evolution presents multiple problems: infectious, preferably reaching the lung, and simultaneously non-infectious, requiring immunomodulatory therapy. This therapy may contribute to an additional susceptibility to infections usually less frequent in these patients. The conjunction with concomitant infections by different agents makes extremely difficult therapeutic options. This case is illustrative of the complexity of some of these patients may present, and the extreme importance of following a referral center and interdisciplinary collaboration, particularly with the Pulmonology.

Key words: *Primary immunodeficiency. Aspergillosis. Non-tuberculous mycobacteria. Lung destruction.*

PRCC-039. NODULAR IDIOPATHIC PLEUROPARENCHYMAL FIBROELASTOSIS MAY BE A PITFALL IN PULMONARY BIOPSY. CASE REPORT

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Introduction: Idiopathic pleuroparenchymal fibroelastosis (IPPFE) is an infrequent and recent well defined disease characterized by fibrosis and thickening of subpleural lung parenchyma, mainly in the upper lobes. Clinical and radiological presentation may mimic other idiopathic interstitial pneumonias (IIP) and pathological evaluation has a core role in providing the correct diagnosis.
Methods: We present a case of an 82-years-old-male with complains of right thoracic pain and haemoptysis. High resolution computerized tomography detected a solitary pulmonary lesion in the upper right lobe, with 18-Fludeoxyglucose (^{18}F-FDG) uptake. Transthoracic needle biopsy revealed possible adenosquamous carcinoma with solid and acinar pattern in fibro-elastic stroma with supporting immunohistochemistry. Lobectomy was performed.
Results: Histological examination of the resected specimen revealed nodular subpleural thickening with collagenous fibrosis, pleural architectural distortion with elastosis and intra alveolar fibrosis with curled, short and randomly oriented elastic fibers. There was an abrupt transition to the remaining lung where emphysema, constrictive bronchiolitis and organizing pneumonia were registered. Grocott methanamine silver stain and PAS did not show fungal infection. The diagnosis of pleuroparenchymal fibroelastosis was established with nodular sub-pleural presentation. The patient was revaluated three weeks after surgery. A chest X-ray showed minimal pneumothorax that disappeared in two weeks, without need for chest drainage.
Conclusions: IPPFE is a distinct and well characterized entity, classified as a rare IIP, which runs an aggressive and rapidly progressive course with poor prognosis. The intense fibroelastosis may be associated with prominent inflammatory and reactive changes, making the differential diagnosis with other IIPs challenging and occasionally mimicking oncologic diseases, especially in small biopsies. The combination of clinical presentation, radiological patterns and histological characteristics is the key for diagnosis. The early recognition of this disease will provide more accurate diagnosis and proper management by expert multidisciplinary teams.

Key words: *Pleuroparenchymal fibroelastosis. Fibrosis. Elastosis.*

PRCC-040. BRONCHIAL SCHWANNOMA AS CAUSE OF OBSTRUCTIVE PNEUMONIA

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Introduction: Bronchial schwannomas are very rare pulmonary lesions, with few cases reported in literature. Clinical and radiological characteristics are nonspecific and pathological examination usually provides the definite diagnosis. For small lesions endoscopic approach may be sufficient, but larger lesions associated with organizing pneumonia, surgical intervention may be required. Prognosis is typically favourable.
Methods: We describe a case of a 66-years-old female with complaints of productive cough and sporadic haemoptysis, dyspnoea, anorexia, excessive sweating and weight loss within 2 months of evolution. Occupational history contamination was excluded and allergic patterns were denied. Pathological background included hiatus hernia, glaucoma, arterial hypertension, vertigo and atrial fibrillation. Regular medication was lansoprazole, idabenone, propafenone, diazepam, betahistine hydrochloride, triflusal and latanoprost. Physical evaluation showed absence of skin lesions or colour abnormalities and lower limb oedema. Arterial blood pressure was 130/70 mmHg and O_2 saturation of 97%. Cardiac auscultation revealed arrhythmic beatings without murmurs, with 80 bpm. Pulmonary auscultation discovered diminished lung sounds on the left apex. Blood tests did not demonstrate relevant findings. Computed tomography (CT) showed a soft tissue dense lesion on the left hilum with 3.75 cm with 18-Fludeoxyglucose (^{18}F-FDG) uptake.
Results: Left upper lobectomy was performed. Gross examination revealed an endobronchial polypoid mass with 3.7cm of diameter, lobulated and tan-brownish in cut section, without necrosis. Histological evaluation revealed an expansive mesenchymal lesion with cellular dense (Antoni A) and less dense (Antoni B) areas with slightly pleomorphic spindle cells, but without mitotic activity. Verocay bodies were evident as well as cystic areas, some hyaline walled blood vessels and iron pigment (evidenced by Prussian blue stain) retention. Surgical margins were negative. The remaining lung showed organizing pneumonia secondary to bronchial obstruction. Immunohistochemistry studies showed diffuse nuclear and cytoplasmatic positivity for S100 protein; Vimentin, Synaptophysin and neuron specific enolase (NSE) were also expressed and AE1/AE3, Chromogranin A, CD117, Melanosome, CK7 and TTF1 were negative. The proliferative index evaluation with Ki67 was also studied, with a low result (2%). The diagnosis of bronchial schwannoma was definite. The patient was evaluated three weeks after surgery. No clinical complains were registered and there were no signs of tumour relapse.
Conclusions: Bronchial schwannomas are rare, but awareness of the entity is important in order to reach a correct diagnosis and decide proper bronchoscopic/surgical intervention, based on patient clinical picture.

Key words: *Bronchial schwannoma. Obstructive pneumonia.*

PRCC-041. PULMONARY AMYLOIDOSIS. CASE REPORT

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Introduction: Amyloidosis is a disease of unknown etiology, characterized by extracellular deposition of amyloid material in any organ or tissue. It occurs usually in male patients aged between 55 and 60 years-old. The respiratory system achievement is not

frequent, and has even less presentation in the intraparenchymal nodular form. It is, therefore, one of lung cancer differential diagnosis.
Case report: The authors present the case of a patient, male, 71 years old, Caucasian, retired from construction, with known personal history of hypertension, benign prostatic hyperplasia, skin basal cell carcinoma. The patient was also followed in urological medical appointment due to a mesenteric inflammatory process non- clarified. No smoking habits or pulmonary pathology known. The patient is sent to the pulmonology appointment to search for micro pulmonary nodules, complaining at that time only of bilateral low back pain, with no relevant findings on the physical examination. He denied respiratory symptoms and/or systemic. Analytically stood out sedimentation speed of 120 mm/h and beta 2 microglobulin 4.3. Bronchoscopy exam did not reveal endobronchial lesions, however, chest and abdominal CT revealed important adenopatic involvement of all the ganglion chains of the mediastinum, with larger nodes at subcarinal space and azygos-esophageal recess. It also co-existed adenopatic involvement in both pulmonary hilum. CT also revealed areas of increased lung parenchymal density in the slope within the right lung field. In the upper abdomen protrudes marked densification of fat surrounding the root of the mesentery and retro peritoneal level involving the structures of the renal hilum. In this context, it was assumed the existence of lymphoproliferative disease. The patient medulograma and bone biopsy were proved inconclusive. At 9.5.2016 he was submitted to cervical mediastinoscopy with lymphadenopathy biopsy of the right paratracheal group, the result of pathological examination revealed extensive deposits of amyloid substance in the interstitium and in the vessel wall. The immuno-histo-chemical suggested it was amyloid AL without lymphoproliferative disease.
Discussion: Pulmonary amyloidosis is a disease that despite being infrequent should always be included in the differential diagnosis for clinical pictures/radiological lung involvement.

Key words: *Amyloidosis. Pulmonary amyloidosis. Lung cancer.*

PRCC-042. SUBGLOTTIC STENOSIS IN ADULTS. THREE CASE REPORTS

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Introduction: Subglotic stenosis is a difficult airway problem with multiple causes which require experience and judgment for satisfactory treatment outcome. It may be classified based on cause: congenital or acquired and location of narrowed segment. The clinical manifestations of tracheal disorders are determined by extend and location of the abnormalities. Dyspnea, inspiratory wheezing and stridor are the main form of presentation when significant obstruction. Beyond the physical examination, evaluation often includes a combination of physiologic, radiographic, and endoscopic assessments. We report 3 cases of different etiologies for subglottic stenosis.
Case reports: Case 1: non smoker 68 years old women with long history of upper airway obstruction since childhood, possible due to congenital subglottic stenosis. She has required 3 tracheostomies in the past and had laser surgery but continues to have significant stenotic segment situated about 1,5 cm below the glottis and about 1,5 cm in length. She underwent a tracheal resection with end-to-end anastomosis with removal of the anterior ring of the cricoid, which was also part of the stenotic segment. Postoperatively, the patient developed progressive stridor. The CT of the neck showed a thin septum obstructing the airway in the region of the cricotracheal anastomosis. The patient was taken to the operating room and the rigid bronchoscope was utilized. The portion of the obstructing septum from 10 o'clock to 5o'clock was removed. She holds regular evaluation and sporadic need of forceful dilatation. Case 2: non smoker 32 years old women with history of tonsillectomy in childhood. She presented dyspnea and wheezing, with no response to bronchodilators. Functional tests - fixed obstruction airflow. CT of the neck-pharyngolaryngeal air column and antero-posterior tracheal diameter of 6 mm on the projection of tongue's base. Autoimmunity and ACE- negative. Flexible bronchoscopy - subglottic stenosis with reduction about 60% of the lumen. She underwent forceful dilatation with bronchoscopes and balloon. Case 3: nom smoker 34 years old men, presents with dyspnea and stridor at rest. Functional tests - fixed obstruction airflow. CT of the neck and thorax - linear band on the subglottic space, from 11 o'clock to 6 o'clock. 11 mm nodule on the periphery of the right upper lobe. Autoimmunity - positive c-ANCA (84.4 UQ). Flexible bronchoscopy - concentric subglottic stenosis associated with a pearly, fibrotic band. He underwent rigid bronchoscopy with removal of membranous septum and forceful dilatation with bronchoscopes and balloon. Suspected diagnosis is granulomatosis with polyangiitis - clinical presentation and positive serology – we are waiting for histology.
Discussion: Tracheal pathology evaluation depends on multiple studies that complement each other. The subglottic stenosis is a complex disease and the decision about the therapy should consider the underlying disease, severity of symptoms and expected objectives. Therapeutic evaluation should rely on an experienced and multidisciplinary team. Bronchoscopy provides a key role in characterization of the lesions, its extension, as well as performing biopsies and therapeutic approach.

Key words: *Subglotic stenosis. Dyspnea. Bronchoscopy.*

PRCC-043. IDIOPATHIC TRACHEAL STENOSIS: A RARITY WITHIN A RARITY

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Introduction: Tracheal stenosis is an uncommon disease, potentially fatal. Etiology may be malignant or benign. Idiopathic progressive subglottic stenosis is a benign cause, and rare, of central airway obstruction, with marked predominance of women, predominantly in perimenopausal period. Symptoms of obstruction are nonspecific and may be acute or subacute.
Case report: A 69-year-old woman, Caucasian, secretary, non-smoking, without relevant personal history or allergies. In 2013 started symptoms of dyspnea and wheezing. At the time, was assumed a diagnosis of asthma, having been medicated with oral corticotherapy, with clinical improvement. In January 2015 presents progressive clinical worsening, characterized by dyspnea, wheezing and sporadic dry cough, with improvement in decubitus. Physical examination showed no relevant changes. Laboratory tests revealed: alpha 1-antitrypsin 146 mg/dL; IgE 12.8 UI/mL; IgA, IgG e IgM unchanged; negative Phadiatop and RAST. Simple espirometry was performed, that showed no changes. Lung function study was repeated and revealed obstruction of the upper airways with mild inflation. Chest TC was performed and showed: "Mediastinum centered; proximal tracheobronchial permeability; without lymphadenopathy displaying; discrete linear densifications in the middle lobe and língula, probably fibro-sequelae; subpleural calcified micronodule in the left upper lobe, probably residual (granuloma); millimeter enphysematous bleb in the middle lobe". Cervical CT was requested and showed: Thickened trachea with filling of anterior commissure extending to a distance of 2.5 cm of infraglottic region. Bronchofibroscopy (May 2015) revealed a ring covered by mucosa without changes or infiltration at 1 cm of

glottis, conditioning reduction of trachea caliber, without other distal changes of the tracheobronchial tree. Biopsies of mucous ring were performed, presenting no significant changes in pathological, microbiological and mycobacteriological exams. After 1 month, patient was attended in a Bronchology Intervention Centre and was submitted to a new flexible bronchoscopy, which revealed fibrotic retraction in the anterior wall of the subglottic region, followed distally by spiral stenosis of 1.5 cm length and 8 mm caliber, with elastic and fibrous consistency in the sidewalls. Biopsies were performed with increased of tracheal caliber to 10-12 mm. Furthermore, instillation of intramucosal Dexamethasone and small cuts on the right wall were performed. In August 2015 was performed rigid bronchoscopy, which demonstrated luminal narrowing by 80%. Cuts were made with rigid plate (2,5 and 8 hours), as mechanical balloon dilation and subsequente Mitomycin-C application. After 1 month was verified a tracheal diameter decrease of 40%, remaining stable until the last bronchoscopy (April 2016). Currently, the patient shows symptoms improvement.
Discussion: In the diagnosis course, the idiopathic tracheal stenosis can be documented by imaging exams, respiratory function tests or bronchoscopy. In the latter, fibrotic or inflammatory signs can be observed. In benign tracheal stenosis, the treatment must be curative, including surgical resection. However, due to the specific nature of this procedure, the therapeutic option often results in intervention bronchology with good results, as in the presented case.

Key words: *Idiopathic Tracheal stenosis. Intervention bronchology.*

PRCC-044. ACUTE STRIDOR: A RARE MANIFESTATION OF A "BIRD'S BEAK" MEGAESOPHAGUS

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Introduction: Stridor is a symptom, not a diagnosis or a disease, and the underlying cause must always be determined. The more common pathologies causing acute stridor are infection, trauma, foreign body, airway burns, neurological injury and tumour growth. Oesophageal achalasia is rarely included in the differential diagnosis of acute stridor. We describe a challenging case of an elderly patient who presented to emergency department with an acute stridor as the initial presentation of oesophageal achalasia.
Case report: An autonomous 87 years-old female patient, without pertinent previous diagnosis, was admitted to emergency room with an acute onset, after feeding, of respiratory distress with stridor and cyanosis. Endotracheal intubation was difficulted by the presence of food waste and nasogastric intubation allowed abundant food aspiration. Laryngoscopy excluded high obstruction caused by foreign body or neoformation and vocal cordes oedema. Bronchoscopy showed food remains present in trachea and bronchial tree. Chest radiograph showed enlargement of the upper mediastinum. Patient was admitted with the hypothesis of aspiration pneumonia. Our first attempt of feeding the patient triggered a new episode of stridor, requiring nasogastric intubation that was not successful. Second bronchoscopy revealed normal mobility of the larynx and patency of airway defense reflex, with an extensive inflammatory involvement of the tracheobronchial tree. Brain computed tomography (CT) displayed no changes. Neck and chest CT ruled out Zenker diverticulum, tumours or cervical collections, showing marked distension of the oesophagus in all its extension. Upper gastrointestinal endoscopy objectified a very dilated oesophagus with abundant food remains. After multidisciplinary discussion, the patient was transferred to Gastroenterology Department with the definitive diagnosis of oesophageal achalasia.
Discussion: Achalasia is a rare disorder that affects approximately one in every 100,000 people. Achalasia is a primary oesophageal motility disorder characterized by the absence of oesophageal peristalsis and impaired relaxation of the lower oesophageal sphincter in response to swallowing. The condition commonly presents between the ages of 30 and 60 years with dysphagia being the primary symptom. Chest pain, regurgitation and weight loss also frequently occur. These symptoms can be insidious, and the condition can exist for years before medical attention is sought. Stridor and respiratory distress in patients with achalasia is thought to be as a result of extreme oesophageal distension causing compression of the trachea against the sternum. There are few published case reports of achalasia presenting initially with respiratory distress and stridor, often after eating and curiously more in the elderly female population. Whilst this is an uncommon presentation, it is life threatening, therefore early management of the compromised airway is critical, along with timely decompression of the mega-oesophagus. In conclusion, the management of this case proves the true importance of a multidisciplinary approach.

Key words: *Oesophageal achalasia. Oesophageal sphincter. Airway obstruction. Stridor.*

PRCC-045. COULD IT BE AN EXPECTED EVENT?

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Case report: A 71 years old man, with small cell lung cancer, T4N2M0, submitted to chemotherapy, neoplasm radiation and prophylactic cranial radiotherapy, currently under second-line chemotherapy with topotecan because of progression of disease. With cardiovascular risk factors, hypertensive heart disease, atrial flutter, active smoking. The patient developed pancytopenia secondary to chemotherapy and is admitted to our hospital with pneumonia, right pleural effusion, fluid overload due to heart failure and tachyarrhythmia. During hospitalization, although optimal medical treatment for heart failure, clinical conditions deteriorate and right pleural effusion get bigger. Because of this situation, the patient is submitted to diagnostic and therapeutic thoracentesis, finding out active tuberculosis infection. Although tuberculosis was not the first diagnostic hypothesis contemplate for our patient, this case reflects the importance of invasive investigation because of the significant prevalence of tuberculosis.
Discussion: This case represents an interesting example of connection between tuberculosis and pulmonary neoplasm. On the first line is known that tuberculosis, as an inflammatory disease and because of the TNF alpha production, promotes oxidative stress and development of pulmonary neoplasm, particularly adenocarcinoma. On the second line, neoplasm itself and its treatment, because of local and systemic effects and associated immunosuppression, promotes the spread of tuberculosis, most frequently as reactivation.

Key words: *Pulmonary neoplasm. Tuberculosis.*