R. Saúde-Conde[a,*], S. Cristóvão-Ferreira[a], E. Campoa[b], M.T. Almodovar[c]

[a] *Medical Oncology, Instituto Português de Oncologia de Lisboa Francisco Gentil, E.P.E., Lisbon, Portugal*
[b] *Medical Oncology, Centro Hospitalar Universitário do Algarve, Faro, Portugal*
[c] *Pulmonology, Instituto Português de Oncologia de Lisboa Francisco Gentil, E.P.E., Lisbon, Portugal*

* Corresponding author.
E-mail address: rconde@ipolisboa.min-saude.pt (R. Saúde-Conde).

13 April 2020

https://doi.org/10.1016/j.pulmoe.2020.11.004

Secondary organizing pneumonia after Varicella-Zoster virus infection: a rare association

Dear Editor,

Organizing pneumonia (OP) is a histologic pattern of the lungs' response to a wide variety of insults, including infectious, non-infectious, or no apparent reason (cryptogenic).[1,2] It is usually a great mimicker showing a wide variety of signs, symptoms, and high-resolution computed tomography (HRCT) findings, which makes it a frequent differential diagnosis. The usual distribution in the HRCT is patchy, peribronchiolar with the presence of numerous buds of granulation tissue within alveoli, often involving alveolar ducts and small airways; areas of consolidation are also the characteristics of organizing pneumonia.[2] Anatomopathological study is needed for final diagnosis.[1,3,4] Although duration and initial symptoms depend on the underlying aetiology, OP usually presents with a several-month history of non-productive cough, low-grade fever, malaise, and shortness of breath. It is mostly seen in patients with pulmonary infection, drug reactions, transplantation, collagen vascular disease, granulomatosis with polyangiitis, after toxic-fume inhalation, e-cigarettes usage and, rarely, lung cancer of unknown primary site.[1,5,6] When the underlying cause is not found, it is idiopathic and then called cryptogenic. Among the pulmonary infections, OP could be present after a bacterial, fungal, mycobacterial or even viral infection. Response to therapy is dependent on the treatment of the underlying cause but, usually, there is a good response to corticosteroid therapy with a good prognosis.[3,4]

We report a case of a 35-year-old male, smoker of 15 packs per year, who went to the emergency department with pruriginous cutaneous eruption. Physical examination revealed vesicular cutaneous lesions, and on pulmonary auscultation a reduction of vesicular murmur and crepitations on the left hemithorax. Analysis identified respiratory failure, thrombocytopenia, hepatic dysfunction, and elevation of inflammatory parameters (C-Reactive Protein = 9.73 mg/dL (Normal < 0.5 mg/dL) with normal procalcitonin). The chest radiograph revealed bilateral opacities with air bronchogram (Fig. 1A). After the identification of epidemiological context for varicella-zoster infection (son with chickenpox), patient was admitted to hospital stay with the diagnosis of pneumonia due to Varicella-Zoster virus and medicated with intravenous acyclovir and levofloxacin for 7 days. A good clinical, analytical, and imaging response was observed (Fig. 1B). During the hospitalization, a thoracic computed tomography (CT) scan was done, showing multiple small nodules scattered in the pulmonary parenchyma, some forming small conglomerates and surrounding ground glass. They presented bilateral distribution, but predominantly in the lower lobes. The largest conglomerate measured was 15 mm in diameter and was located in the lower-left lobe. (Fig. 1C). The patient was discharged asymptomatic.

After one year, the patient returned to the emergency department with fever, odynophagia, cough, and haemoptysis, showing no changes in physical examination or blood analysis. The following thoracic CT revealed a micronodular pattern with mostly calcified nodules. In the lower right lobe, a dense 19 mm nodule with ground-glass pattern and, juxtaposed to this, other calcified 19 mm and 5.8 mm nodules were found. There was a growth of the previously reported lesion in the lower-left lobe, which measured 21 mm. (Fig. 2A).

Blood and sputum culture were negative and bronchoalveolar lavage showed no changes. CT guided biopsy of the lower left lobe lesion was performed, with anatomopathological exam revealing focal lesions of organizing pneumonia with foamy macrophages in intralveolar localization (Fig. 2B). After the exclusion of other causes of organizing pneumonia and considering the previous diagnosis of varicella-zoster pneumonia, this cause was assumed. Corticosteroid therapy was started with an initial dose of oral prednisolone of 30 mg (0,5 mg/kg) daily for 6 months. Symptoms and imaging gradually improved and, after 1 year, clinical or imaging recurrence was not observed (Fig. 2C).

The present report describes a case of OP associated with varicella-zoster infection. Hypersensitivity pneumonitis (HP) could have a very similar clinical presentation such as fever, cough, and ground glass consolidations on CT and has a high incidence among Portuguese population in contact with birds, mould, cork or isocyanates.[7] However, besides an identifiable exposure history, imaging changes are usually upper lobe predominant. Despite the usually good prognosis with response to antigen avoidance, some chronic forms, mainly fibrotic, could have the same prognosis as idiopathic pulmonary fibrosis.[8] As previously stated, OP is an histological pattern associated with a variety of disorders,[1,4] however, OP is rare after viral infections. There are a few cases reported of influenza

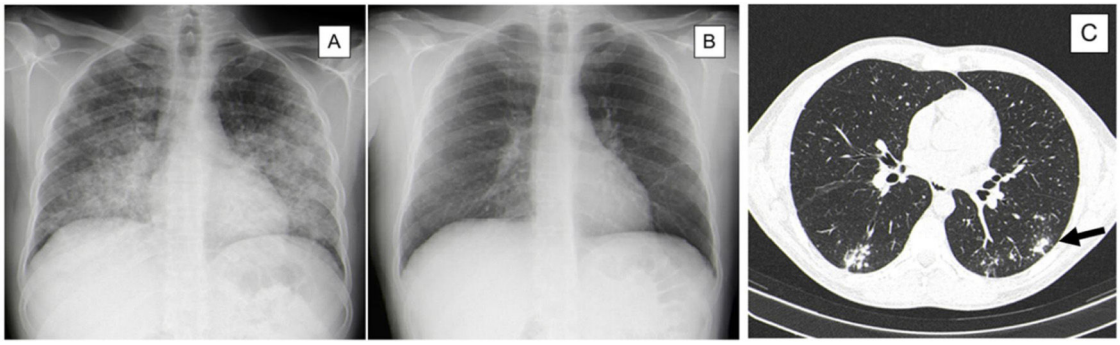

Figure 1 Imaging evolution of Varicella-Zoster pneumonia.
Posteroanterior chest radiograph evolution during Varicella-Zoster Pneumonia. A) On admission with bilateral opacities with air bronchogram. B) On discharge day showing almost complete resolution of the previously described lesions.
Thoracic computed tomography. C) On final days of hospitalization by varicella-zoster pneumonia showing multiple small nodules scattered in the pulmonary parenchyma, some forming small conglomerates, and surrounding ground glass with bilateral distribution, but predominantly in the lower lobes. The largest conglomerate measuring 15 mm in the lower left lobe (arrow).

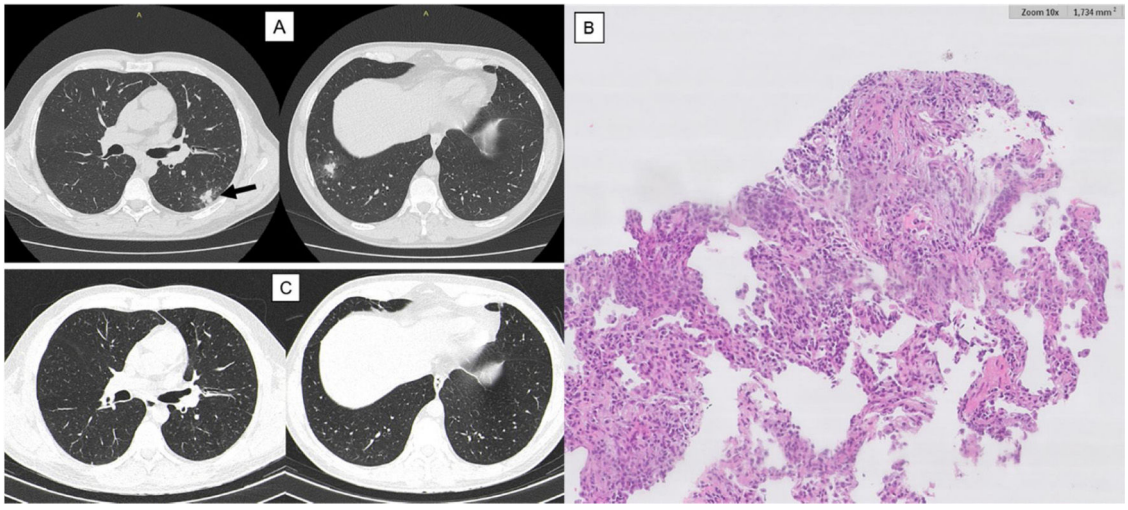

Figure 2 Imaging evolution and histology of Organizing Pneumonia secondary to Varicella-Zoster infection.
Thoracic computed tomography (CT). A) One year after varicella-zoster infection showing a micronodular pattern with mostly calcified nodules. In the lower right lobe, a dense 19 mm nodule with ground-glass pattern and, juxtaposed to this, other two calcified nodules. Growth of the previously reported lesion in the lower-left lobe, measuring 21 mm (arrow). **CT guided biopsy of the lower left lobe lesion.** B) Anatomopathological exam showing focal lesions of organizing pneumonia with foamy macrophages in intralveolar localization. **Thoracic CT.** C) One year after corticosteroid treatment maintaining micronodular pattern with calcified nodules, but with complete resolution of the previously described lesions.

association,[5] while the association with varicella-zoster is even rarer. The few cases reported of OP after varicella-zoster infection occurred in patients with known risk factors for varicella-zoster infection, such as cigarette smoking, pregnancy, immunosuppression and male sex.[9,10] Early diagnosis and treatment contribute to a favourable prognosis. We acknowledge the need for the physicians' awareness of the secondary OP after varicella-zoster infection, even if the disease had previously been cured.

Patient's consent

Patient's informed consent was obtained.

Conflicts of interest

The authors have no conflicts of interest to declare.

References

1. American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias. Am J Respir Crit Care Med. 2002;165(2):277–304.
2. Travis WD, Costabel U, Hansell DM, King TE, Lynch DA, Nicholson AG, et al. American Thoracic Society Documents An Official American Thoracic Society / European Respiratory Society Statement: Update of the International Multidisciplinary Clas-

sification of the Idiopathic Interstitial Pneumonias. Am J Respir Crit Care Med. 2013;188(6):733–48.
3. Zhou H, Gu W, Li C. Post-infectious organizing pneumonia: an indistinguishable and easily misdiagnosed organizing pneumonia. Clin Lab. 2015;61(11):1755–61.
4. Schwarz MI, King TE. Interstitial lung disease. People's Medical Pub. House; 2011. p. 1161.
5. Asai N, Yokoi T, Nishiyama N, Koizumi Y, Sakanashi D, Kato H, et al. Secondary organizing pneumonia following viral pneumonia caused by severe influenza B: a case report and literature reviews. BMC Infect Dis. 2017;17:572.
6. Khan MS, Khateeb F, Akhtar J, Khan Z, Lal A, Kholodovych V, et al. Organizing pneumonia related to electronic cigarette use: a case report and review of literature. Clin Respir J. 2018;12(3):1295–9.
7. Santos V, Martins N, Sousa C, Jacob M, Padrão E, Melo N, et al. Hypersensitivity pneumonitis: main features characterization in a Portuguese cohort. Pulmonology. 2020;26:130–7.
8. Alberti ML, Malet Ruiz JM, Fernández ME, Fassola L, Caro F, Roldán IB, et al. Comparative survival analysis between idiopathic pulmonary fibrosis and chronic hypersensitivity pneumonitis. Pulmonology. 2020;26(1):3–9.
9. Hockberger RS, Rothstein RJ. Varicella pneumonia in adults: a spectrum of disease. Ann Emerg Med. 1986;15(8):931–4.
10. Ferreira Santos C, Gomes A, Garrido A, Albuquerque A, Melo E, Barros I, et al. Pneumonia a Varicella zoster. Rev Port Pneumol (English Ed.). 2010;16(3):493–505.

M. Oliveira[a,*], S. Braga[a], F. Fernandes[a,b], J.M. Silva[a,b]

[a] *Pulmonology Department, Unidade Local de Saúde da Guarda, Guarda, Portugal*
[b] *Faculdade de Ciências da Saúde da Universidade da Beira Interior, Covilhã, Portugal*

*Corresponding author at: Avenida Rainha Dona Amélia, 6300-259 Guarda, Portugal.
E-mail address: marcosandre.oliveira90@gmail.com (M. Oliveira).

6 October 2020

https://doi.org/10.1016/j.pulmoe.2020.10.006

Daytime non-invasive ventilatory support via intermittent abdominal pressure for a patient with Pompe disease

To the editor:

Late onset Pompe disease (LOPD) is a glycogen storage disorder characterised by progressive skeletal and respiratory (i.e. diaphragmatic) muscle weakness and ventilatory pump failure (VPF).[1] Enzyme replacement therapy (ERT) can prolong survival.[2] Noninvasive ventilation (NIV) is often used with low bi-level positive airway pressure (PAP) which may be inadequate for optimal respiratory muscle rest and ventilator support. Higher pressure support (PS) of 14 cm H2O or more often called Noninvasive ventilatory support (NVS) needs to be used. While NIV can improve gas exchange, sleep, and quality of life (QOL) for mild cases, only increased pressures can be used for continuous support to prolong survival.[3] Both NIV and NVS can be provided during sleep via different interfaces. The nasal mask or mouth-piece is better during daytime. Intermittent abdominal pressure ventilator (IAPV) can also provide daytime support. It consists of an air-sack inside a corset inflated by a ventilator. The sack compresses the abdomen to raise the diaphragm and to increase tidal volumes.[4,5] IAPV use by LOPD patients has not yet been described, and we aimed to report its feasibility in this study.

Case Study: A 22-year-old male university student, diagnosed with LOPD at age two, began bi-weekly ERT at age 11. The ERT was adjusted over the years based on his body weight. At age 13 he began nocturnal CPAP for obstructive sleep apnoea (OSA). Within two years, due to hypercapnia, he was transitioned to NIV, and since then continued with limited follow-up. Settings: inspiratory (I)PAP 12, expiratory (E)PAP 4 cm H2O, respiratory rate 14/min which he presented to us using in 2019. During our first visit, he had severe scoliosis, emaciation (BMI 12.8 kg/m^2), and recent onset of hyper-somnolence and morning headaches. Pulmonary function testing (PFT), arterial blood gases (ABG), and polysomnography (PSG) using NIV data are in Table 1. His QoL was assessed by the McGill Questionnaire.[6] Over the previous 30 days mean nocturnal use was 9 h with average Vt 350 mL with 85% of the breaths triggered. Then, IPAP

Table 1 Baseline characteristics.

	Baseline (T0)
SB	
pH	7.41
pCO2 (mmHg)	51
pO2 (mmHg)	87
HCO3- (mmHg)	32,3
BE (mol/l)	6.7
SNIP (cmH2O)	30
PEF (L/min)	250
SVC (L, %)	1,10 (20)
FVC (L, %)	1,10 (21)
FEV1 (L, %)	1 (22)
NIV	
AHI	11,4
TST90 (%)	0

SB, spontaneous breathing; BE, base excess; SNIP, sniff nasal inspiratory pressure; PEF, peak expiratory flow; SVC, slow vital capacity; FVC, forced vital capacity; FEV1, forced expiratory volume in 1 s; NIV, Non-invasive ventilation; AHI, apnea-hypopnea index; TST90, total sleep time with oxyhemoglobin saturation below 90%.