Case 16425

Pulmonary alveolar proteinosis
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Section: Chest imaging
Area of Interest: Lung Thorax
Procedure: Diagnostic procedure
Imaging Technique: CT
Imaging Technique: CT-High Resolution
Case Type: Clinical Cases
Authors: Filipa Vilas Boas, António Valente1, João Amorim2, Pedro Ninitas1, Leonor Moutinho3, Isabel Sapeira3
Patient: 31 years, female

Clinical History:
A 31-year-old female teacher, non-smoker, with no relevant past medical history and HIV-negative, presented with a 3-month history of progressive dyspnoea and cough with minimal mucoid sputum. Physical examination revealed crackles in pulmonary auscultation, and the rest of the examination was unremarkable. Laboratory workup showed an elevated value of lactate dehydrogenase.

Imaging Findings:
A chest radiograph was initially performed, which revealed bilateral ground-glass opacities, more expressive in the middle and lower lung fields. High resolution computed tomography (HRCT) of the thorax showed bilateral alveolar ground-glass attenuations with superimposed interlobular septal thickening, in a mosaic pattern called “crazy-paving”. There were no other noteworthy imaging findings. The radiological manifestations were unspecific and a bronchoscopy with a transbronchial biopsy was performed for clarification. A histopathologic diagnosis of pulmonary alveolar proteinosis was made. The bronchoalveolar lavage fluid showed abundant acelular material. The patient underwent whole lung lavage without significant improvement.

Discussion:
Pulmonary alveolar proteinosis (PAP) is a rare disease characterised by alveolar accumulation of lipoproteinaceous material, which blends with periodic acid-Schiff (PAS) [1, 2]. It may be congenital, primary or idiopathic and less commonly it presents secondarily to various conditions, that can be divided into three categories: lung infections, haematological diseases or conditions that alter the patient’s immunity and exposure to various types of chemicals, such as silica particles [2].

The underlying pathophysiological mechanism corresponds to an alteration of pulmonary surfactant homeostasis and lung immune function. Idiopathic PAP has antibodies against granulocyte-macrophage colony-stimulating factor (GM-CSF), which results in decreased surfactant degradation and its accumulation [3]. PAP is more frequent during the third and fourth decades of life [4]. A strong relationship with smoking is established, with men being more affected than women [3, 5]. The clinical presentation of PAP is unspecific. Frequently, respiratory symptoms are
moderate and have an insidious onset. Dyspnoea and dry or minimally productive cough are the most common presenting symptoms. Physical examination is commonly normal, but may reveal auscultatory crackles, clubbing and cyanosis. The predominant abnormality in serologic tests is an increased lactate dehydrogenase level [3]. The typical chest radiography findings are central, bilateral and symmetrical alveolar opacities yielding a “butterfly” distribution pattern, with relative sparing of the apices and costophrenic angles. A less common appearance may correspond to asymmetrical opacities, extensive diffuse consolidation and a mixed alveolar-interstitial or nodular pattern. The most common findings on chest HRCT include ground-glass opacities with geometric distribution and smooth thickening of the interlobular septa, which together result in the so-called "crazy-paving" pattern, which is highly suggestive but not pathognomonic. The thickened interlobular septa are present only within the areas of ground-glass opacities and correspond to septal oedema. The distribution is typically bilateral and, unlike secondary forms, there is no predominance of any specific lung region. There is often a marked discrepancy between the patient's clinical and radiological manifestations, which are more pronounced [2-5]. Treatment depends on the form of PAP: pulmonary support or lung transplantation in the congenital disease; removal of causative agent in the secondary PAP and whole-lung lavage and GM-CSF supplementation, with varying degrees of effectiveness, in the idiopathic form [3, 5]. Prognosis is variable, ranging from clinical improvement with treatment to a chronic and terminal course that results from respiratory failure or pulmonary infection [1, 4].

Written patient consent for this case was waived by the Editorial Board. Patient data may have been modified to ensure patient anonymity.

Differential Diagnosis List: Pulmonary alveolar proteinosis, Organizing pneumonia, Pneumocystis pneumonia, Cardiogenic pulmonary oedema - Diffuse alveolar haemorrhage

Final Diagnosis: Pulmonary alveolar proteinosis

References:
Figure 1

Description: Chest radiography, posteroanterior view, showed symmetric, perihilar ground-glass opacities, predominant in middle and lower lung fields. Origin: Department of Radiology, Centro Hospitalar de Lisboa Central, Portugal, 2019
Description: HRCT scan of the thorax with axial sections (lung window) demonstrates bilateral widespread ground-glass opacities with focal areas of sparing and strikingly prominent septal lines, corresponding to a “crazy paving” pattern. Origin: Department of Radiology, Centro Hospitalar de Lisboa Central, Portugal, 2019
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**Figure 3**

**Description:** HRCT scan of the thorax with coronal section (lung window) demonstrates the diffuse distribution of the “crazy-paving” pattern. **Origin:** Department of Radiology, Centro Hospitalar de Lisboa Central, Portugal, 2019
Description: HRCT scan of the thorax with sagittal section (lung window) demonstrates the diffuse distribution of the “crazy-paving” pattern. Origin: Department of Radiology, Centro Hospitalar de Lisboa Central, Portugal, 2019