Case 13963

Eurorad

Knowledge of shadows: Pulmonary alveolar proteinosis

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Section: Chest imaging
Area of Interest: Lung Thorax
Procedure: Diagnostic procedure
Procedure: Instrumentation
Procedure: Comparative studies
Imaging Technique: Experimental
Imaging Technique: Conventional radiography
Imaging Technique: CT-High Resolution
Special Focus: Acute Case Type: Clinical Cases
Authors: Parra-Fariñas C, Andreu J, Persiva O, Varona E, Pallisa E, Almazán E, Ortiz-Andrade C, Prat JA, Hernandez C, Dyer Hartnett S, Comet R.
Patient: 39 years, female

Clinical History:

A 39-year-old male patient, known smoker, presented with fever, dry cough, and progressive breathlessness for two months.

Initial evaluation confirmed hypoxia, and pulmonary function tests (PFT) were suggestive of a restrictive pattern (Fig. 1).

Imaging Findings:

Plain chest radiography demonstrated bilateral hazy airspace opacities with perihilar infiltrates and diffuse reticulonodular lesions in both lung fields, mostly centrally located (Fig. 2).

High resolution computed tomography (HRCT) showed bilateral interstitial confluent pulmonary infiltrates, ground-glass opacities, and smooth thickening of interlobular and intralobular septal lines. The combination of these features is termed "crazy-paving" pattern (Fig. 3).

The patient was intubated with a double-lumen endotracheal tube for whole lung lavage (WLL). The specimens of fluid contained large amounts of granular acellular eosinophilic proteinaceous material with morphologically abnormal "foamy" macrophages engorged with periodic acid-schiff (PAS) positive intracellular inclusions, findings compatible with primary pulmonary alveolar proteinosis (PAP). The procedure was terminated once the effluent had cleared significantly (Fig. 4).

Chest radiography and HRCT were performed one week after the WLL. They showed marked decrease in the extent of the opacities noticed in the previous studies (Fig. 5, 6).

Discussion:

PAP, also known as pulmonary alveolar phospholipoproteinosis, is a rare disorder of unknown aetiology, first described in 1958 by the physicians Samuel Rosen, Benjamin Castleman, and Averill Liebow, in which lipoproteinaceous material accumulates within alveoli, interfering with gas exchange [1].

Three main categories of PAP have been defined depending on the aetiology: genetic, primary (also called idiopathic), and secondary [2, 3].

Patients are typically aged 20-50 years at diagnosis, more common in males and tobacco smokers [4, 5]. They usually present with progressive dyspnoea and cough. Less common symptoms include fever, chest pain, or haemoptysis [1, 2].

Physical examination signs can be unremarkable, but there are inspiratory crackles, cyanosis, and digital clubbing. In uncomplicated cases, chest radiography usually reveals bilateral ill-defined nodular or confluent pattern, suggestive of pulmonary oedema but without other findings of left-sided heart failure [1]. HRCT shows patchy, ground-glass opacities with septal thickening, a pattern commonly referred to as “crazy-paving” [5].

PFT can be normal but typically show a restrictive ventilatory defect with a disproportionate and severe reduction of the carbon monoxide diffusing capacity [5, 6]. The impairment of gas exchange is secondary to filling of the alveoli with proteinaceous material leading to ventilation and perfusion mismatch.

Clinical and radiological findings suggest the diagnosis of PAP in suspected cases, while findings on examination of a bronchoalveolar lavage specimen can establish the diagnosis. The lavage fluid has an opaque and milky appearance. It contains large alveolar macrophages filled with PAS positive material and increased numbers of lymphocytes [7]. On light-microscopy, the normal alveolar architecture is generally preserved unless there is infection. Immunohistochemical staining reveals abundant accumulation of surfactant protein [8].

Therapy for all types of PAP remains WLL, although for the primary form of the disorder, successful lung transplantation has been reported [9]. Idiopathic PAP has been treated successfully since the early 1960 by WLL, and this procedure remains the standard of care today [10]. Secondary PAP treatment involves WLL and underlying condition therapy.

TEACHING POINTS:
1. Abnormal processing of surfactant by macrophages with amorphous proteinaceous material deposition in the alveoli.
2. Subacute presentation with a gradual onset of symptoms.
3. Imaging findings
• Chest radiography: Symmetric and bilateral alveolar opacities
• HRCT: “Crazy-paving” pattern
4. WLL: First line of treatment.

Differential Diagnosis List: Primary pulmonary alveolar proteinosis (PAP)., Cardiogenic pulmonary oedema., Hypersensitivity pneumonitis, Non-small cell lung cancer, Pneumocystis jirovecii pneumonia, Sarcoidosis, Small cell lung cancer

Final Diagnosis: Primary pulmonary alveolar proteinosis (PAP).

References:

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Ben-Dov I, Kishinevski Y, Roznman J, Soliman A, Bishara H, Zelligson, et al (1999) Pulmonary alveolar proteinosis in Israel: Ethnic clustering. Isr Med Assoc J 1:75–8 (PMID: 10731299)

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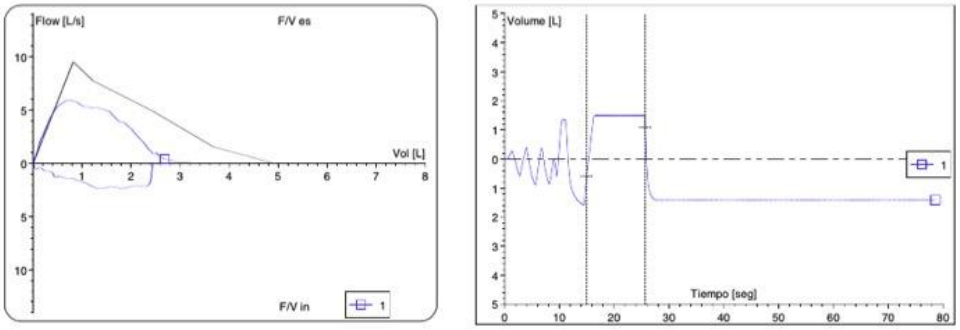
Figure 1

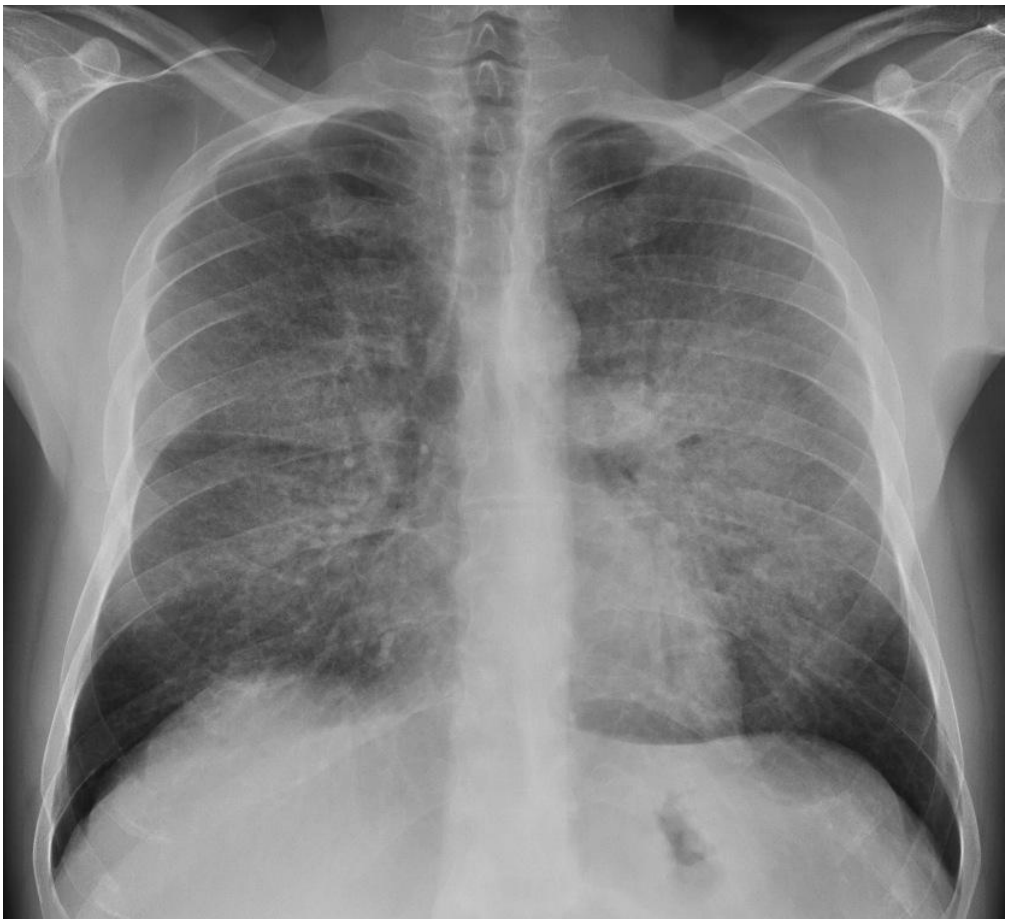
a



Description: Pulmonary function tests showed restrictive pattern. **Origin:** 2016 Department of Radiology, Vall d´Hebron Hospital. Barcelona, Spain.

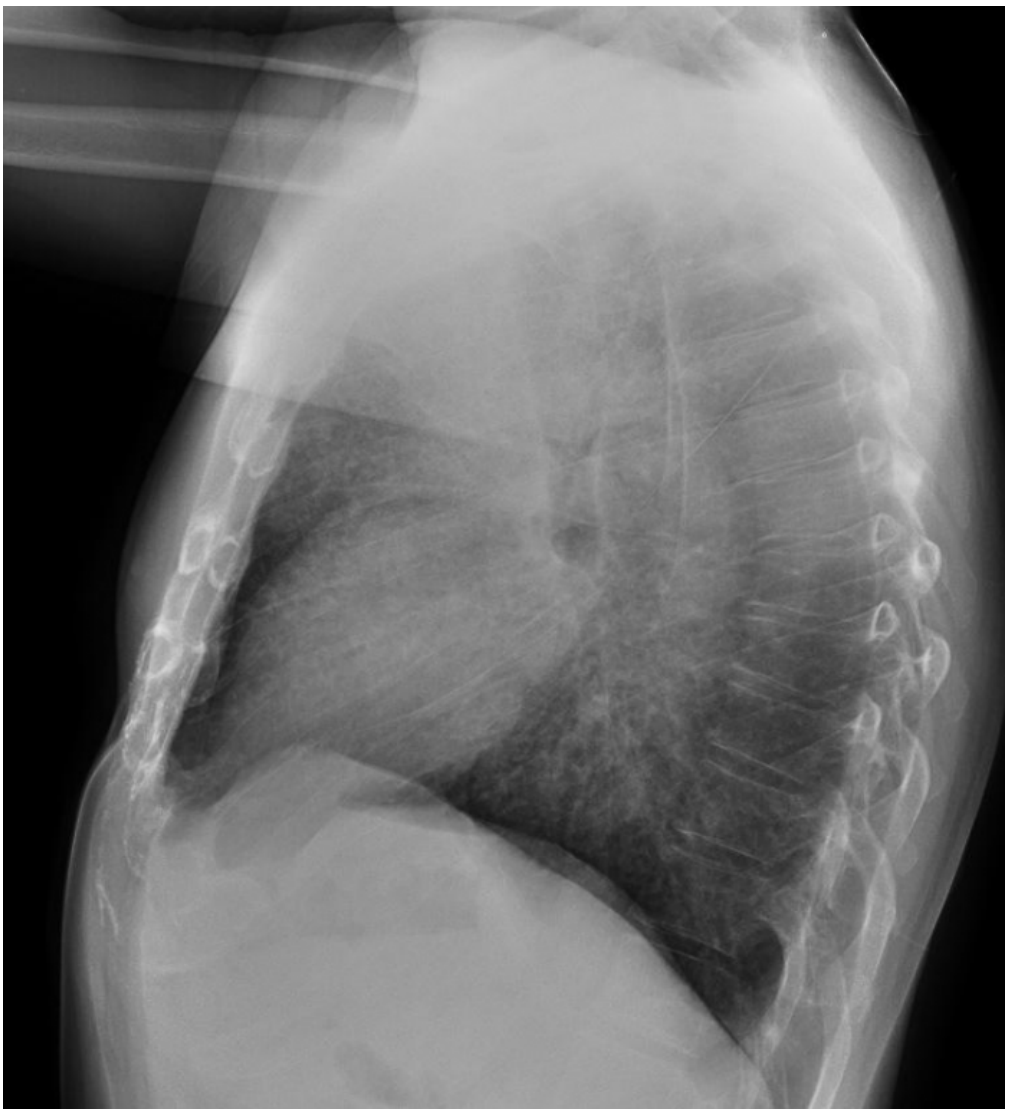
Figure 2

a



Description: Posteroanterior conventional chest X-ray revealed symmetric, bilateral alveolar opacities, without air bronchogram, showing a perihilar and basal distribution. **Origin:** 2016 Department of Radiology, Vall d´Hebron Hospital. Barcelona, Spain.

b



Description: Left lateral conventional chest X-ray revealed symmetric, bilateral alveolar opacities, without air bronchogram, showing a perihilar and basal distribution. **Origin:** 2016 Department of Radiology, Vall d´Hebron Hospital. Barcelona, Spain.

Figure 3

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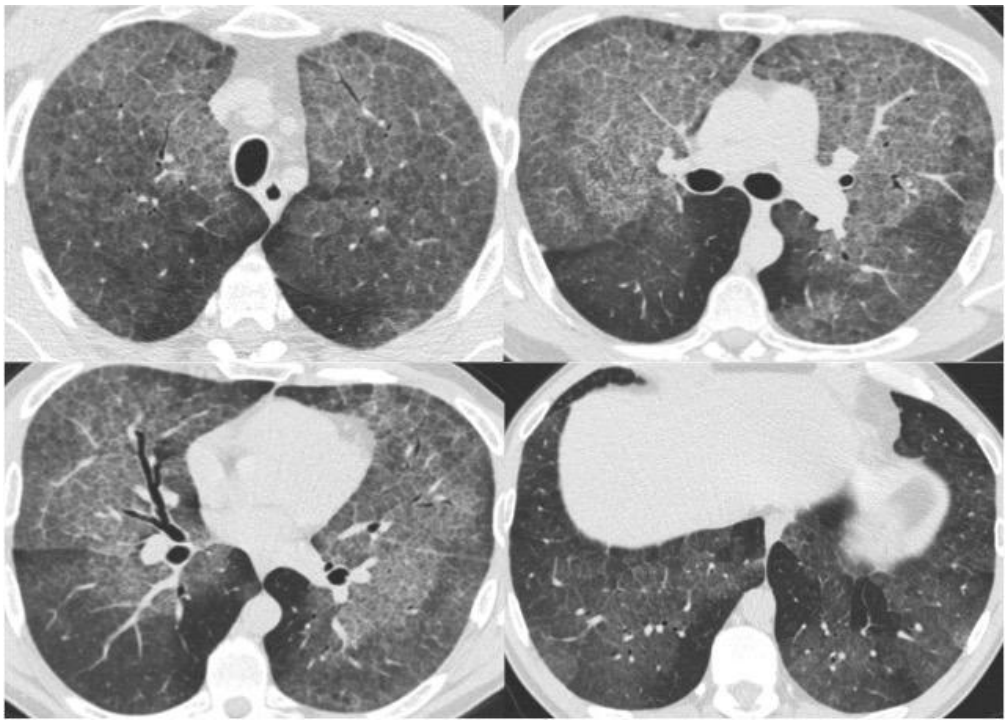
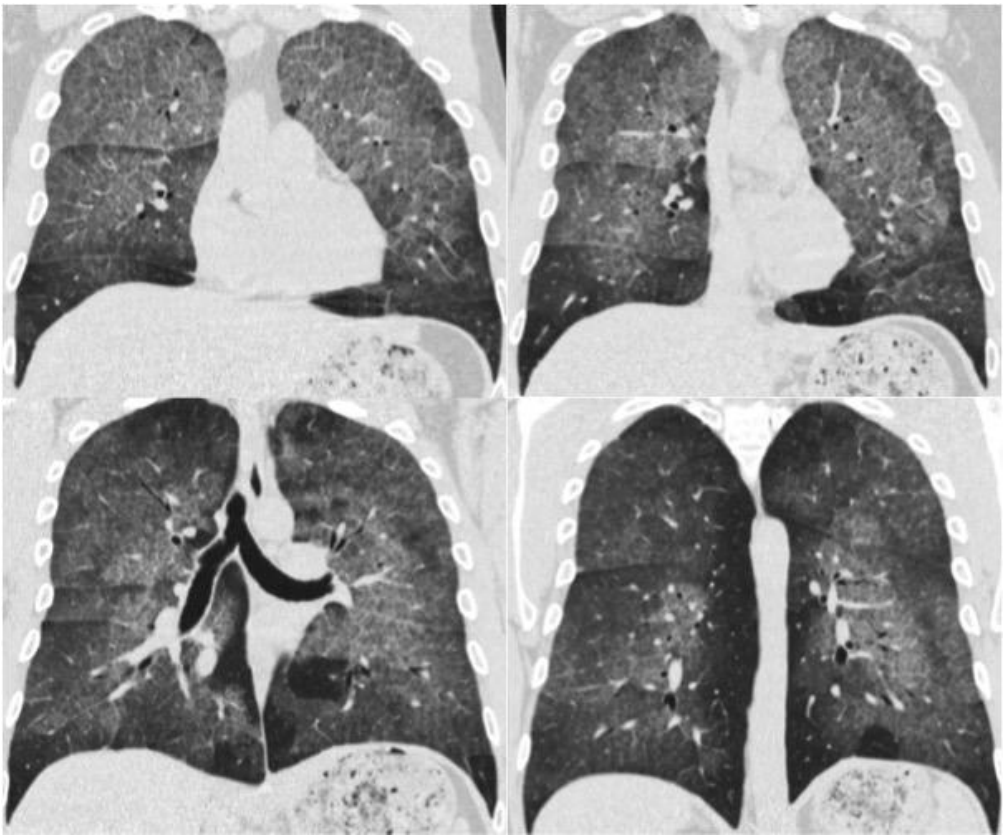


Description: Axial HRCT showed reticulations superimposing on ground-glass opacities forming a “crazy-paving” pattern with a geographic distribution: juxtaposition of healthy and sick zones. **Origin:** 2016 Department of Radiology, Vall d´Hebron Hospital. Barcelona, Spain.

b



Description: Axial HRCT, coronal reconstruction, showed reticulations superimposing on ground-glass opacities forming a “crazy-paving” pattern with a geographic distribution: juxtaposition of healthy and sick zones. **Origin:** 2016 Department of Radiology, Vall d´Hebron Hospital. Barcelona, Spain.

Figure 4

a

Description: Photograph of WLL demonstrating an opaque, milky-appearing fluid with sediments.
Origin: 2016 Department of Radiology, Vall d´Hebron Hospital. Barcelona, Spain

Figure 5

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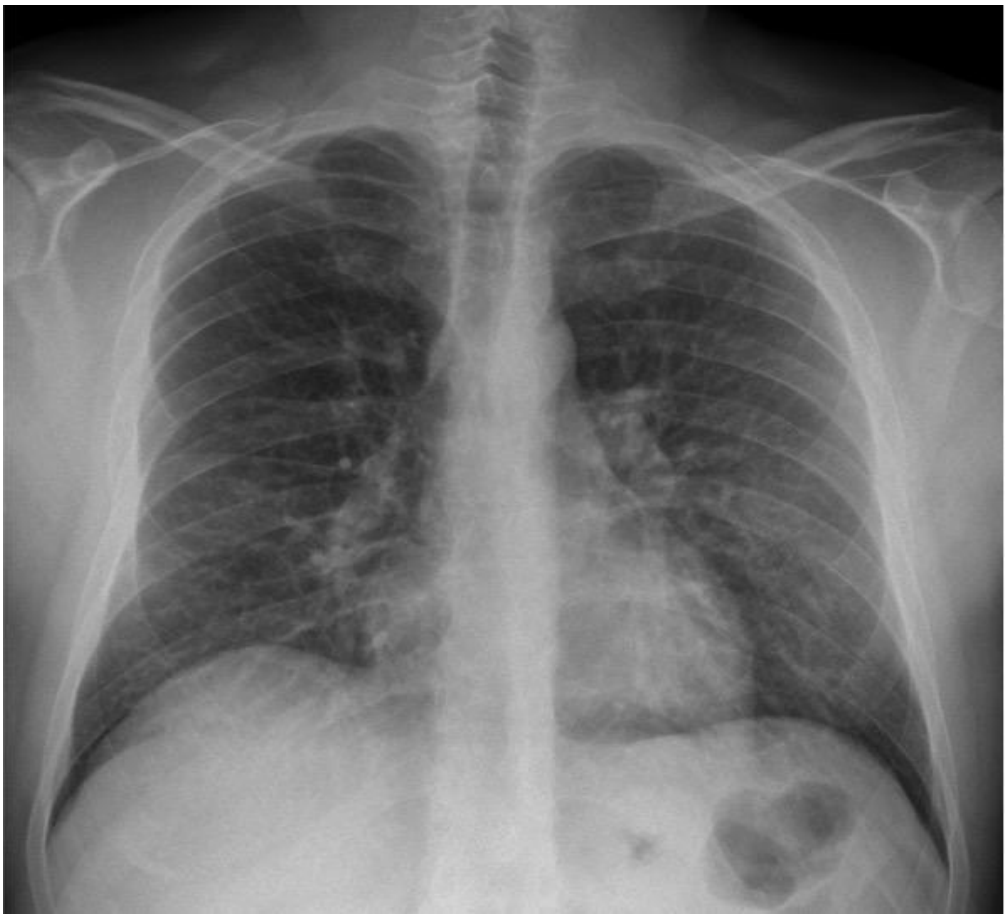
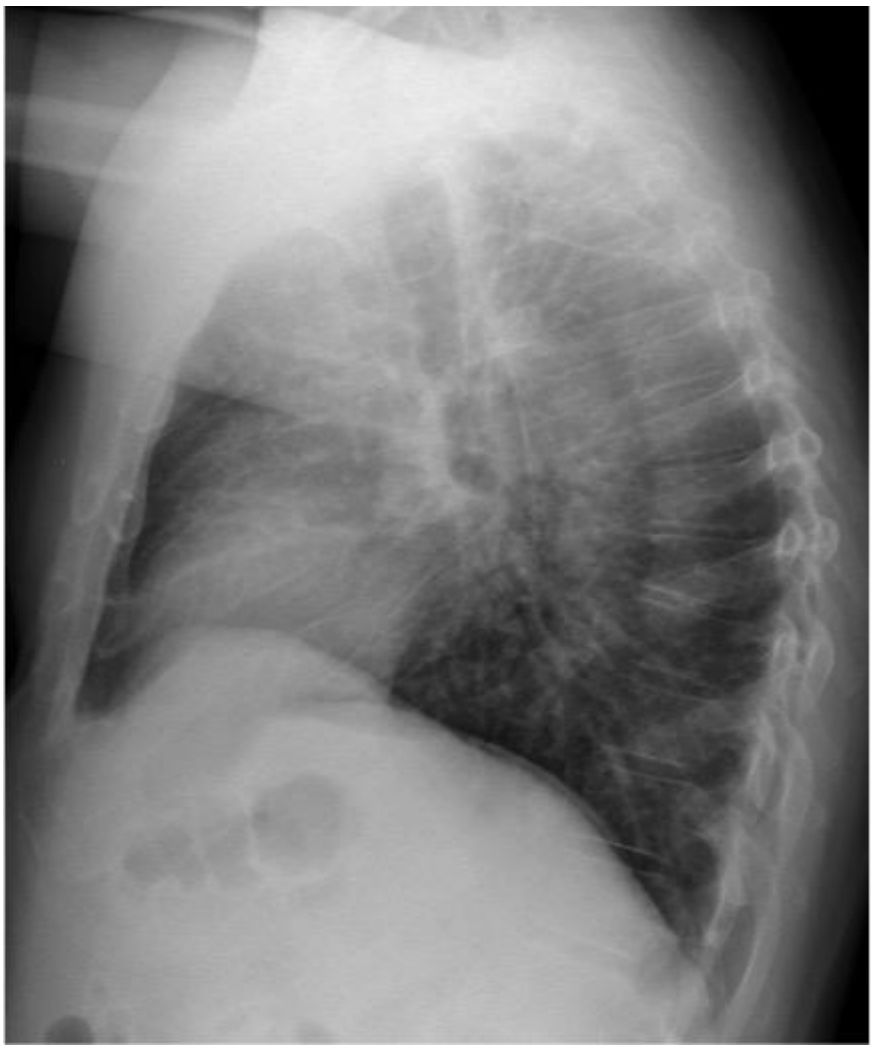


Description: Posteroanterior conventional chest X-ray after one week of WLL showed improvement of the infiltrative process. **Origin:** 2016 Department of Radiology, Vall d´Hebron Hospital. Barcelona, Spain.

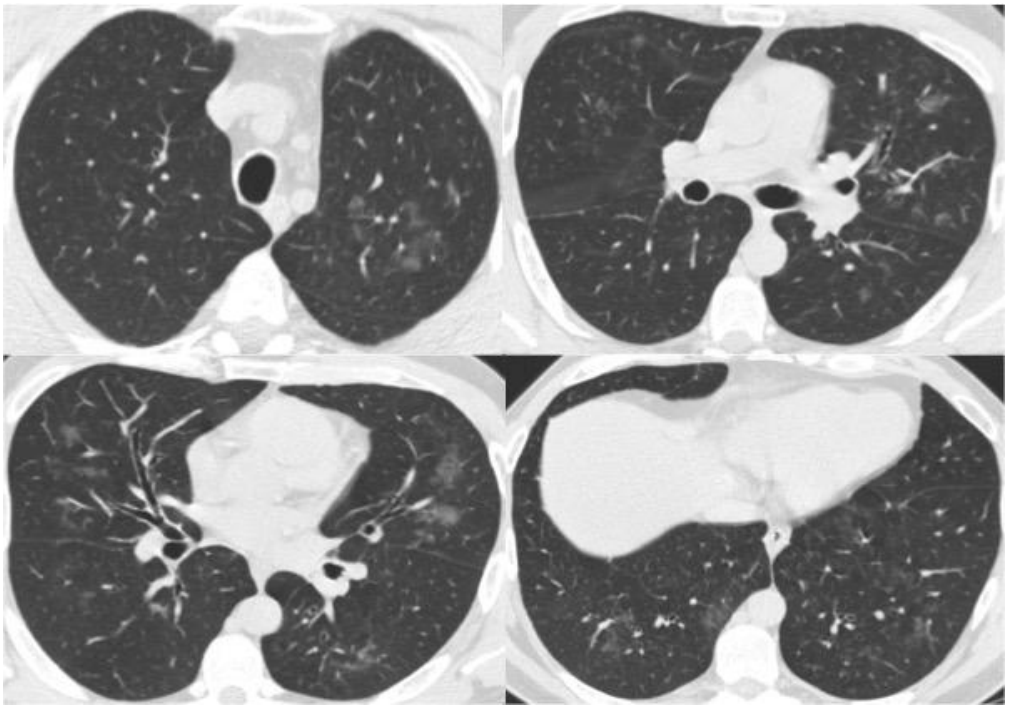
b



Description: Left lateral conventional chest X-ray after one week of WLL showed improvement of the infiltrative process. **Origin:** 2016 Department of Radiology, Vall d´Hebron Hospital. Barcelona, Spain.

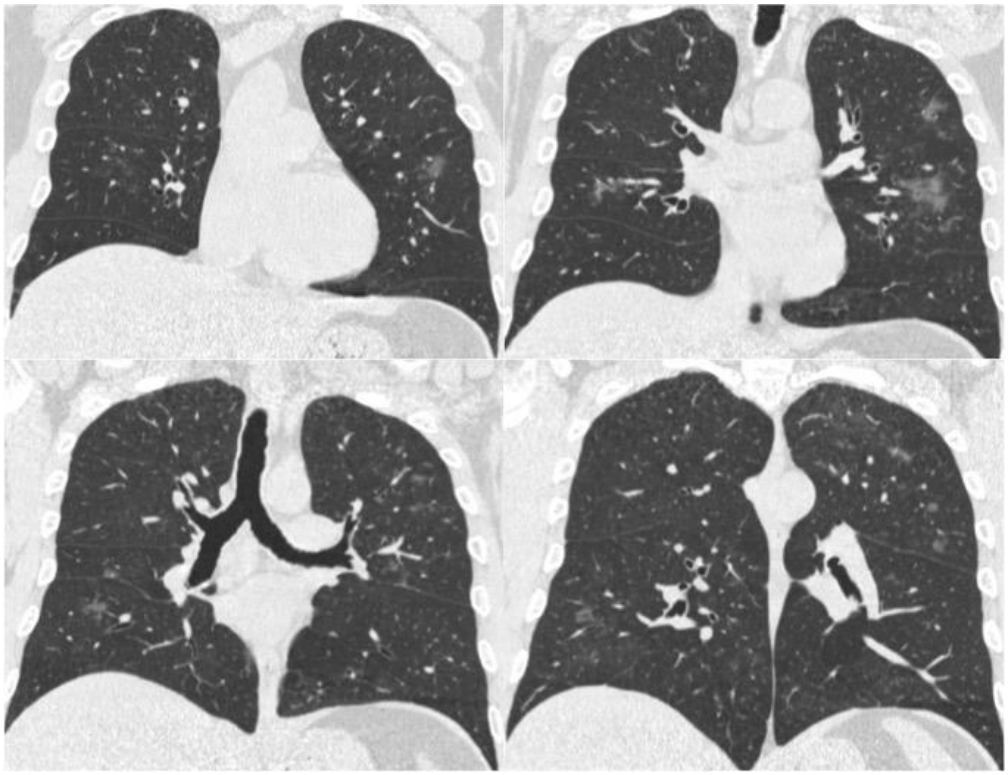
Figure 6

a



Description: Axial HRCT after one week of WLL showed marked decrease in the extent of the opacities. **Origin:** 2016 Department of Radiology, Vall d´Hebron Hospital. Barcelona, Spain.

b



Description: Axial HRCT, coronal reconstruction, after one week of WLL showed marked decrease in the extent of the opacities. **Origin:** 2016 Department of Radiology, Vall d´Hebron Hospital. Barcelona, Spain.