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Pleuroparenchymal Fibroelastosis: Case Study

M. Moutahir, C. Benrakkoun, W. Omari, N. Rguig, M. Aharmim , M.Soualhi , H.Charaf, R. Zahraoui
Department Of Pneumology, Moulay Youssef Hospital, Rabat-Morocco

Abstract
Pleuroparenchymal fibroelastosis is a rare and recently recognized condition, first described by Amitani et al. in the 1990s as fibrosis of the upper lobes and later recognized as a new clinicopathological entity by Frankel. In 2013, idiopathic pleuroparenchymal fibroelastosis was classified as a rare subtype in the international classification of idiopathic interstitial pneumonias. The disease can occur at any age, and the diagnosis is suspected in a patient with dyspnea and radiological presentation, characterized by fibrosis of the upper lobe of the pleural and subpleural pulmonary parenchyma. A definitive diagnosis is histopathological, characterized by dense intra-alveolar fibrosis with elastosis of the alveolar walls and fibrous thickening of the subpleural pleura, predominantly in the upper lobes. The disease progresses, with a poor prognosis, and currently, the only therapeutic option is lung transplantation. We describe a rare case of pleuroparenchymal fibroelastosis diagnosed radiologically in an 83-year-old man.

Keywords: *pleuroparenchymal fibroelastosis, pleural thickening, subpleural fibrosis, lungtransplantation*

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I. Introduction

Pleuroparenchymal fibroelastosis is a rare, slowly progressive idiopathic interstitial pneumonia recently included in the updated classification of idiopathic interstitial pneumonias. It is characterized by predominant pleural fibrosis in the upper lobes and underlying parenchyma. Complications such as pneumothorax are common. The origin ofpleuroparenchymal fibroelastosis is not clearly understood; it may be associated with underlying conditions or be idiopathic (1). We describe a case of pleuroparenchymal fibroelastosis diagnosed radiologically in an 83-year-old man.

Case Observation

This case involves an 83-year-old male, a former farmer, who had never been treated for tuberculosis and had no recent history of tuberculosis exposure or toxic habits. He reportedprogressive worsening dyspnea over the past year, reaching stage 4 on the mMRC scale, associated with a dry cough, chest pain, and mechanical low back pain, all in a context of apyrexia, anorexia, and significant weight loss. Clinical examination revealed a cachectic patient with a BMI of 15 kg/m², a WHO performance score of 4, slightly polypneic with a respiratory rate of 22 cycles/minute, correctly saturated on ambient air, slight digital clubbing, and pleuropulmonary crackles on auscultation. A chest radiograph showed bilateral nodular opacities with pleural thickening and bilateral blunting of both pleural recesses (Figure 1). Thoracic ultrasound did not reveal pleural effusion. A thoracic CT scan showed pleural thickening with bilateral nodular opacities and septal thickening, with a predominance of lesions in the upper lobes (Figure 2). Biologically, there was an inflammatory syndrome with a sedimentation rate of 60 mg/L, normal HLA. Functionally, plethysmography showed a severe restrictive ventilatory defect with a total lung capacity of2,40 L (43%), and arterial blood gas analysis showed hypoxemia at 53 mmHg with an indication for long-term oxygen therapy. A lung biopsy was not performed due to the suggestive CT scan appearance. The patient is following a respiratory rehabilitation program.

Figure 1: Bilateral nodular opacities with pleural thickening, bilateral blunting of the twopleural recesses

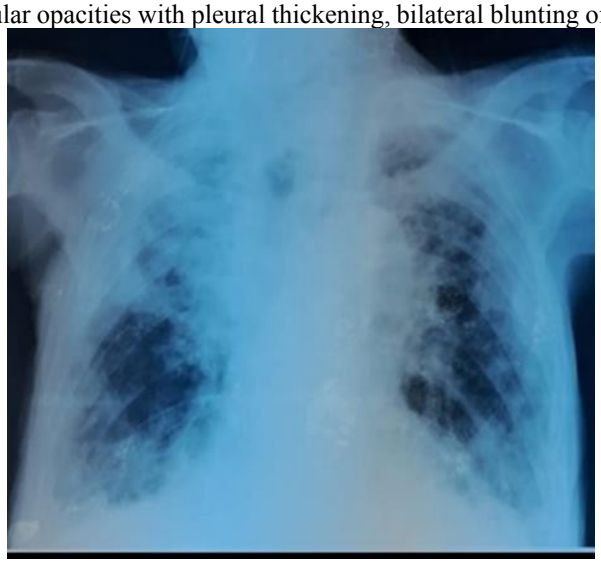
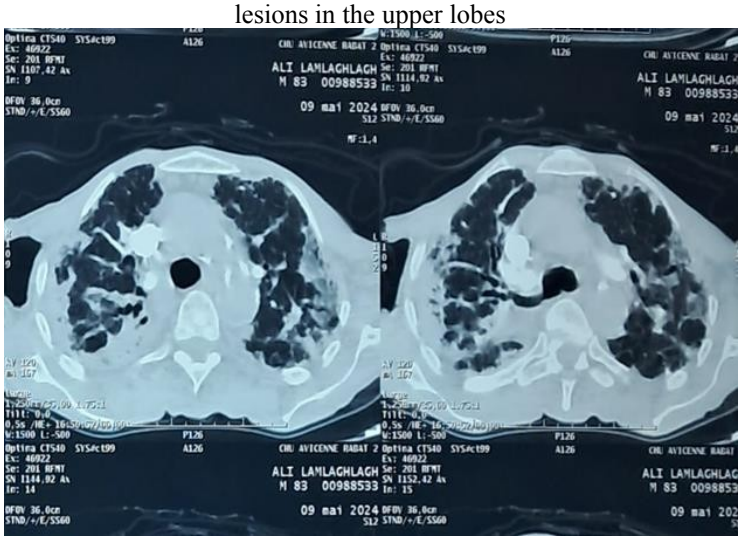


Figure 2 : pleural thickening with bilateral nodular opacities and septal thickening, with apredominance of lesions in the upper lobes



II. Discussion

Idiopathic pleuroparenchymal fibroelastosis first appeared in the ATS/ERS classification ofidiopathic interstitial pneumonias in 2013(tableau 1) (3). It is a rare clinicopathological entity first described under this term in a 2004 publication, which grouped the first five formally diagnosed cases of pleuroparenchymal fibroelastosis, all sharing a pattern of chronic interstitial and pleural fibrosis not corresponding to other categories of idiopathic interstitial pneumonias (2). However, it was first described in 1992 by Amitani et al. in Japanese literature as Amitani disease or other names like idiopathic pulmonary upper lobefibrosis (4).

Table 1. Revised American Thoracic Society/European Respiratory Society Classification Of Idiopathic Interstitial Pneumonias

Major idiopathic interstitial pneumonias
Idiopathic pulmonary fibrosis
Idiopathic nonspecific interstitial pneumonia
Respiratory bronchiolitis–interstitial lung disease
Desquamative interstitial pneumonia
Cryptogenic organizing pneumonia
Acute interstitial pneumonia
Rare idiopathic interstitial pneumonias
Idiopathic lymphoid interstitial pneumonia
Idiopathic pleuroparenchymal fibroelastosis
Unclassifiable idiopathic interstitial pneumonias*

The etiologies are mainly idiopathic and often iatrogenic, particularly after lung transplantation. Pleuroparenchymal fibroelastosis is associated with lung transplants as a late complication with a prevalence of 7,54% (6). After chemotherapy, a study highlighted the potential role of alkylating agents in the development of fibroelastosis, particularly cyclophosphamide (7,14). Other causes include bone marrow transplantation, hematopoietic stem cell transplantation (5), recurrent infections (for example pulmonary aspergillosis or Mycobacterium avium complex), cancers, radiotherapy, autoimmunity (1), occupational exposure to asbestos and aluminum. Smoking does not appear to be a risk factor (9).

The median age of disease onset is 57 years, with no sex predominance (8). Patients with pleuroparenchymal fibroelastosis may remain asymptomatic for a long time. Clinically, they present with progressively worsening dyspnea, cough, weight loss, and chest pain, such us our patient , sometimes indicative of pneumothorax(10). The presence of a flat chest is often noted on clinical examination, which is characteristic of the disease (11) and may be related to fibrosis and volume loss in the upper lobe. Digital clubbing, often seen in patients with idiopathic pulmonary fibrosis, is rarely reported in pleuroparenchymal fibroelastosis(12). Crackles are less frequent in pleuroparenchymal fibroelastosis than in idiopathic pulmonary fibrosis(8,5).

In the early stages of the disease, thoracic CT reveals reticular and nodular opacities limitedto the lung apices. As the disease progresses, pleuro-parenchymal thickening, septal thickening, traction bronchiectasis, and reduced lung volume are noted (12). In 2012, Reddy et al. proposed radiological criteria for diagnosing pleuroparenchymal fibroelastosis. A definitive diagnosis is made when thoracic CT shows pleural thickening of the upper lobesand subpleural fibrosis, with less or no involvement of the lower lobes. A diagnosis compatible with pleuroparenchymal fibroelastosis is when pleural thickening and subpleural fibrosis are present but not concentrated in the upper lobes(8). Histologically, Kusagaya et al. defined histological criteria for diagnosing pleuroparenchymal fibroelastosis, later expanded by Reddy et al (8,13).(table 2)

Pneumothorax and pneumomediastinum are common complications of pleuroparenchymal fibroelastosis (15). Pulmonary function tests reveal a predominant restrictive syndrome with an altered carbon monoxide diffusing capacity of the lungs . The disease progresses to restrictive respiratory failure, often hypercapnic. Differential diagnoses include tuberculosis sequelae, sarcoidosis type 4, interstitial lung disease associated with connective tissue diseases, particularly ankylosing spondylitis, and asbestosis (1).

There is no effective pharmacological treatment for pleuroparenchymal fibroelastosis; however, one study showed that antifibrotic treatment with nintedanib could reduce the rate of decline in functional vital capacity in patients with idiopathic or secondary pleuroparenchymal fibroelastosis (16) , but lung transplantation remains the definitive treatment for the disease. Chronic respiratory failure should be managed: respiratory rehabilitation, oxygen therapy, and non-invasive ventilation are necessary.

Table2: Diagnostic criteria for pleuroparenchymal fibroelastosis

Category	Histopathology	CT high resolution
Definitive	Upper lobe pleural fibrosis with subjacent intraalveolar fibrosis accompanied with alveolar septal elastosis	Pleural thickening with associated subpleural fibrosis in the upper lobes without involvement of the lower lobes
Consistent with PPFE	Presence of intraalveolar fibrosis but 1) not accompanied by significant pleural fibrosis, 2) not supleural predominance or 3) not present in a biopsy of the upper lobe	Pleural upper-lobe thickening with associated subpleural fibrosis but 1) not distributed in the upper lobes, or 2) with characteristics of coexistent disease in other sites
Inconsistent with PPFE	Absence of features of definitive and consistent diagnosis	Absence of features of definitive and consistent diagnosis

CT, computed tomography; PPFE, pleuropulmonary fibroelastosis.

III. Conclusion

Pleuroparenchymal fibroelastosis is a rare, slowly progressive disease whose first symptomis dyspnea or dry cough. Chest pain due to pneumothorax may be the first symptom in somepatients. The prognosis is poor, and the decline in vital capacity is rapid, with no effective treatment available except for lung transplantation.

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