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Case Report

Pleuroparenchymal fibroelastosis associated with aluminosilicate dust: a case report

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Abstract: Idiopathic pleuroparenchymal fibroelastosis (IPPFE) is a recently described rare condition, characterized by pleural and subpleural parenchymal fibrosis, predominantly in the upper lobes. The clinical course of this disease is progressive and prognosis is poor, with little information regarding the etiology of IIPPFE. This report describes an IPPFE patient with convincing evidence of inhalational dust and suggests that dust exposure should be considered as a new causative factor of IPPFE.

Keywords: Pleuroparenchymal fibroelastosis, idiopathic interstitial pneumonias, dust exposure

Introduction

Idiopathic pleuroparenchymal fibroelastosis (IPPFE) is a recently reported group of disorders, first described by Frankel et al. in 2004 [1]. The authors identified five cases of a unique idiopathic pleuroparenchymal lung disease characterized by fibrotic thickening of the pleural and subpleural parenchyma, predominantly in the upper lobes. Such clinicopathological entity did not fit any previously established category of idiopathic interstitial pneumonia. More cases have been reported later and share similar clinical, radiographic and pathologic features. Consequently, in the updated American Thoracic Society/European Respiratory Society classification of idiopathic interstitial pneumonias [2], IPPFE is included as a separate but well-defined category of rare entities. The authors report a new potential etiology of IPPFE in a female patient.

Case report

A 49-year-old female who is a never-smoker was referred to our hospital with a complaint of chest pain and dyspnea. She has worked in a refractory plant as a cremator for 9 years and had convincing evidences of industrial dust exposure. The patient had no known family history of pulmonary disease. Physical examination showed reduced breath sounds and fine crackles in both lower lobes. Serology tests were negative for autoantibodies, rapid plasma regain and rheumatoid factor. Lung function tests showed restrictive ventilatory defects.

Chest computed tomography (CT) revealed bilateral irregular pleural thickening mainly represented in the upper lobes (**Figure 1**). An open lung biopsy (Video-Assisted Thoracoscopic Surgery) was performed on the right upper lobe. Histopathology examination of the surgical lung biopsy specimen showed markedly thickened visceral pleura and prominent subpleural fibrosis characterized by abnormal increase of elastic tissue and dense collagen, abrupt transition to normal parenchyma was also seen (**Figure 2**). The clinical imaging and histological features of this patient were compatible with idiopathic pleuroparenchymal fibroelastosis (IPPFE).

Transmission electron microscopy (TEM) has been used to identify a few suspicious particles that were discovered through polarized optical microscope (POM). The TEM examination showed dust-like crystal structures in the lung

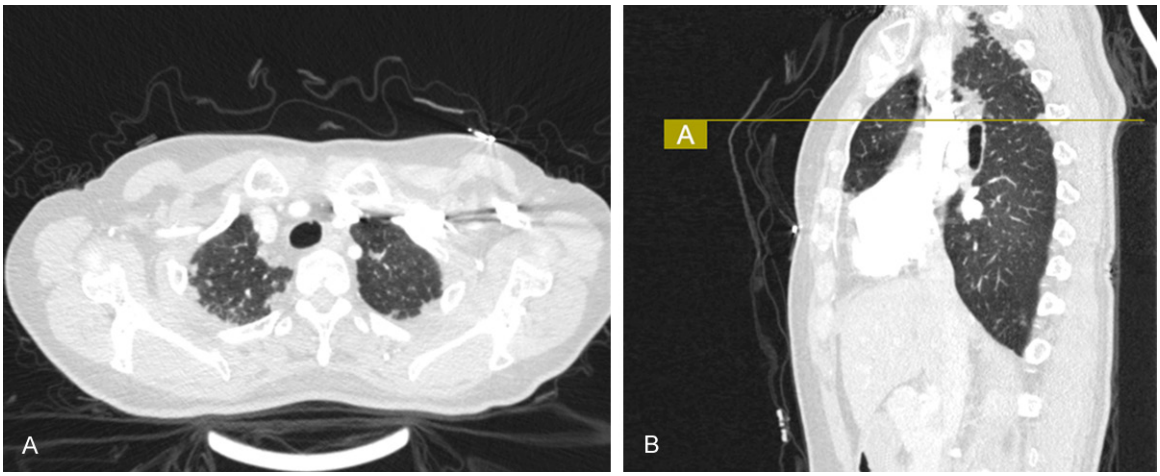

Figure 1. Chest CT scan showed: A. Bilateral pleural thickening and irregular pleural-based opacities. B. The radiographic changes mainly represented in the upper lobes. CT, computed tomography.

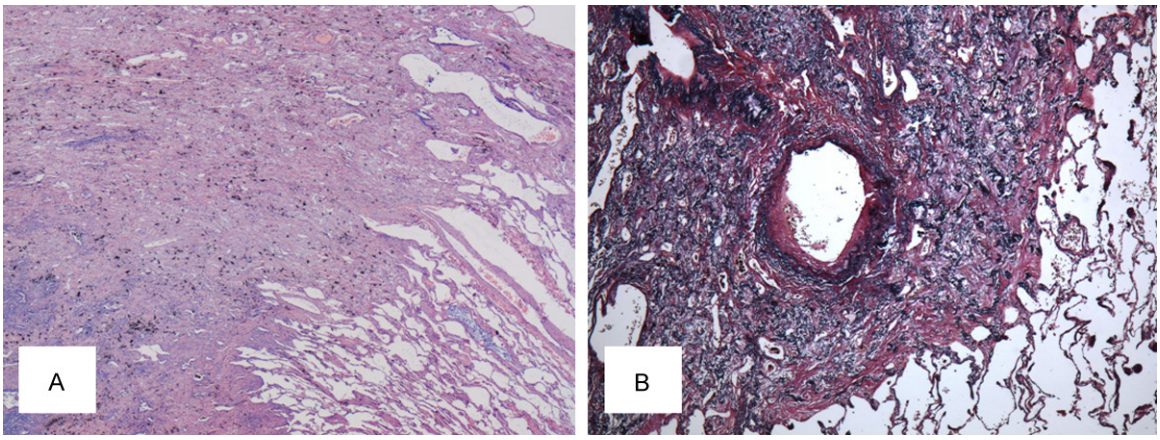

Figure 2. Histopathological findings in the surgical lung biopsy specimen. A. Hematoxylin-eosin, original ×20) showed fibrous thickened visceral pleura with an abrupt transition to normal lung parenchyma. B. Elastic fiber staining, original×40) demonstrated an abundance of short, amorphous elastic fibers in the thickened interstitium.

tissue (**Figure 3A**), and X-ray microelemental analysis revealed an abnormally high concentration of aluminum, silicon and oxygen (**Figure 3B**, **3C**). It can be speculated that these particles were inhalational dust, probably aluminosilicate (the main crude material for refractories industries), since the patient once worked in a refractory plant.

Discussion

We report a patient with well-documented dust exposure in which clinical presentation, imaging, and histopathological features are compatible with IPPFE. In this case, IPPFE may have been causally related to prior exposure to dust. Based on an extensive review of the literature, the case reported here suggests a new potential etiology of IPPFE.

Although there is insufficient knowledge about the etiology of IPPFE, several observations that may be of relevance to cause this condition have been reported. A few cases have underlying diseases or conditions such as bone-marrow transplantation [3, 4], lung transplantation [5] or antineoplastic therapy [1, 6]. Reddy et al. believed that repeated inflammatory damage in a predisposed individual have a role in its pathogenesis [7]. Frankel et al. reported two cases that were siblings revealed that genetic predisposition is probably another factor [1]. New data has been published suggesting that asinine herpesvirus may associate with some

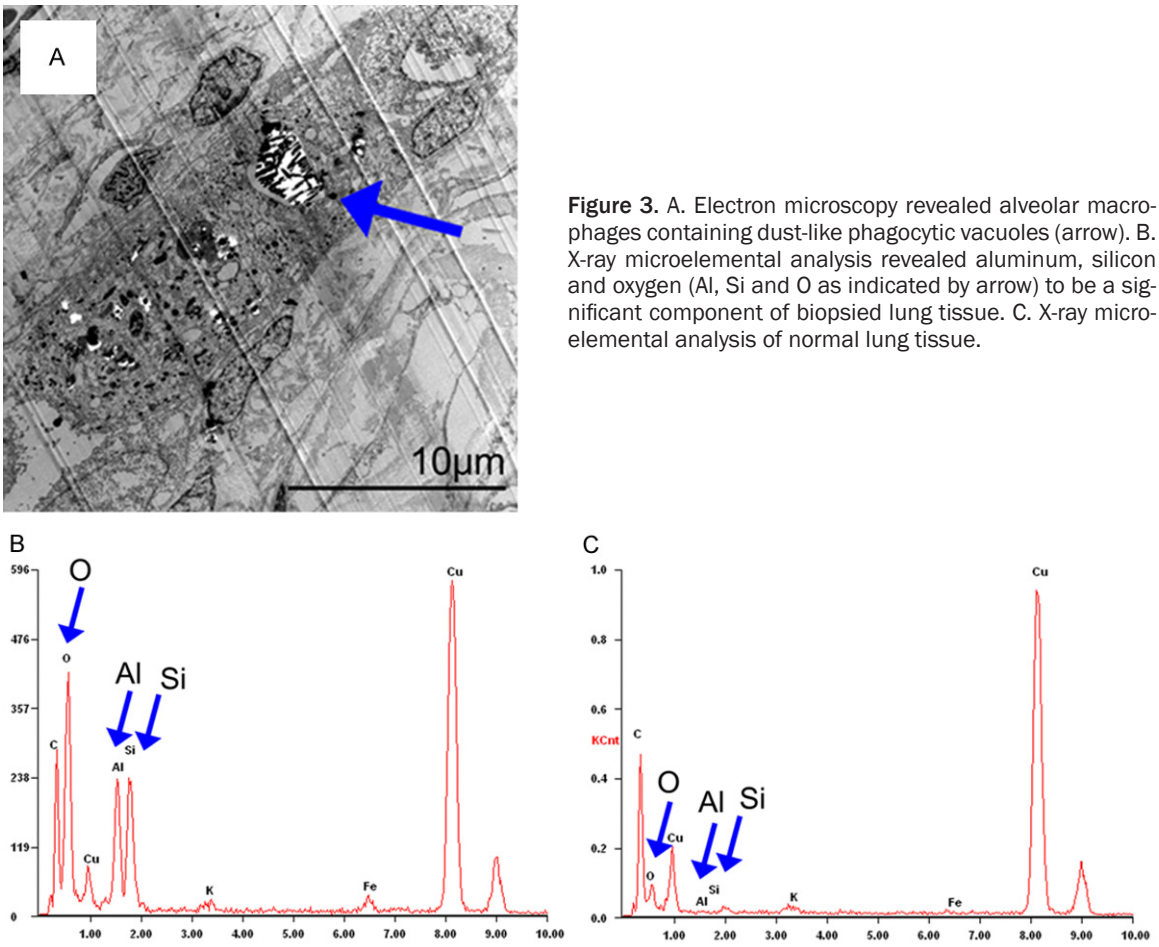

Figure 3. A. Electron microscopy revealed alveolar macrophages containing dust-like phagocytic vacuoles (arrow). B. X-ray microelemental analysis revealed aluminum, silicon and oxygen (Al, Si and O as indicated by arrow) to be a significant component of biopsied lung tissue. C. X-ray microelemental analysis of normal lung tissue.

donkey pulmonary fibrosis which was categorized as being consistent with IPPFE [8]. The potential role of dust exposure as a cause for IPPFE has not been discussed previously. Although two patients reported suspected asbestos exposure [9, 10], none of them offered convincing evidence of asbestos exposure, and the ferruginous bodies were universally absent.

In our patient, previous causes of IPPFE were ruled out according to her clinical presentation and laboratory findings. Based on the presence of dust-like crystal structures and the increased aluminum, silicon and oxygen in the lung biopsy specimen that compatible with the patient's history of occupational exposure to aluminosilicate, we feel it is reasonable to speculate that dust exposure should be considered as a possible causative factor in the pathophysiology of IPPFE.

Kang et al. and Jung et al. reported TGF-α increased in the mice stimulated by Asian sand dust which has similar composition of aluminosilicate [11, 12]. While TGF-α upregulation causes pleural and parenchymal fibrosis with markedly increased elastin expresses in a murine model [13, 14]. This gives us a hint that TGF-α may play an important role in the pathogenesis of dust-related IPPFE.

Conclusion

We report the first case of IPPFE probably attributable to dust exposure. Identification of the etiology can help us understand the disease mechanisms and put forward effective treatments since supportive care and ultimately lung transplantation are only options for these patients at present.

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Disclosure of conflict of interest

None.

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