Idiopathic pleuroparenchymal fibroelastosis presenting as bilateral spontaneous pneumothorax: A case report

Sir,

Pleuroparenchymal fibroelastosis (PPFE) is an under-recognized clinicopathological entity characterized by fibroelastosis of the pleura and subpleural lung parenchyma with striking upper lobe predominance.[1] Pneumothorax and pneumomediastinum complicate the course of the disease and often can be the initial presenting manifestation. To the best of our knowledge, this is the second case report of PPFE from the Indian subcontinent.[2] This is the first case

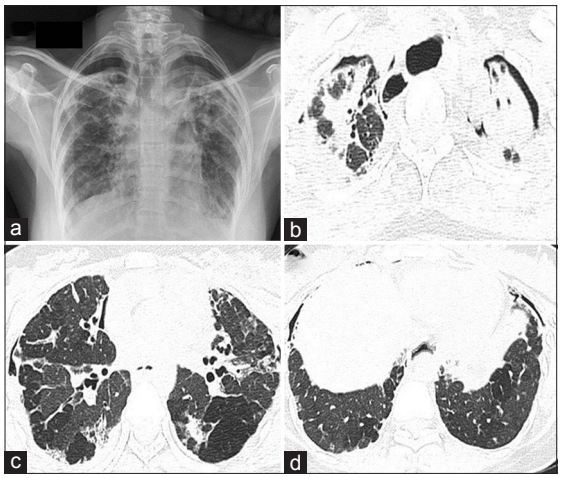

Figure 1: Chest radiograph showing bilateral pneumothoraces and bilateral apical fibrosis (a). High-resolution computed tomography of the chest showing bilateral pneumothorax and subpleural fibrosis and consolidation of the upper and middle lobes (b and c) with relative sparing of the lung bases (d)

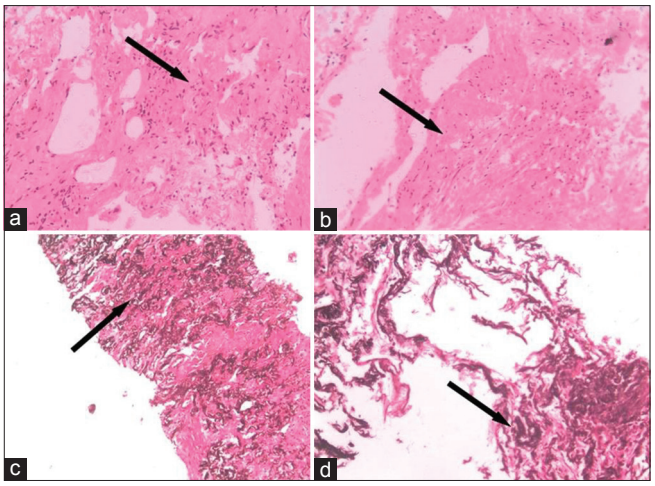

Figure 2: Photomicrograph of lung biopsy showing fibrocollagenous tissue (H and E, ×200) (a and b). Photomicrograph highlighting the presence of excessive elastin fibers in the same areas using the elastin van Gieson stain (c and d)

from our subcontinent where the diagnosis was established antemortem and treated successfully.

A 31-year-old female, a homemaker, presented with dry cough and progressively worsening dyspnea of 5 months duration in the postpartum period. She also lost 6 kg weight over the same period. Her history was insignificant for any connective tissue diseases. On auscultation, she had bilateral fine end inspiratory crackles more in the suprascapular areas. Arterial blood gas analysis was suggestive of Type I respiratory failure. Chest radiograph showed bilateral pneumothoraces and bilateral upper zone infiltrates [Figure 1a]. A high-resolution computed tomogram (CT) of the chest was done which revealed bilateral apical pneumothorax and pneumomediastinum along with bilateral apical fibrosis. There were also areas of subpleural consolidation and mosaic attenuation [Figure 1b-d]. A differential diagnosis of chronic hypersensitivity pneumonitis, sarcoidosis, tubercular sequelae-related lung disease, connective tissue-associated lung disease, drug-induced lung injury, and atypical interstitial lung disease were considered. A CT-guided biopsy was performed from the right upper lobe, which showed visceral pleural thickening with collagenous fibrosis, subpleural elastosis, and intra-alveolar collagenous fibrosis [Figure 2]. A diagnosis of idiopathic PPFE was made and she was initiated on systemic steroids (1 mg/kg prednisolone equivalent). Following the biopsy, there was worsening of the pneumothorax with persistent air leak which was managed with pigtail placement. Her arterial oxygen saturation gradually improved to 96% on room air and was discharged. On follow-up, as her symptoms worsened while tapering steroids, she was also started on mycophenolate mofetil as a steroid-sparing agent. One year since diagnosis, she continues to perform her daily activities and remains independent of oxygen support.

PPFE was first described in Japan by Amitani *et al*.[3] as idiopathic pulmonary upper lobe fibrosis. The findings consistent in their cases were (a) slender stature with flattened thoracic cage, (b) progressive subpleural fibrosis without honeycombing, (c) recurrent pneumothorax, (d) no extra thoracic lesions, and (e) absence of acid-fast bacilli and lack of response to antitubercular therapy. It had been called as Amitani disease until the term PPFE was coined by Frankel *et al*. in 2004.[1]

Although the etiology is unknown, most cases have shown association with lung, bone marrow, and hematopoietic cell transplantations, chemotherapy drugs, occupational exposures, and recurrent lower respiratory tract infections.[4] The common symptoms at presentation include dyspnea, dry cough, weight loss, and chest pain. Patients often have slender body habitus and a flat chest.[1] Spontaneous or iatrogenic pneumothoraces which are generally small and often recurrent and bilateral are common in the course of disease. The elastic pleura has limited healing capacity and this leads to persistent bronchopleural fistulae.[5] Earlier age of onset, low body mass index, presence of a flat chest, upper lobe predominance, high incidence of pneumothorax, and bronchopleural fistulae differentiate this entity from idiopathic pulmonary fibrosis.[4,6]

The unique pathologic feature of PPFE is intense, predominantly elastic fibrosis of the visceral pleura, particularly in the upper lobes. Marked elastin deposition within the areas of fibrosis, very few or rare fibroblastic foci, and homogeneous intra-alveolar fibrosis with preserved alveolar structure rather than temporal heterogeneity differentiate PPFE from usual interstitial pneumonia (UIP) pattern.[7] Both UIP and nonspecific interstitial pneumonia have subpleural-predominant interstitial fibrosis, which consists of more of collagen than elastic fibers, and have a lower lobe predilection unlike PPFE.[1]

Differential diagnoses include asbestos-related disease, advanced fibrosing sarcoidosis, connective tissue-associated disease, radiation- and/or drug-induced lung injury, and organizing pneumonia (OP). A relevant exposure history and absence of sarcoid granulomas and asbestos bodies on histopathological examination can differentiate PPFE from asbestosis and sarcoidosis. Involvement of lung bases and more peribronchial distribution rather than predominant subpleural and paraseptal contiguous areas of fibrosis differentiates OP from PPFE.[7]

PPFE is a rare form of interstitial lung disease, and differentiating this entity from other idiopathic interstitial pneumonias is of paramount importance to study the natural history and to guide the treatment regimen. Performing elastin fiber stains routinely in patients with radiological features suggestive of PPFE is recommended to establish diagnosis.[5] Clinicians should anticipate complications such as both spontaneous and secondary pneumothoraces during the management of this entity. Lung transplantation remains the only option for refractory disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/lungindia.lungindia_248_18

How to cite this article: Maturu VN, Rajendran P, Narahari NK. Idiopathic pleuroparenchymal ibroelastosis presenting as bilateral spontaneous pneumothorax: A case report. Lung India 2019;36:75-7.