disseminated forms of histoplasmosis usually occur in immunocompromised hosts.¹,² Lung biopsy and fungal culture has been widely recognized as the gold standards for diagnosing pulmonary histoplasmosis. Unlike with the literature, our patient was presented with large solid mass of the right lower lobe abutting the surrounding structure with extension toward left lung. The diagnosis of histoplasmosis was established by histopathological examination and special staining of tissue obtained by percutaneous CT-guided biopsy. The patient was put on antifungal treatment in the form of Itraconazole and shows clinical as well as radiological improvement. The case was more interesting because no factor responsible for immunosuppression could be demonstrated in the patient.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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There are no conflicts of interest.

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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
PPFE was first described in Japan by Amitani et al. 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.

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. The common symptoms at presentation include dyspnea, dry cough, weight loss, and chest pain. Patients often have slender body habitus and a flat chest. 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. 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.

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. 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.
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.[3] 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.

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