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CASE REPORT

Idiopathic pleuroparenchymal fibroelastosis (PPFE) – A case study of a rare entity

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Abstract Idiopathic pleuroparenchymal fibroelastosis (IPPFE) was recognized as a rare new entity. We report the case of a 63 years old female suffering from progressive dyspnea and dry cough for three years. Two years before admission to our hospital, idiopathic pulmonary fibrosis (IPF) was diagnosed in another hospital and treatment with prednisolone and N-acetylcysteine (NAC) was commenced. At admission HRCT showed upper lobe dominant fibrosis and associated pleural thickening. Surgical biopsies were re-evaluated and revealed fibroelastosis with pleural thickening and a probable UIP pattern, consistent with idiopathic PPFE. Treatment with pirfenidone was initiated due to progression under prednisolone and NAC. Upper lobe predominant pleural thickening with associated subpleural fibrotic changes should raise suspicion of PPFE.

Introduction

Pleuroparenchymal fibroelastosis (PPFE) is a rare entity, characterized by upper lobe dominant subpleural fibroelastosis and dense fibrous thickening of the visceral pleura. Most cases are idiopathic. Evidence based treatment options do not exist.[1] In the update of the international consensus classification of idiopathic interstitial pneumonia (IIP), idiopathic PPFE has been included within the group of rare IIP.[2,3] We here report a case of PPFE, which was initially misdiagnosed as IPF.

Case presentation

A 63 year old female patient presented with progressive dyspnea and dry cough for 3 years. There was no significant occupational exposure, allergy or smoking history. There was no family history of pulmonary fibrosis.

Two years before the admission to our hospital high-resolution computed tomography (HRCT) in a regional hospital had shown fibrotic changes of the lung parenchyma. Subsequently the patient underwent a video assisted thoracoscopic surgery (VATS) for histological assessment,

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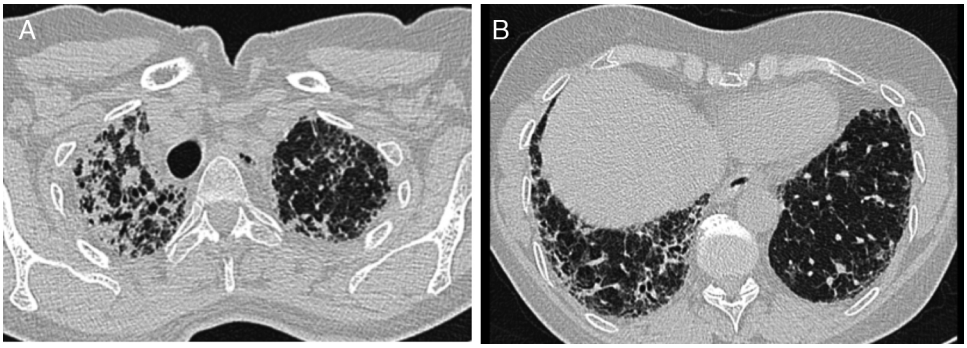

Figure 1 (A) Pleural thickening, dense fibrosis and consolidation in the upper lobes. (B) Traction bronchiectasis, reticular opacities and minor honeycombing in the lower lobes.

Table 1 Pulmonary function tests in follow up.

	2 years before admission	At admission	6 months follow up under pirfenidone	1 year follow up under pirfenidone
FVC [% pred.]	72	55	51	37
FEV1 [% pred.]	80	57	39	n.a.[a]
TLCO [% pred.]	46	35	25	n.a.[a]
paO_2 [mmHg]	82	67	68	65

[a] n.a. – not available.

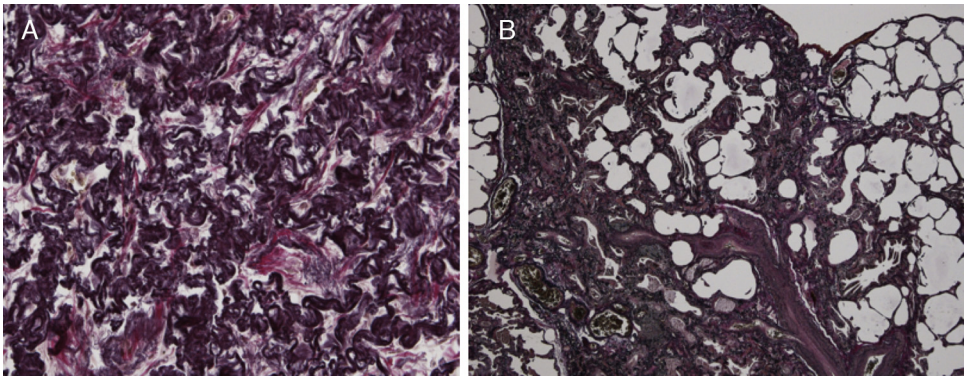

Figure 2 (A) The biopsy from the upper lobe showed severe fibrosis in the subpleural space with a prominent elastosis in the EvG stain; the adjacent pulmonary parenchyma revealed non-characteristic alveolar-septal fibrosis without honeycombing. (B) The biopsy from the lingular lobe revealed a patchy fibrosis with paraseptal and subpleural predominance. Honeycombing and mucus plugging were seen focally. Fibroblastic foci were not detected. Inflammatory infiltrates were scarcely developed. In some parts proliferation of smooth muscle cells was found. A mesh-like fibrosis was seen focally.

fibrotic changes were interpreted as normal interstitial pneumonia (UIP) pattern. A diagnosis of IPF was made and a treatment with prednisolone and NAC started.

HRCT performed in our hospital demonstrated upper lobe predominant pleural thickening and associated reticular opacities, traction bronchiectasis, subpleural and peribronchial consolidation. There was upper lobe shrinkage and architectural distortion (Fig. 1A). The lower lobes showed minor honeycombing (Fig. 1B). Bronchoalveolar lavage cell differential was normal. There was no clinical or serological evidence of a connective tissue disease, vasculitis or extrinsic allergic alveolitis. Lung function showed a significant decrease in forced vital capacity (FVC) (55% of predicted) in comparison to two years earlier (72% of predicted) (Table 1). Six-minute walking test demonstrated a walking distance of 475 m with significant desaturations (76%). Transthoracic echocardiography showed mild pulmonary hypertension with an estimated systolic pulmonary artery pressure of 35 mmHg.

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