Idiopathic pleuroparenchymal fibroelastosis
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DESCRIPTION
A woman in her 60s presented with chronic cough, dyspnea on exertion and progressive weight loss. She was a never smoker and had a history of recurrent pneumothorax (one bilateral, one right sided and one left sided). There was no history of exposure to organic dust. She had a slender stature with a flattened thoracic cage. At the time of initial examination, she was 154 cm tall and weighed 30.4 kg with a body mass index of 12.8. She had an oxygen saturation of 93% while breathing ambient air. Sounds on auscultation of the lungs were normal. Arterial blood gas analysis on ambient air showed hypoxemia with mild hypercapnia (PaO₂, 58 torr; PaCO₂, 49 torr; and pH, 7.46). Laboratory investigations revealed a normal Krebs von Lungen-6 level (216 U/mL; normal range of 0–449 U/mL) and a normal β-D glucan level. Antinuclear antibodies and rheumatoid factor were negative. Repeated sputum examinations were sterile without Mycobacterium or Aspergillus. Posteroanterior (figure 1A) and lateral chest radiography (figure 1B) showed bilateral upper lobe-predominant consolidations, a rightward tracheal deviation (arrowheads) and a flattened thoracic cage. Chest CT (figure 2A–C) revealed upper lobe-dominant, subpleural wedge-shaped consolidations, an upward shift of hilar structure and a mild pneumothorax (arrow). There were no mediastinal or hilar lymphadeno pathy. No underlying diseases of interstitial pneumonia were evident. She was diagnosed with idiopathic pleuroparenchymal fibroelastosis (PPFE) according to a multidisciplinary discussion.

Idiopathic PPFE is a rare subtype of idiopathic interstitial pneumonia, which is characterised by upper lobe-dominant pulmonary fibrosis with peculiar clinical features of recurrent pneumothorax, flattened thoracic cage and progressive emaciation.1 The disorder was named ‘pleuroparenchymal fibroelastosis’ because of its histopathological characteristics of increased elastic and collagen fibres in the subpleural area of the lungs with collagenous thickening of the visceral pleura.1

Idiopathic PPFE is usually progressive and results in restrictive ventilatory impairment. The prognosis of idiopathic PPFE is poor, with a 5-year survival rate of 23–58%.2 Some investigators have described a poorer prognosis of idiopathic PPFE than idiopathic pulmonary fibrosis.3 Antifibrotic agents including nintedanib and pirfenidone have been reported to be effective in a broad range of interstitial lung diseases. However, there is no established treatment for idiopathic PPFE. Clinicians should be aware of this interstitial pneumonia type, as it is sometimes mistaken for obsolete tuberculosis or residual scarring. Retrospective review or prospective investigation of imaging data is necessary to differentiate idiopathic pleuroparenchymal fibroelastosis from obsolete tuberculosis or residual scarring.

Patient’s perspective
I will be glad to be of help for future healthcare.

Learning points
- Idiopathic pleuroparenchymal fibroelastosis is a rare subtype of idiopathic interstitial pneumonia, which is characterised by upper lobe-dominant pulmonary fibrosis with peculiar clinical features of recurrent pneumothorax, flattened thoracic cage and progressive emaciation.
- Idiopathic pulmonary fibrosis is sometimes misdiagnosed as obsolete tuberculosis or residual scarring.
- Retrospective review or prospective investigation of imaging data is necessary to differentiate idiopathic pleuroparenchymal fibroelastosis from obsolete tuberculosis or residual scarring.
misdiagnosed as obsolete tuberculosis or pulmonary apical cap. The obsolete tuberculosis or pulmonary apical cap are localised and have quite slow or no progression, while PPFE is extensive and progressive. During one year of follow-up, the patient’s pulmonary lesions gradually progressed, and home oxygenation therapy was introduced.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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