

THE FISSURAL INVOLVEMENT IN PLEUROPARENCHYMAL FIBROELASTOSIS: SOMETHING BEYOND THE LUNG PARENCHYMAL ELASTOSIS

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To the editor,

A 58-year-old man, former smoker, with history of exposure to wood in a sawmill, came to our attention because of dyspnoea and dry cough occurring in the last six months.

The chest CT scan documented a diffuse interstitial lung disease characterized by pleuroparenchymal fibroelastosis in the upper lobes, associated with reticulation and traction bronchiectasis in the mid and lower lung zones (Figure 1, a-c). A significant thickening, with solid density, triangular in shape, was also present along the upper portion of the right major fissure (Figure 1, d-e). Scattered pulmonary ossifications were finally detectable in the context of lung fibrosis.

Due to the history of exposure, the patient underwent a transbronchial cryobiopsy.

Biopsies were carried out in the dorsal segment of the right upper lobe and in the anterior segment of the right lower lobe.

Soon after the end of the procedure, a right pneumothorax was documented, requiring insertion of a chest tube.

Due to the persistence of a moderate right pneumothorax and subcutaneous emphysema one week later, a new CT scan was performed. The fissural

lesion present in the prior CT scan decreased in size showing an airfluid level in its context (Figure 1, f).

The patient was subsequently placed in the prone position, and the lesion became a flat band connecting the parietal and visceral pleura (Figure 1, g and h).

The cryobiopsy samples from the right upper lobe confirmed a leaf of fibroelastotic tissue (6.5 x 4 mm) with margins covered partially by mesothelial cells; dilated lymphatics, smooth muscle cells and few alveolar structures were also present. In the samples from the lower lobe a typical UIP pattern was observed (Figure 1, I and l). The histopathologic aspects in the cryobiopsy samples, associated with the aspects of the pneumothorax in CT scan, confirmed that this band of tissue represented a fibroelastotic strip connecting the visceral to the parietal pleural.

Pleuroparenchymal fibroelastosis (PPFE) is a radiologic-pathologic entity characterized by fibrosis with upper lobe and subpleural predominance, involving both the visceral pleura and the subjacent subpleural lung parenchyma, and it's histologically characterized by elastin deposition in the alveolar septa, in the parenchymal interstitium and dense visceral pleural and subpleural elastotic fibrosis¹⁻⁴

An emerging concept is the relevance of the fissural involvement by this elastotic process.

In a recent paper, the fissural elastotic thickening has been demonstrated having a significant correlation with the PPFE score extent and with the fibrosis score in those cases in which UIP pattern was associated⁵.

The fissures (most involved were the right ones) were classically triangular shaped and, once measured, showed a correlation with PPFE extent and,

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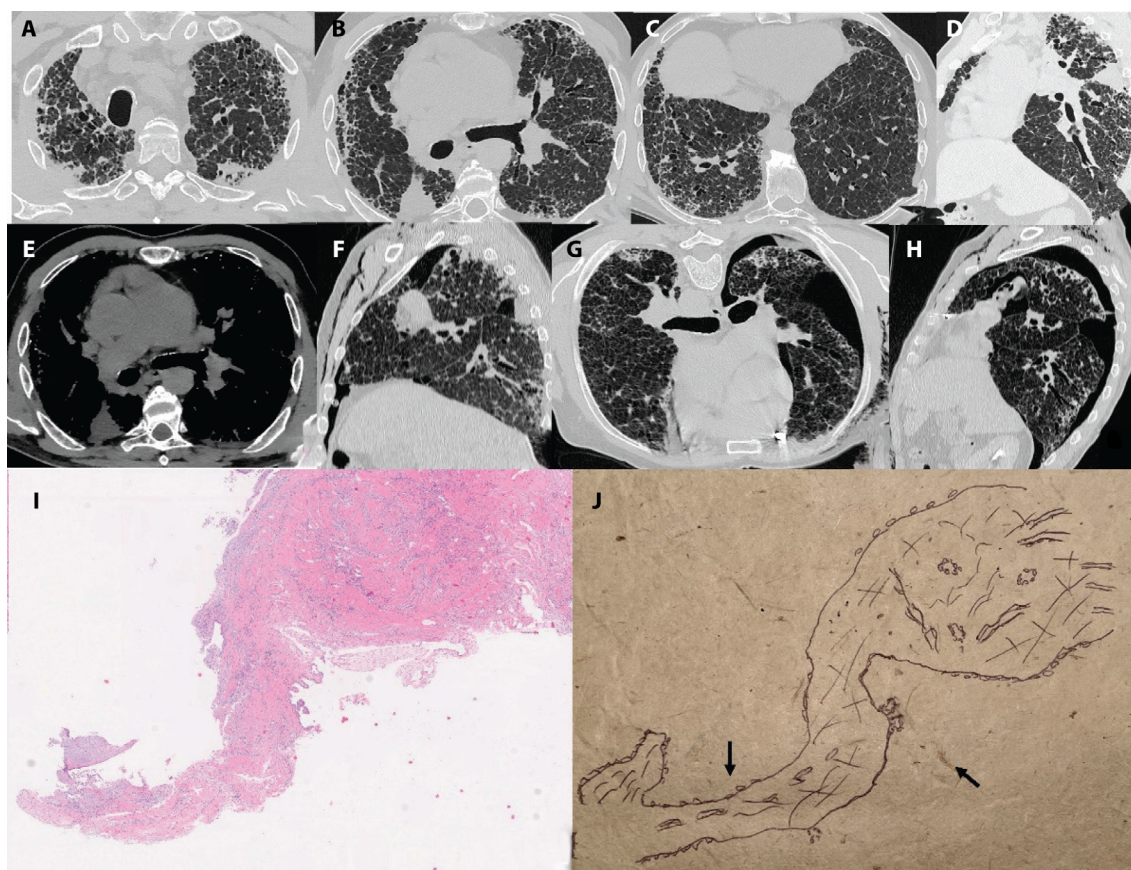


Figure 1. (a-e) CT scan before cryobiopsy. Fibrotic interstitial pneumonia characterized by severe pleural and subpleural fibrosis, traction bronchiectasis and pulmonary ossifications in both upper lobes. A severe fibrotic thickening of the fissures is visible bilaterally, more on the right side where it is triangular (d, sagittal view), and hypodense in the mediastinal window (e, red arrowhead). No pleural effusion is present. A marked right and posterior deviation of the trachea is also visible present (a). In both lower lobes peripheral reticulation, architectural distortion, traction bronchiectasis and foci of honeycombing are visible (d, orange circle), with prevalence for the right side. Of interest, a severe pulmonary artery dilatation is present (e). (f-h) Post-cryobiopsy CT scan: turning from the supine position (f, sagittal MPR reconstruction) to the prone position (g,h) the pleural thickening mimicks an air-fluid level (f) that changes position on the prone image and is seen as fibrotic band connecting the chest wall and the visceral pleura (g,h). No pleural effusion is present, as it's visible in the prone sagittal view (h). (i) Cryobiopsy samples from the right upper lobe showed a leaf of fibroelastotic tissue (6.5 x 4 mm) with margins covered partially by mesothelial cells; in the larger portion smooth muscle cells and few alveolar structures were also identifiable (H&E, x 10). (j) Ink on paper. Arrows indicating mesothelial cells partially covering the margins.

more interestingly, with fibrosis score of the associated ILD.

Based on the aspects herein described, the fissural involvement suggests that the fibroelastotic tissue could be accumulated, at least in some extent in the parietal pleural and not only in the visceral one as suggested so far.

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