

Pleuroparenchymal Fibroelastosis: A Review of Histopathological Features

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Purpose of the study: Pleuroparenchymal Fibroelastosis (PPFE) is a rare disease recognised in the revised 2013 ATS/ERS classification of idiopathic interstitial pneumonias, with major histopathological features being predominantly subpleural intra-alveolar fibrosis and elastosis (IAFE), and visceral pleural fibrosis. This recognition has led to a significant increase in cases presenting at our institution and we have therefore reviewed this cohort to assess proposed histological criteria (IAFE and visceral pleural fibrosis), as well the incidence of coexistent individual histological features and patterns.

Methods: 41 cases of PPFE were reviewed, 12 cases having been previously reported. Each case was scored semi-quantitatively as (mild (% cases)/moderate (% cases)/severe (% cases)) for IAFE, visceral pleural fibrosis, vascular changes, inflammation and fibroblastic proliferation, and present/absent for granulomas. Coexistent histological patterns were also documented.

Summary of results: All cases showed IAFE (17%/27%/56%), associated inflammation (14%/54%/32%) and fibroblastic proliferation (41%/39%/20%) with 93% showing fibro-intimal vascular thickening (17%/49%/27%) and 65% showing visceral pleural fibrosis (27%/19%/19%). Granulomas were seen in 29% of cases, with a histological pattern of hypersensitivity pneumonitis seen in 15% and a pattern of usual interstitial pneumonia in 10%. One aspergilloma and one case of coexistent Wegener's granulomatosis were also seen.

Conclusions: Whilst IAFE, associated inflammation and fibroblastic proliferation were always present in PPFE, visceral pleural fibrosis was less frequently seen. Vascular changes are prominent and there is wide variation in the extent of all of these features, which may reflect potential aetiologies and could provide prognostic data. Coexistent histologic patterns are also not uncommon.