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# Coexistence of Pleuroparenchymal Fibroelastosis and Hypersensitivity Pneumonitis: Case Report

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Pleuroparenchymal fibroelastosis (PPFE) is a rare fibrosing lung disease, included as a distinct clinicopathologic entity in the latest international multidisciplinary classification of the idiopathic interstitial pneumonias. The etiology of PPFE is unclear at this juncture, with many cases being considered as idiopathic forms of the disease. However non-idiopathic PPFE has been increasingly reported in association with several interstitial lung diseases including idiopathic pulmonary fibrosis, hypersensitivity pneumonitis (HP) and familial forms of pulmonary fibrosis. Some of its clinical features are dyspnea and dry cough, similar to other interstitial pneumonias. Radiologically, it is mostly characterized by pleural and subpleural parenchymal fibrosis in the upper lobes. Pathologically, PPFE is characterized with intra alveolar dense fibrosis with prominent elastosis in alveolar walls, dense fibrous thickening of visceral pleura. Here, we present a more rare case with co-existence of PPFE with HP. A thirty seven year old male patient who had been working as an operating room staff in the hospital, presented to our clinic with a shortness of breath that had gradually worsened by exercise and a year history of cough. He had a history of pneumonia 20 years ago. He noted having a pet pigeon when he was young. He has no other past medical history. In physical examination pectus excavatum deformity was observed and the antero-posterior diameter of chest was decreased. Breath sound was diminished in both lung fields. Chest computed tomography revealed bilateral air bronchogram and consolidations in the upper lobes. Pleural thickening especially in apical subpleural regions and bullous emphysema was observed. Laboratory findings were normal. Review of pulmonary function testing shows severe obstruction and reduction in diffusing capacity. He was consulted with rheumatology for differential diagnosis, and connective tissue disease was not considered. Bronchoscopy was performed and bronchial lavage and transbronchial biopsy results were non diagnostic. Video-Assisted Thoracoscopic Surgery was performed. Histological features revealed the characteristic findings of PPFE with marked elastosis and alveolar-septal fibrosis and HP. PPFE is a rare idiopathic interstitial pneumonia, characterized by different clinical, radiological and pathological features. PPFE was identified in 23% of HP patients and was independently associated with a reduced lung capacity and diffusing capacity. In our case, PPFE was assumed to be associated with hypersensitivity pneumonitis.

**Keywords:** Pleuroparenchymal fibroelastosis, hypersensitivity pneumonitis, rare interstitial lung disease