Pleuroparenchymal fibroelastosis (PPFE) is a rare condition characterised by predominantly upper lobe pleural and subjacent parenchymal fibrosis, the latter being intra-alveolar with accompanying elastosis of the alveolar walls. We report the case of a 71-year-old woman, ex-smoker, who had a left lower lobectomy 5 years ago for a Stage One adenocarcinoma. In the following years, she started complaining of worsening breathlessness, weight loss and had on average, 12 lower respiratory tract infections a year. She was on no medication, had no significant family history and had no occupational exposure to chemicals. Positive clinical findings on examination revealed that she was hypoxic and had bibasal crackles. Her pulmonary function test showed a restrictive pattern with a low transfer factor. Surveillance CT thorax discussed at the lung multidisciplinary team showed features of pleuroparenchymal fibroelastosis with bronchiectatic changes that was confirmed on CT core biopsy of her lung. Following her diagnosis, she had multiple hospital admissions for her repeated chest infection. Her last CT thorax done showed that bilateral subpleural peripheral reticulation, traction bronchiectasis and volume loss involving all lobes due to fibrosis from known diagnosis of pleural-parenchymal fibroelastosis were stable. However there was interval development of extensive mass like right apical nodular pleural thickening but there was no evidence of metastatic disease in relation to previous left lower lobectomy for adenocarcinoma. Furthermore interval development of multiple bilateral loculated pneumothoraces were seen. The patient was managed conservatively with oxygen, intravenous antibiotics, and chest physiotherapy.

PPFE is a distinct clinicopathological entity, with clinical data suggesting a link to recurrent pulmonary infection. Genetic and autoimmune mechanisms may also contribute to the development of these changes. Going through the literature reviews, the data of 12 patients with PPFE were analysed. Of the 12 patients (seven females, median age 57 yrs), the presenting symptoms were shortness of breath (11 out of 12 patients) and dry cough (six out of 12 patients). Seven patients reported recurrent infections during the course of their disease. Five demonstrated nonspecific autoantibody positivity. Two patients had a family history of interstitial lung disease. Of the 10 patients with follow-up data, seven demonstrated disease progression. Five of these patients died, with the time interval between diagnosis of PPFE and death ranging from 4 months to 2 years. Four of these five patients reported recurrent infections during the course of their disease. One of the five patients tested positive for Aspergillus sp. Four of these patients also had positive autoantibody screens. Treatment administered to these patients was highly variable and largely empirical, reflecting the lack of experience in treating patients with PPFE. The clinical course was progressive in many of these patients, a finding concordant with previous studies, despite aggressive treatment in some cases, including corticosteroids and immunosuppressants.

The dilemma in our case was a lack of evidence based medicine regarding the management of the
Therefore more clinical trials are required in order to have a better understanding of the pathophysiology and treatment of this condition.

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Reference

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