

Case Report

A Rare Manifestation of Asbestos Induced Lung Disease

D McErlean, B Praveen & E Reid

Dane McErlean
Stepping Hill Hospital

Bhatia Praveen
Consultant Respiratory
Physician
Stepping Hill Hospital

Emma Reid
Consultant Radiologist
Stepping Hill Hospital

Correspondence
Praveen Bhatia
Consultant Respiratory and
General Physician
Stepping Hill Hospital
Stockport
SK2 7JE

Abstract

Pleuroparenchymal fibroelastosis (PPFE) is a rare pulmonary condition that is often clinically characterised by lung upper-lobe predominant fibrosis. This article describes a case of a gentleman with shortness of breath over years who has subsequently been diagnosed with PPFE.

A 79 years old gentleman is referred to the chest clinic with progressive breathlessness and cough of a few years' duration. He has never smoked, but has worked with asbestos in the past. He has never kept any birds as pets. He has a past medical history of aortic valve replacement and hiatus hernia. His medications are bisoprolol, a statin, salbutamol inhaler, a DOAC and amlodipine. His pulmonary function tests are as follows

- FEV1= 1.39=51%
- FVC= 1.67= 45%
- Ratio= 110%
- TLCO= 3.82= 49%
- KCO=1.30=99%
- TLC=3.02=45%
- RV=1.81= 49%

He had a chest x-ray and CT thorax.

Asbestos is a naturally occurring fibrous silicate mineral. It was widely used in the building industry as it an electrical insulator and fire retardant, before its harmful effects were recognised. It was in the 1970s that its adverse effects on health were recognised and since then it has been banned in various parts of the world. This article will focus on asbestos related lung disease with particular reference to the rare PPFE.

Pleuroparenchymal fibroelastosis (PPFE) is a rare pulmonary condition that is often clinically characterised by lung upper-lobe predominant fibrosis. Patients typically present with exertional shortness of breath and/ or dry cough and the clinical course is slow, much like idiopathic pulmonary fibrosis. However one of the classic differences clinically between PPFE and IPF is the upper lobe predominance associated with PPFE, compared with a lower lobe predominance associated with IPF.

The histopathology of PPFE is characteristic and classically described as visceral pleural thickening with collagenous fibrosis, subpleural elastosis, and intraalveolar collagenous fibrosis. While PPFE is a rare condition, the literature has reported many different conditions over the past few decades of simply upper lobe predominant fibrosis. Commonly cited causes of upper lobe pulmonary fibrosis include fibrosis associated with autoimmune conditions; rheumatoid arthritis, ulcerative colitis, ankylosing spondylitis, radiation, TB, aspergillus, chemotherapy, stem cell or lung transplant, as a feature of graft versus host disease, and occupational exposure such as silicosis and of course asbestos, as we have seen with the patient discussed. As the interstitial connective tissue response in asbestosis is *fibroelastotic* rather than fibrotic, with pleural thickening, it is proposed that asbestos exposure directly induces the pathology of PPFE. However with infective causes of upper lobe lung fibrosis such as TB or aspergillus, it is unknown if these pathogens may induce PPFE, or if pathological lungs with PPFE provide an environment more susceptible for such infections. Clinical examination of patients with PPFE is reported to be less abnormal than such patients with IPF, namely finger clubbing is rarely reported and chest crackles on auscultation is also rarely reported for patients who go on to be diagnosed with PPFE.

Autoimmunity and genetic predisposition are both considered contributing factors in the development of PPFE as mentioned. However studies trying to pinpoint specific causes of this condition often conclude that the development of PPFE is likely multifactorial. Nonetheless it is widely accepted that asbestos exposure is a contributing factor.

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I draw your attention to Figures 1-3, images of the discussed patient

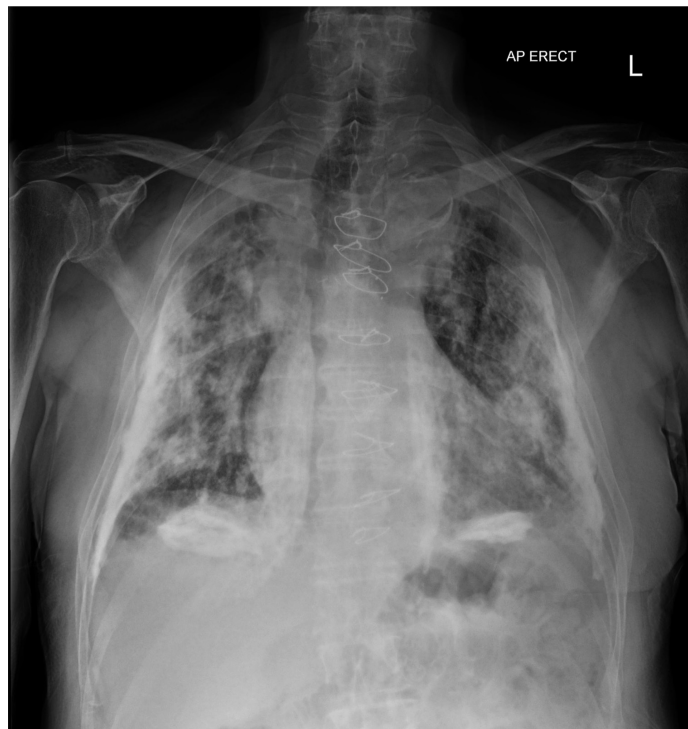


Figure 1. Plain chest radiograph showing extent of the pleural plaques as well as upper lobe volume loss

Management

This patient has subsequently been referred to local specialist ILD clinic and pulmonary rehab. Financial compensation may be available for asbestos related lung disease via council or government schemes. Idiopathic PPFE is slowly progressing and can be difficult to treat, often proving resistant to corticosteroids or other immunosuppressants. Supportive therapy such as long term O₂ and pulmonary rehab can prove beneficial. Furthermore, treatment of infections including *Aspergillus* is very important to prevent worsening of the PPFE.

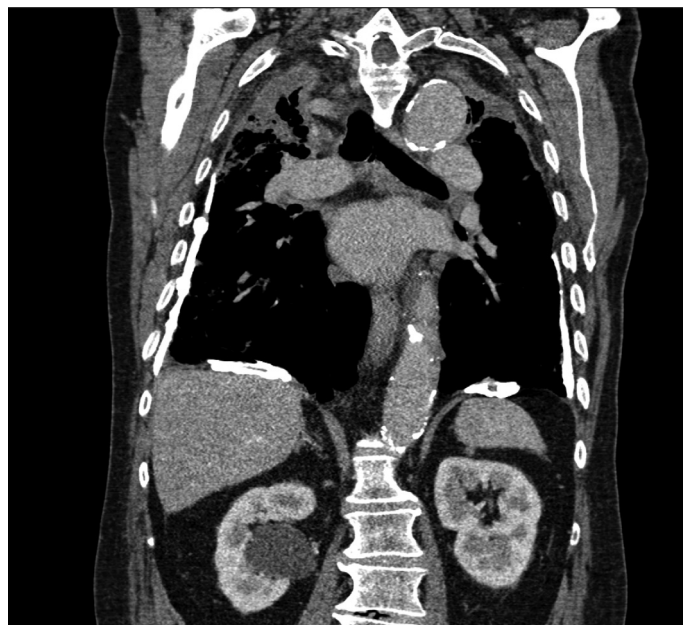


Figure 2. CT thorax scan showing extensive calcified plaques as well as upper lobe fibrosis and volume loss

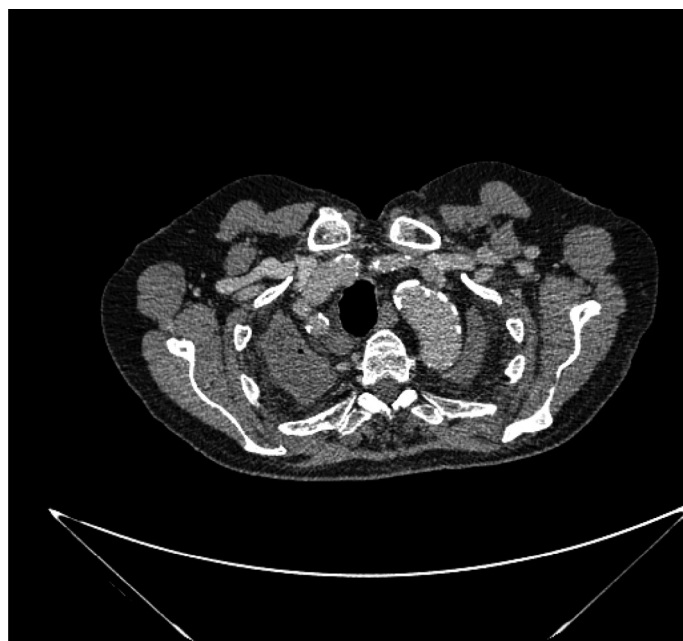


Figure 3. CT thorax showing platythorax and deep suprasternal notch -one of the key findings in PPFE. In this feature, you get posterior retraction of the trachea such that the posterior wall of the trachea is behind the level of the vertebral body as well as drawing in of the suprasternal notch. Platythorax is a morphological descriptor for the variation in chest shape where there is a flat chest with a comparative reduction in the AP diameter when compared with the lateral diameter

References

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