

## Case report

# Severe and progressive platythorax disproportionate to lung fibrosis: A rare variant of idiopathic pleuroparenchymal fibroelastosis

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## ABSTRACT

A 57-year-old man was referred to our department because of progressive shortness of breath and emaciation. He had experienced pneumothorax three times in the past five years. The patient radiologically showed mild upper-lobe predominant airspace consolidation and severe platythorax and was clinically diagnosed with idiopathic pleuroparenchymal fibroelastosis (PPFE). Although the wedge-shaped shadows in the bilateral lung apices did not significantly progress, his platythorax gradually worsened during the clinical course. He ultimately died of chronic respiratory failure 1.2 years after the diagnosis. This case demonstrates a rare variant of idiopathic PPFE with progressive platythorax disproportionate to the extent of upper-lobe fibroelastosis.

## 1. Introduction

Pleuroparenchymal fibroelastosis (PPFE) is a chronic fibrosing interstitial pneumonia with predominant fibroelastosis in the bilateral upper lobe [1–5]. PPFE is categorized as either idiopathic or secondary due to an underlying disease [6]. PPFE is radiologically characterized by upper-lobe-predominant airspace consolidation and platythorax, but both usually worsen as the disease progresses [1,7,8].

We herein report a patient with idiopathic PPFE who died of chronic respiratory failure, probably due to severe and progressive platythorax disproportionate to the extent of upper-lobe fibroelastosis.

## 2. Case report

A 57-year-old man presented to our department complaining of progressive shortness of breath for the past 6 months. He was an ex-smoker with a 30-pack-year for 15 years and had no abnormalities at birth and throughout development. His weight gradually decreased from 50 kg (51 years old) to 34.5 kg (57 years old). There was no respiratory or neuromuscular disease in his family history. He had experienced pneumothorax three times over the past five years.

At the time of the initial visit, his body mass index (BMI) was 12.4 kg/m<sup>2</sup> (166 cm tall and 34.5 kg weighed). There were no abnormalities in his vital signs and chest auscultation. Chest X-ray showed the upward shift of hilar structures (Fig. 1A). Chest computed tomography (CT) showed multiple subpleural wedge-shaped airspace consolidations at

the apex of both lungs, but the lesions were relatively mild and limited to the upper lobes (Fig. 1B and C). No fibrous lesions were found in the lower lobes (Fig. 1D and E). The thoracic cage was flattened with a flat chest index of 0.50 (reference value in IPF:  $0.649 \pm 0.006$  [9]), which was measured as the anteroposterior diameter of the thoracic cage/transverse diameter of the thoracic cage at the level of the 6th thoracic vertebra (Fig. 1D). On chest X-ray and CT, the platythorax and elevation of the hilar structures gradually worsened throughout the clinical course of the disease (Fig. 2), while the wedge-shaped shadows in the bilateral lung apices did not progress significantly (Fig. 2).

The laboratory data showed Krebs von den Lungen-6 (KL-6) of 214 U/mL (normal value < 500 U/mL), surfactant protein-D of 80.4 ng/mL (normal value < 110.0 ng/mL), rheumatoid factor of 7 IU/mL (normal value < 15 U/mL), antinuclear antibody of <x40, anti-aminoacyl-tRNA synthetase antibody index of <5.0 index (normal value < 25.0 index), aldosterone of 292 pg/mL (29.9 < normal value < 159 pg/mL), renin activity of 1.1 ng/mL/h (0.3 < normal value < 2.9 ng/mL/h), adrenaline of 9 pg/mL (normal value < 100 pg/mL), noradrenaline of 923 pg/mL (100 < normal value < 450 pg/mL), dopamine of 26 pg/mL (normal value < 20 pg/mL), thyroid stimulating hormone of 1.135  $\mu$ IU/mL (0.5 < normal value < 4.8  $\mu$ IU/mL), and free thyroxine of 1.52 ng/dL (0.83 < normal value < 1.77  $\mu$ ng/dL). An arterial blood gas analysis showed a pH of 7.350, PaO<sub>2</sub> of 91.0 Torr, PaCO<sub>2</sub> of 44.0 Torr, and HCO<sub>3</sub><sup>-</sup> of 24.3 mmol/L in room air. The results of respiratory function testing were as follows: forced vital capacity 1190 mL (30.8%), forced expiratory volume in 1 s 1190 mL (36.6%), FEV<sub>1</sub>/FVC 100%, functional residual

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capacity 3800 mL (116%), total lung capacity (TLC) 4560 mL (80.6%), residual volume (RV) 3380 mL (182%), RV/TLC 74.1% (199%), and diffusion capacity of the lung for carbon monoxide 7.58 mL/min/mmHg (54.8%); these findings indicated marked restrictive ventilatory dysfunction with an increased RV/TLC.

Since the patient showed mild muscle weakness in the extremities and severe emaciation, we suspected amyotrophic lateral sclerosis or myotonic dystrophy as a differential diagnosis and performed evoked electromyography. However, no obvious neurogenic changes were evident. Myositis was ruled out by the laboratory data and physical examinations. We also considered endocrine disorders, including hypothyroidism, primary aldosteronism, and pheochromocytoma as a differential diagnosis of emaciation, but the laboratory data and radiologic examinations did not show any abnormal thyroid function or obvious adrenal tumors. After ruling out possible diseases, we concluded that the muscle weakness and emaciation were related to lung disease.

We clinically diagnosed the patient with idiopathic PPFE because of the presence of emaciation, chronic respiratory symptoms, recurrent pneumothorax, upper-lobe-predominant fibrotic lesions, platythorax, and restrictive ventilatory dysfunction with an increased RV/TLC [9, 10]. We were unable to perform a surgical lung biopsy because of the high risk of intractable pneumothorax. Since the patient had severe restrictive ventilatory failure, the indication for lung transplantation was discussed. However, the patient was not considered to be a candidate for a lung transplant because his lung lesions were relatively mild, and lung transplantation would not improve his respiratory function.

Even though the lung lesions were mild and did not progress much after the diagnosis, the flat chest index and BMI decreased over time (Fig. 2D). Non-invasive mechanical ventilation was started a year after the diagnosis because of the retraction of CO<sub>2</sub> in an arterial blood gas

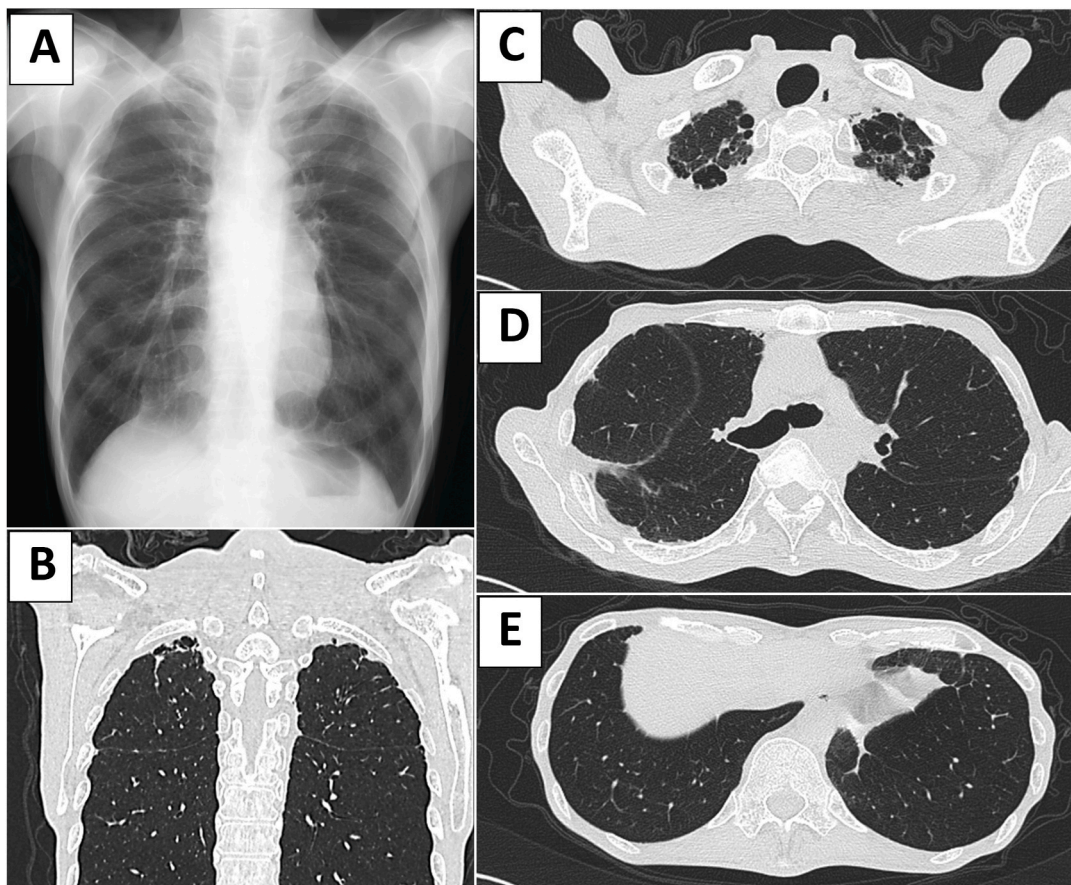
analysis (PaCO<sub>2</sub> 60.0 Torr) but was discontinued because of the incidence of pneumothorax. He ultimately died of chronic respiratory failure 1.2 years after the diagnosis.

### 3. Discussion

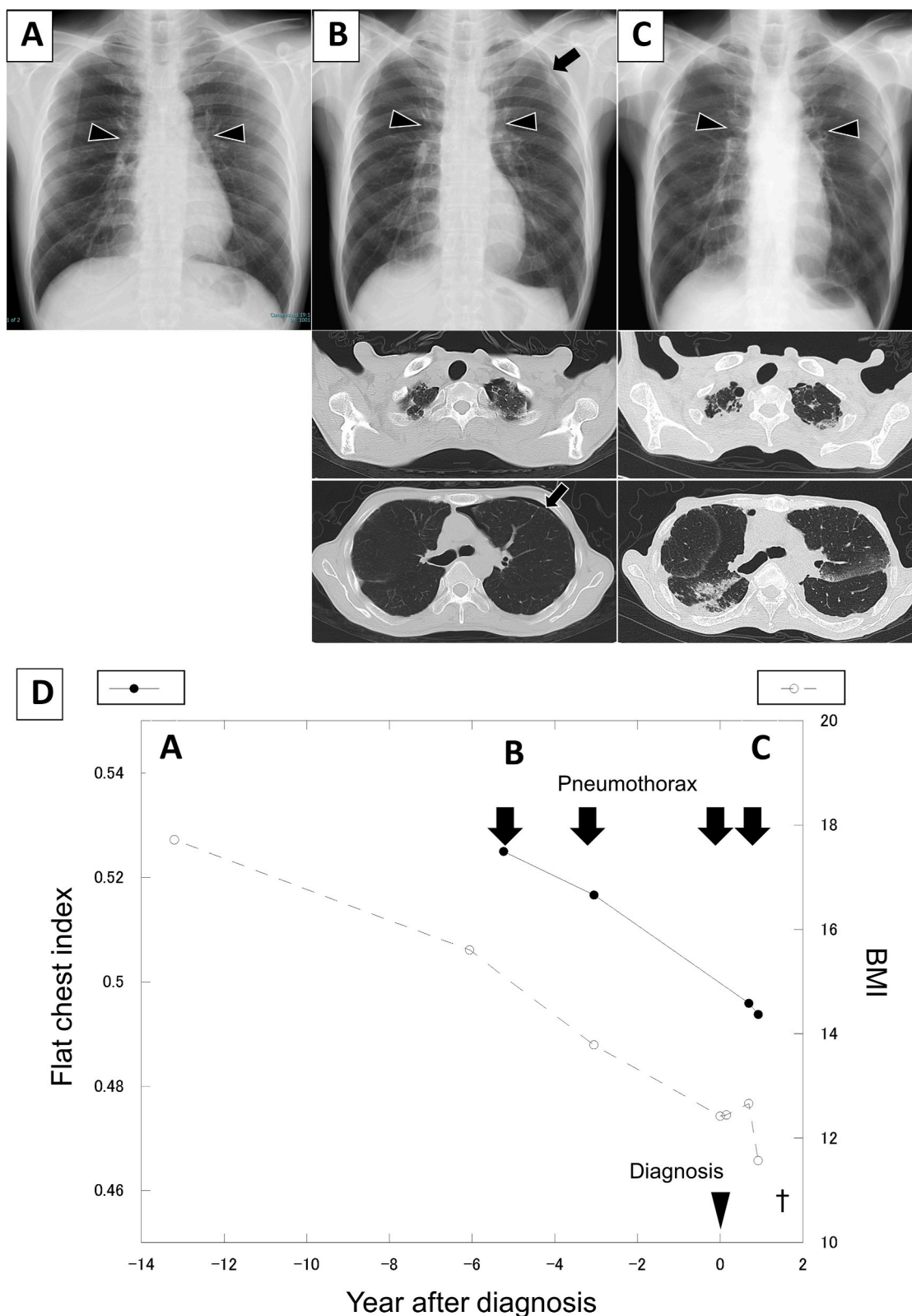
We herein report a patient with idiopathic PPFE who died of progressive respiratory failure, probably due to severe and progressive platythorax disproportionate to the extent of upper-lobe fibroelastosis. PPFE has two pathophysiologic aspects: upper-lobe-predominant fibroelastosis and platythorax (or flat chest) [1,7,10]. In patients with PPFE, which of the two is the primary event is debatable, as they usually progress simultaneously. In the present case, although our patient had upper-lobe-localized fibrosis, it was difficult to explain the progressive respiratory impairment by the pulmonary fibrosis alone, and his platythorax was deemed to be mainly associated with the progressive respiratory impairment.

Platythorax might arise secondary to fibrotic shrinkage of the upper lung lobes [1,8]. Harada et al. showed that platythorax in PPFE is often acquired and progresses during the clinical course [8]. They hypothesized that the thoracic cage was forced inward by long-standing fibrosis and gradual shrinkage of the lungs, resulting in progressive flattening of the thoracic cage [8]. However, platythorax might be the initial event prior to upper-lobe lesions of PPFE [1,7]. We previously demonstrated that early fibroelastosis lesions in idiopathic PPFE can start with subpleural zonal elastosis consisting of alveolar collapse [11]. A narrowed thoracic space in PPFE can prevent the lungs from fully expanding, possibly resulting in the collapse of subpleural alveoli [11,12].

The restriction of lung expansion due to thoracic cage deformity can cause impaired emptying of the lungs and restricted respiratory



**Fig. 1.** Chest X-ray (A) and computed tomography (B–E) at the diagnosis of pleuroparenchymal fibroelastosis showing mild wedge-shaped consolidations in bilateral lung apices (A–C), overinflation of the lower lobes (A), and a flattened chest cage (D) with elevated hilar structures (A). Lower lobes fibrosis was not evident (E).



**Fig. 2.** Chest X-ray and computed tomography findings from 13 years before the diagnosis (A), 6 years before the diagnosis (B), and 1 year after the diagnosis (C). The hilar structures (arrowheads) on chest radiography were gradually elevated throughout the disease course (B and C). While the wedge-shaped shadows in the bilateral lung apices did not change significantly, the chest cage gradually became flattened on chest computed tomography (flat chest index of 0.52 in B and 0.49 in C). Pneumothorax in the left lung was evident (arrows). The chest cage was flattened, and the body mass index (BMI) decreased over the course of the disease (D). † indicates death.

impairment, regardless of the presence or absence of lung fibrosis [13]. Clay et al. defined this type of restrictive respiratory impairment as a “complex restrictive pattern” and distinguished it from a “simple restrictive pattern” due to pulmonary diseases such as interstitial pneumonia [13]. In the present case, the patient’s physique was normal by nature, so platythorax was presumed to have been acquired. Acquired chest wall deformities account for less than 1% of all chest wall deformities [14]. Three major types of acquired chest wall deformities have been described based on their etiology: i) primary chest wall disease due to infection or tumor invasion, ii) iatrogenic deformities due to prior surgery, and iii) post-traumatic deformities [14]. Ankylosing spondylitis or kyphoscoliosis may also result in an impaired expansion of the thoracic cage [14–16]. Ankylosing spondylitis is sometimes associated with upper-lobe fibrosis or atelectasis [15,17]. However, the present patient had no underlying disease that might cause platythorax, as described above.

The upward shift of hilar structures is a characteristic radiologic finding in PPFE [18]. It is also interesting that the lesions at the lung apex were mild but accompanied by progressive elevation of the hilar structures. The upward shift of the hilar structures is presumed to be associated with progressive contraction of the upper lung lobes [18]. Why the hilar structures were elevated disproportionately to the degree of lung involvement is also unclear. In the present case, we may assume that platythorax and the upward shift of the hilar structures progressed regardless of the lung lesion.

Recently, diagnostic criteria for idiopathic PPFE have been proposed [9]. Because a tissue biopsy is currently avoided due to concerns about a prolonged period of post operative iatrogenic pneumothorax in patients with PPFE [19,20], clinical diagnostic criteria that do not depend on a pathological diagnosis have been included in the proposal [9]. Although the pathology could not be examined in the present case, the patient met all of the clinical diagnostic criteria for idiopathic PPFE. After 13 years of follow-up, the patient presented with restrictive respiratory impairment and ultimately died of chronic respiratory failure, probably due to progressive platythorax. This report describes a new variant or rare phenotype of PPFE in which platythorax is noticeable, but upper-lobe fibrosis is inconspicuous. This is an important case for understanding the significance of platythorax in PPFE.

#### Declaration of competing interest

The authors have no conflicts of interest directly relevant to the content of this article.

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