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Case report

Emphysema formation in a never-smoker with scleroderma-related interstitial pneumonia

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ABSTRACT

We report a case of pathological emphysema formation in a never-smoker with scleroderma-related interstitial pneumonia. We realized the importance of pulmonary vasculopathy causing the coexistence of emphysema and fibrosis in scleroderma. Our discovery provides novel information that emphysema can occur in patients with scleroderma-related interstitial pneumonia who have never smoked.

1. Introduction

Although emphysema combined with systemic sclerosis-associated interstitial lung disease (SSc-ILD) is present in 7.5% of never-smokers [1], the mechanism of emphysema formation in patients with SSc-ILD has never been clarified.

We herein report a case report of the coexistence of emphysema and fibrosis in a patient with SSc.

2. Case presentation

A 64-year-old male never-smoker without significant environmental or occupational exposure complained of cough and dyspnea for 6 months. He was diagnosed as having SSc from physical examination findings of Raynaud's phenomenon, diffuse skin hardening, and positive serum anti-scleroderma-70 antibody test. Alpha-1 antitrypsin deficiency was excluded. Chest computed tomography revealed traction bronchiectasis, reticulation, and ground-glass opacities (Fig. 1A). A mean pulmonary artery pressure of 32 mmHg on right heart catheterization indicated pulmonary arterial hypertension (PAH). Surgical lung biopsies revealed a fibrosing pattern indicative of nonspecific interstitial pneumonia (Fig. 1B), prominent splitting of the secondary alveolar walls (i.e., pulmonary emphysema) (Fig. 1C), and pulmonary vasculopathy (Fig. 1D and E).

3. Discussion

Pulmonary emphysema combined with SSc-ILD was present in some never-smokers [1]. However, the mechanism of emphysema formation in SSc-ILD has never been clarified. This previous report indicated the presence of both emphysema in the upper lobes and pulmonary fibrosis in the lower lung zones as the radiological findings. Because our case revealed the coexistence of pathological emphysema and fibrosis, our case showed novel findings different from this previous report. Recently, we suggested that peripheral vasculopathy frequently seen in SSc patients may occur that can result in destruction of the fibrously thickened alveolar walls, resulting in the emphysematous change seen in SSc-ILD [2]. Pulmonary vasculopathy characterized by a thickening of the small vessel walls may begin early in the process of smoking-related emphysematous change [3,4]. In addition, Overbeek et al. reported that most SSc patients with or without PAH have intimal fibrosis in the small vessels such as arterioles, venules, and interlobular veins [5].

Our patient showed pulmonary vasculopathies in SSc-ILD, and moreover, we could confirm the area of emphysema formation by the presence of fibrotic alveolar walls, along with the vasculopathy of muscularisation of the arterioles. Therefore, we again realized the importance of pulmonary vasculopathy causing the coexistence of emphysema and fibrosis in SSc. However, immunohistochemical analysis could not be performed in our analysis. Moreover, vasculopathy and fibrosis in SSc appear to be different pathogenetic processes [6], determination of vasculopathy's role in accelerating emphysema is

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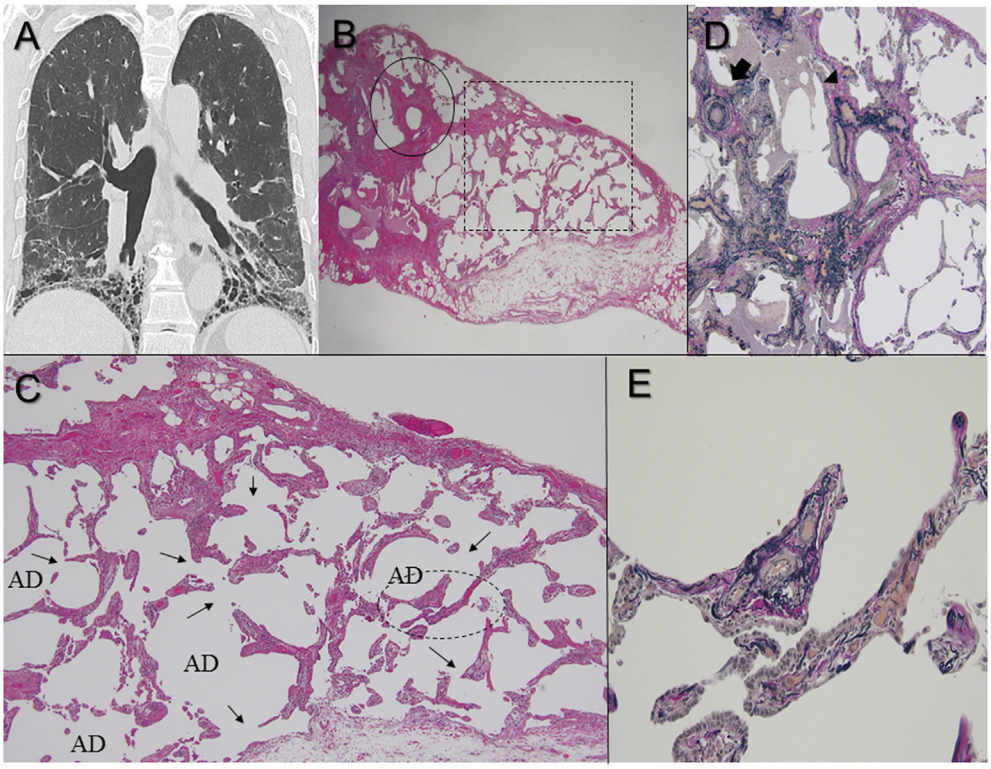

Fig. 1. (A) Chest CT shows reticulation and volume loss in the lower lungs along with severe traction bronchiectasis predominantly in the lower lungs. (B) Panoramic view of a specimen from the left lower lobe shows fibrosing nonspecific interstitial pneumonia with emphysematous change (H&E stain, × 1.25). (C) High-power magnification view of the square in B shows frequent splitting of the secondary alveolar walls (arrow) (H&E stain, × 4); AD: alveolar duct. (D) Magnified view of the circle in B shows concentric intimal fibrosis and medial thickening (Heath-Edward grade 3) of a medium-sized pulmonary artery (arrow) and intimal fibrosis of a pulmonary venule (arrowhead) (Elastica van Gieson stain, × 10). (E) Magnified view of the dotted circle in C shows muscularisation of an arteriole (arrow) of < 50 μm in diameter (Elastica van Gieson stain, × 20).

required in the future.

Conflict of interest statement

The authors declare no Conflicts of Interest (COI) in association with this article.

Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.rmcr.2018.09.009.

References

[1] K.M. Antoniou, G.A. Margaritopoulos, N.S. Goh, et al., Combined pulmonary fibrosis and emphysema in scleroderma-related lung disease has a major confounding effect on lung physiology and screening for pulmonary hypertension, Arthritis Rheumatol. 68 (4) (2016) 1004–1012.

[2] H. Yamakawa, T. Takemura, T. Iwasawa, et al., Emphysematous change with scleroderma-associated interstitial lung disease: the potential contribution of vasculopathy? BMC Pulm. Med. 18 (1) (2018) 25.

[3] N. Awano, M. Inomata, S. Ikushima, et al., Histological analysis of vasculopathy associated with pulmonary hypertension in combined pulmonary fibrosis and emphysema: comparison with idiopathic pulmonary fibrosis and emphysema alone, Histopathology 70 (6) (2016) 896–905.

[4] K. Hueper, J. Vogel-Claussen, M.A. Parikh, et al., Pulmonary microvascular blood flow in mild chronic obstructive pulmonary disease and emphysema. The MESA COPD study, Am. J. Respir. Crit. Care Med. 192 (5) (2015) 570–580.

[5] M.J. Overbeek, M.C. Vonk, A. Boonstra, et al., Pulmonary arterial hypertension in limited cutaneous systemic sclerosis: a distinctive vasculopathy, Eur. Respir. J. 34 (2) (2009) 371–379.

[6] E.F. de Carvalho, E.R. Parra, R. de Souza, et al., Arterial and interstitial remodelling processes in non-specific interstitial pneumonia: systemic sclerosis versus idiopathic, Histopathology 53 (2) (2008) 195–204.