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CLINICAL IMAGE

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Pleuroparenchymal fibroelastosis in mycobacterium avium complex lung disease

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Abstract

We report a rare case of pleuroparenchymal fibroelastosis (PPFE) with interstitial lung disease progressed after an onset of mycobacterium avium complex (MAC) lung disease. Clinicians should pay attention to the management for patients with PPFE in MAC lung disease.

KEYWORDS
interstitial lung disease, mycobacterium avium complex, pleuroparenchymal fibroelastosis

CLINICAL IMAGE

An 80-year-old Japanese woman was diagnosed as having pleuroparenchymal fibroelastosis (PPFE) with interstitial lung disease (ILD) at the age of 74. She was admitted to our hospital because of a 3-month history of persistent cough, dyspnea on exertion and body weight loss. Her radiographic findings showed usual interstitial pneumonia (UIP) pattern (Figure 1). The diagnosis of MAC lung disease was made on the basis of repeated positive sputum cultures. After the

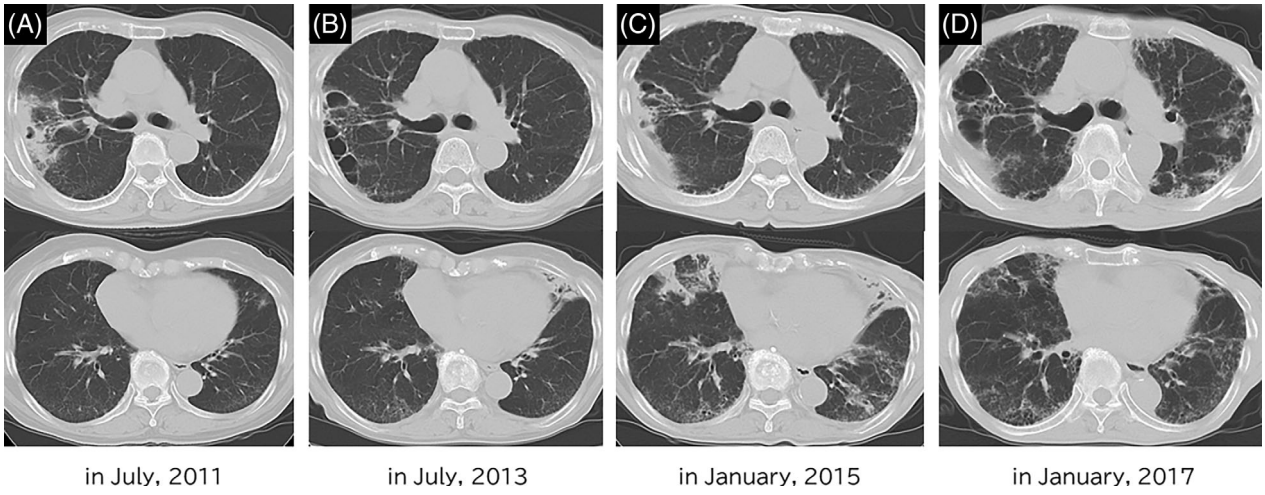

FIGURE 1 Serial changes in computed tomography (CT) of the chest. (A) At the initial visit, subpleural consolidation with cavity and air bronchogram was seen in the right upper lobe. In addition, the segmental bronchus was slightly thickened and dilated. There was a 5 mm diameter small nodule anteriorly in lingular segment of left lung. (B) Two years after antimycobacterial agents therapy began: consolidation in the right upper lobe was improved, but chest CT showed bronchiectasis, peripheral enlarged thick-walled cysts. In lingular segment of left lung, small nodule developed to consolidation. There has been some progression of the asymmetric and unevenly distributed fine reticular opacities. (C) Two years after the end of therapy, chest CT revealed peripheral areas of consolidation in the right upper lobe, and further deterioration of patchy consolidation and multiple nodules in both middle and lower lobes. (D) One year after antimycobacterial agents therapy began again, chest abnormalities were improved totally, but multiple nodular lesions of variable size emerged in the left upper lobe. The predominantly subpleural reticulation has increased in extent and coarsened in the bilateral lower lobes

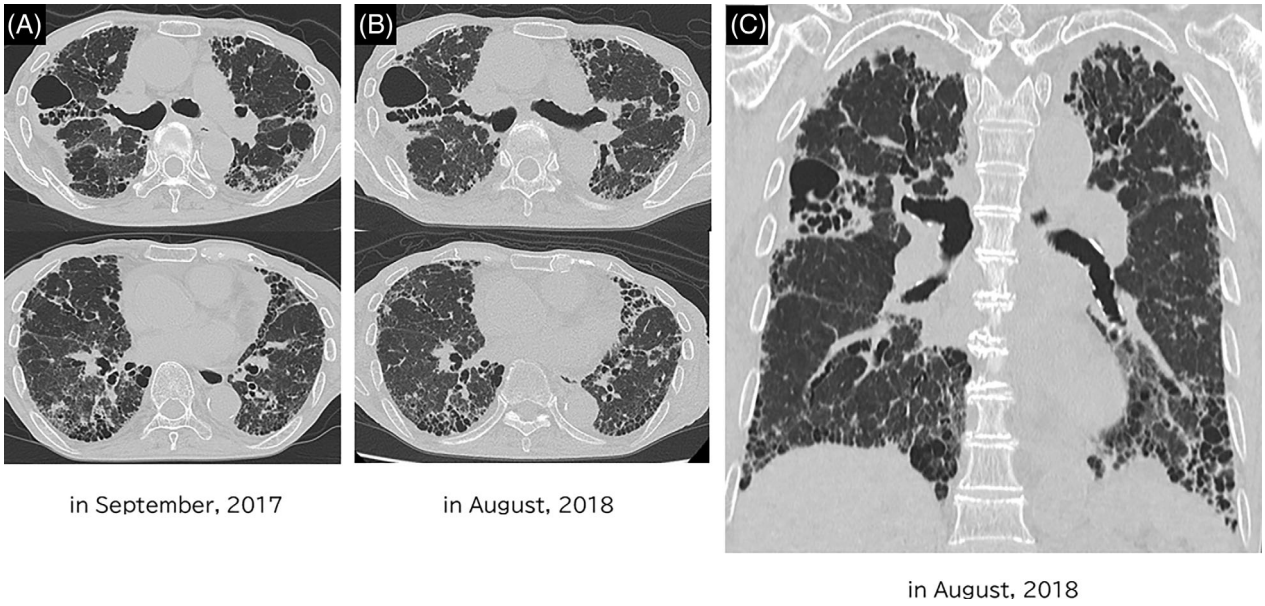

FIGURE 2 Serial changes in high-resolution computed tomography (HRCT) of the chest. (A) Following another 8 months (at first admission), images of chest HRCT showed further deterioration of cystic bronchiectasis and multiple small nodules. Subpleural honeycombing and reticulation with traction bronchiectasis were predominantly in the bilateral lower lobes. In addition, ground-glass opacities (GGOs) were seen in the bilateral lung fields. (B) After 1 year oral corticosteroid and antifibrotic therapy, GGOs and a part of nodules in both lung fields were improved. On the other hand, cystic bronchiectasis and multiple small nodules were gradually extended in the upper and middle zones predominance. (C) Coronal images of chest HRCT at same time (B). Note that subpleural dense consolidation (radiologically pleuroparenchymal fibroelastosis-like lesion) predominantly in the bilateral upper lobes and honeycombing in both lower lobes

initiation of combination of antimycobacterial agents, clinical symptoms improved temporarily, but PPFE with ILD gradually developed despite starting antifibrotic therapy. Her disease progressed and she developed chronic respiratory failure which led to her death (Figure 2). To our knowledge, little has been written and reported on the development of PPFE with ILD in MAC lung disease. The pathogenesis of PPFE with ILD in MAC lung disease remains obscure but it seems likely that triggers such as chronic/recurrent respiratory inflammation due to MAC infection may be involved.[1,2] At present, it is difficult to distinguish between coexistence of MAC lung disease with idiopathic PPFE and PPFE secondary to MAC lung disease. Therefore, further studies are needed to ascertain the pathophysiology.

AUTHOR CONTRIBUTION

Keishi Sugino was responsible for conceptualization and drafting the manuscript. Keishi Sugino, Hirotaka Ono, Masahiro Ando and Eiyasu Tsuboi analysed and interpreted the clinical and radiological data. All authors read and approved the final manuscript.

CONFLICT OF INTEREST

None declared.

DATA AVAILABILITY STATEMENT

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

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