Case 268

## 5. Pleuro-parenchymal fibroelastosis

### 【Discussion】

There are several diseases occupying bilateral upper lobes including tuberculosis, Kansasii (nontuberculous mycobacterium), silicosis, sarcoidosis, Aspergillosis and pleural parenchymal fibroelastosis (PPFE).

PPFE is reported one of the upper lobe fibrosis. It is critical for diagnosing precise PPFE to exclude other diseases.

There is a controversial whether PPFE reported by Franklin is identical to idiopathic pulmonary upper-lobe fibrosis (idiopathic PULF) reported by Amitani (1, 2). Histologically, the document by PPFE (subpleural fibrosis with a mixture of elastic tissue and dense collagen) is identical to that by idiopathic PULF (Subpleural atelectatic induration with the proliferation of elastic fibers and intraluminal) (1-3). However, idiopathic PULF called Amitani disease is clinically characteristic of associating with repetitive pneumothorax, flat thoracic cage, shortening of anterior-posterior diameter of thorax and a prominent suprasternal notch, as time progress and culminate in dying of pulmonary failure (3). Meanwhile, the clinical backgrounds of PPFE defined in English-speaking country, are ambiguous that include drug-induced and transplant-induced pneumonia (3, 4).

CT of PPFE or upper lobe fibrosis irrespective of Amitani disease and PPFE demonstrate apical pleural thickening, subpleural consolidation with coarse reticular pattern and striking traction of bronchiolectasis or bronchiectasis (5, 6).

The wall of trachea and bronchus are assembled mainly by cartilage. Bronchioles does not have cartilage in their own wall that are, however, assembled by smooth muscle and elastic fiber. Apical and upper lobe on PPFE or idiopathic upper lobe fibrosis are replaced by thick fibrosis and elastic fiber called elastosis, indicating degenerative change of lung tissue and collapse of secondary lobules. The trigger of upper lobe fibrosis irrespective of PPFE and Amitani disease is yet to be clarified.

In our case, she was given antibiotics and steroid for skin disease. CT demonstrated ground glass opacity in bilateral upper lobe indicating interstitial pneumonia but not either apical pleural thickening or dense reticular pattern. This pattern might be explained by drug-induced pneumonia. She had neither supraclavicular notch nor thoracic shortening of anterior-posterior diameter (flattening thoracic cage). Then, it indicates that she did not have Amitani disease but it is included in PPFE criteria, because of unknown origin.

## 【Summary】

We presented an eighty three-year-old female presented in our hospital for fever and dyspnea. CT demonstrated interstitial pneumonia in bilateral upper lobes, suspicious of pleural parenchymal fibroelastosis (PPFE). There are several diseases of occupying upper lobes such as: tuberculosis, nontuberculous mycobacteria, silicosis, sarcoidosis, aspergillosis and ankylosing spondylitis. These diseases are, first, excluded for diagnosing PPFE. It is borne in mind that histologically elastin and fibrin are proliferated occupying bilateral upper lobes for PPFE and idiopathic pulmonary upper lobe fibrosis (Amitani disease). CT demonstrated dens reticular pattern and apical pleural thickening associated with traction bronchial ectasia. Amitani disease is clinically characteristic of repetitive pneumothorax, flat thoracic cage, shortening of anterior-posterior diameter of thorax and a prominent suprasternal notch, as time progress. She does not belong to category of Amitani disease but does to that of PPFE.

## 【References】

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