

Case 406

4. Pleuro-parenchymal fibro-elastosis

【Progress】

She was under watchful observation because she had no respiratory symptoms.

【Discussion】

Bilateral apical lesions are often depicted on routine CT such as emphysema, tuberculosis, non-tuberculous mycobacterium called Kansai disease (1), silicosis, pleuro-parenchymal fibroelastosis (PPFE, Amitani disease), and chronic hypersensitivity pneumonia. The apical area includes a lymphatic excreting system. It includes that minute foreign bodies irrespective of organisms or non-organisms which are not excreted by airways, are carried away to lymphatic pathways. However, in the situations of the foreign bodies in volume being over excretion capability, the micro-foreign bodies remain or accept immune response accompanied with fibro-reticular pattern.

PPFE or Amitani diseases have common histological backgrounds: accumulation of thick elastic fibrosis with collapse of pulmonary parenchyma (2-4). Elastic fibers include bronchiole for preserving their lumens, taking place of cartilage that preserves lumens patency of bronchus walls. In fact, the difference of bronchiole from bronchus is whether the presence of absence of cartilage. Then, the presence of elastic fiber implies collapse of pulmonary parenchyma peripheral of bronchioles. However, in case of PPFE, elastic fiber proliferates alveolar walls that are unresolved. Both diseases also include thick pleural thickening. Fibrosis produced by fibroblasts can be absorbable, but fibrosis produced by myofibroblasts, unabsorbable. Myofibroblasts emerge when damages are too great for fibroblasts to overcome. Honeycomb pattern is not found in thick fibrous pleural thickening at apex.

The differences of PPFE from Amitani disease are clinical symptoms. Amitani diseases are often accompanied with pneumothorax and flattening of upper chest that depicts the trachea shifts backwards to the anterior line of thoracic vertebrae on chest CT. However, both diseases are common poorly prognostic for 10 to 20 years survivals. They are susceptible to infect mycotic infection such as Aspergillosis. As a result, Amitani disease might be included one of the extreme PTFE.

As one of the stable thick fibrous diseases with pleural thickening, chronic hypersensitivity disease is listed. The image characteristics of hypersensitivity pneumonia are three density patterns on CT: ground glass, normal density and hyperlucent (5,6). When antigen exposure finalizes, these patterns usually disappear. However, the exposure of allergic foreign bodies is persistent, fibrosis by fibrocytes falls into unabsorbable fibrosis by myofibroblasts, indicating that chronic hypersensitivity add the findings of thick fibrosis to three density patterns.

【Summary】

We presented a seventy-six-year-old female presented in our hospital for hem stool several times without respiratory symptoms. The follow-up CT images depict progressive fibrosis associated with pleural thickening and trachea dorsal margin is the same level of anterior trachea margin indicative of flattening upper chest that is not contradictory with pleuro-parenchymal fibroelastosis. It is borne in mind that when fibrous pleural thickening at apex, chronological CT check is crucial to check whether more thickening or stable, whether trachea dorsal line shifts backward or not or whether pneumothorax is associated or not.

【References】

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