Case 269

5. Pleuro-parenchymal fibroelastosis

【Discussion】

Histology of pleuro-parenchymal fibroelastosis (PPFE) indicate intra-alveolar fibrosis and deposition of elastin and fibrin in the interstitial. Elastin is distributed in all bronchial trees (1-3). The wall of bronchus is reinforced by cartilage and elastin while that of bronchiole alveolar duct and alveoli is done by elastin without cartilage (4, 5). Then, the hypothesis may be the case that elastosis is not produced but the result of collapse of the lobules which are supported by elastin.

Elastin which is one of extracellular materials (ECM), is produced by fibroblasts, smooth muscle cells and endothelial cells, while fibrin is produced by fibroblasts. Fibroblasts work for reparative process and they are differentiated from stem cells: club cells in bronchiole and type II pneumocytes in alveolus (6, 7).

There are usually cystic air spaces in PPFE on CT. Small air spaces are usually surrounded by dens and thick frame. Small air spaces are also found in emphysema and usual interstitial pneumonia (UIP). Air space associated with emphysema and UIP is formed by thin wall like a cyst or accumulated cysts. Air space on emphysema is formed by damages of centrilobular parenchyma or subpleural parenchyma. Air space on UIP is formed by apoptosis of type II pneumocytes, inducing dry up of surfactant and resulting in dilated non-functioning air spaces. Both air spaces on emphysema and UIP are surrounded by thin frame (8).

In short, the difference whether the frame is thick or thin between PPFE and emphysema or between PTFE and UIP depends on whether the lobules collapse is present or absent. It might be the case that the advance of lobules collapse induces thickness of air space frame & the surrounding pleura, leading the apical cage narrowing (9, 10).

Perfusion potency at upper lobe is less than at lower lobe because of gravidity. Ventilation might at upper lobe is less than at lower lobe because of less respiratory muscle strength. Upper lobes are less ventilated because of less intercostal muscle volume and less effect of diaphragm. Then, pathogen of tuberculous bacillus, nontuberculous mycobacterium (Kansasii) and aspergillosis fungus, particles of silicon and smoking are stagnant at the upper lobe, inducing lesion growth at upper lobes.

As for response mechanism against pathogen and foreign body, lymphatic channel develops at the upper lobe. Lymphatic channel of bronchial tree run together with pulmonary artery and bronchus, termed broncho-vascular bundle. In terminal bronchioles and respiratory bronchioles or more peripheral, the lymphatic channel along with broncho-vascular bundle is existing least. In the secondary lobules, another lymphatic channel exists in the interlobular septum along with pulmonary vein. The lymphatic channels both at broncho-vascular bundle and interlobular septum mainly flow out toward pulmonary hilum. The lymphatic channel especially at upper lobe flow out directly to mediastinum. The etiology of PPFE is yet to be clarified.

However, the subpleural fibrosis found in PPFE indicate lymphatic channel might work for the extrinsic pathogen or foreign body.

The collapse of lobules might be the case that club cells which inhabit at the surface of bronchiole are selectively and gradually damaged or falling into apoptosis. Club cells secrete surfactant similar fluid to clean pathogen and foreign body. They function to repair the damages to bronchiole. They are like stem cells. Their damages might fall into ruin for producing and preserving of lobules.

The prognosis of PPFE varies. However, a PPFE case with development of interstitial pneumonia in the lower lobes is relatively poor. In our case, the gradual narrowing thoracic cage and CT findings of pleural thickening and air spaces with thickened frame are characteristic of PPFE. Unfortunately, ground glass opacity in both lower lobes were found on chest CT, implying the possibility of poor prognosis.

【Summary】

We presented an eighty eight-year-old female presented in our hospital suffering from short breath soon after moving. Air spaces surrounded thick and dens frame are found in bilateral upper lobes on chest CT, compatible image with pleuro-parenchymal fibroelastosis (PPFE). Further, ground glass opacity in bilateral lower lobes is also associated, laboratory test revealing elevation of KL-6 values. It is borne in mind that the wall of bronchiole alveolar duct and alveoli is reinforced by elastin without cartilage. The accumulation of elastin might be the case of the result of massive collapse of apical lobules, leading to anterior-posterior narrow thoracic cage. Fibrin proliferation is the reparative process for pathogen or foreign particles by fibroblasts. Air spaces found in emphysema and UIP are surrounded by thin frame, probably by damages of lobules parenchyma.

Club cells usually work for preserving structure of bronchiole. The hypothesis might be the case that club cells situated on the surface of bronchiole fall in dysfunction or apoptosis. The prognosis of PPFE associated with ground glass opacity in the lower lobes are relatively poor, including our case.

【References】

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