Pathological diagnosis of diffuse interstitial lung diseases (ILDs) is based on the lung architecture, i.e., pulmonary lobule and acinus and vascular and lymphatic distribution. The exact correlation between histopathological features and HRCT findings can be achieved by understanding of the morphological basis of the lung.

(1) Pathological basis of fibrosis of interstitial pneumonia

The lung parenchyma is composed of alveolar structure and air. In a limited sense, interstitium of lung parenchyma indicates alveolar septum composed of pneumocytes, capillaries, and septal matrix. Not only alveolar septal fibrosis, but also intra-alveolar fibrosis is important in the pathogenesis of ILDs. Intra-alveolar organization through the alveolar epithelial and capillary injury, and reparative stage of mural incorporation fibrosis, and obliterative fibrosis participate in the fibrosis process of interstitial pneumonia. These patterns of intraluminal fibrosis are clearly demonstrated by elastic fiber staining.

(2) UIP is the basic pathological pattern of IIPs

Among the idiopathic interstitial pneumonias (IIPs), usual interstitial pneumonia (UIP), histology of idiopathic pulmonary fibrosis (IPF) is the most important and basic pathology of IIPs. UIP is characterized by marked fibrosis with architectural distortion, predominantly subpleural and paraseptal distribution with or without honeycombing, patchy involvement of lung parenchyma and fibroblastic foci (4–6). The perilobular fibrosis in the early phase of UIP is closely associated with interlobular vein and intralobular venules and also connective tissue septa. Honeycombing is a major feature of UIP, which is defined as clustered cystic air spaces (3–10 mm) on CT scans, located at subpleural and characterized by well-defined thick walls. Macroscopically, end-stage lung with honeycombing reveals 3–5 mm concavo-convex pleural surface, and round thick-walled clustered cystic lesions on cut sections. Histologically, collapsed alveoli in the thick walls of each cystic space with retracted pleura and bronchiolectasis are observed. “Microscopic honeycombing” reveals periacinar fibrosis with atelectasis of alveoli, dilated alveolar ducts with structural distortion and bronchiolar epithelial lining.
(3) Update of classification of IIPs

The update of the international multidisciplinary classification of the IIPs has been published in 2013\(^6\). There are specific areas in the update as follows; 1) Idiopathic NSIP is accepted\(^5\). 2) RB-ILD is commonly diagnosed without surgical biopsy. 3) The behavior of IPF is acknowledged to be heterogeneous. 4) Acute exacerbation of chronic fibrosing IIPs is well defined. 5) Unclassifiable IIPs are recognized, often because of mixed patterns of lung injury. 6) Clinical algorithm is necessary for classifying and managing IIP cases. 7) Pleuroparenchymal fibroelastosis (PPFE) is recognized as a specific rare entity. 8) Molecular and genetic studies are necessary to diagnose and predict prognosis. PPFE, unclassifiable IIPs and rare histologic IP patterns will be discussed in this session.

(4) Differential diagnosis between IIPs and chronic hypersensitivity pneumonitis

Clinico-radiological and pathological studies on chronic hypersensitivity pneumonitis (CHP) have been accumulated in the past 10 years. Biopsy findings suggesting HP include bronchiolocentric distribution, bridging fibrosis and poorly formed granulomas. However, up to 30% of subjects with histologic HP have no identifiable antigen\(^6\).

(5) Differential diagnosis between IIPs and connective tissue diseases

Connective tissue disease (CTD) complicates high frequency of interstitial pneumonia of NSIP and UIP. Lymphoid follicles with germinal center, pleuritis, bronchiolitis, and plasma cell infiltration are common histologic features of CTD-IP. A substantial percentage of patients with NSIP and UIP have some autoantibodies, but meet the criteria for a defined CTD, which described as lung dominant CTD or autoimmune featuredILD.

In conclusion, pathological diagnosis should be referred to HRCT findings in each case. The diagnosis and management of the IIPs require clinical-radiologic-pathologic correlation with multidisciplinary discussion.

References