Characterizing features of interstitial pneumonias

Pathology type	Clinical findings	Radiologic features
Usual interstitial pneumonia (UIP)	Defining of idiopathic pulmonary fibrosis but also associated with other chronically progressive fibrotic disease — such as connective tissue disease-interstitial lung disease, chronic hypersensitivity pneumonitis and pneumoconioses; portends poorer prognosis in the idiopathic setting when compared to other histology	Dominated by reticular and honeycomb findings; peripheral and bibasilar in distribution
Nonspecific interstitial pneumonia (NSIP)	Frequently associated with autoimmune disease; portends a better prognosis when compared with UIP; generally responsive to directed anti-inflammatory therapy but may be chronically progressive and fibrotic; biopsy confirmation often needed as up to one-third of so-called radiologic NSIP may be pathologic UIP	Dominated by reticular and ground-glass findings; honeycombing rare, though has been reported; peripheral and central distribution, often bibasilar more than upper lobe; characteristic subpleural sparing may be seen
Cryptogenic organizing pneumonia (COP)	May present with waxing and waning infectious-type symptoms, often requiring biopsy assessment to confirm after exclusion of other known causes, such as infection; generally responsive to empiric steroids, though repeat treatment may be needed along with occasional short-term immunosuppression	Migratory, consolidative and ground-glass infiltrates, often bilateral and peripheral with lower lobe predominance; atoll (reverse halo) sign supportive but not frequent; minimal fibrosis or long-term sequelae
Desquamative interstitial pneumonia (DIP)	Smoking related in over 80 percent of cases; prognosis better than UIP, particularly with smoking cessation; shared spectrum of clinical and pathologic overlap with RB-ILD	More centrally located and diffuse ground-glass infiltrates; occasional reticular findings centrally located without peripheral predominance
Respiratory bronchiolitis-interstitial lung disease (RB-ILD)	Younger age predilection in prior or active smokers; nonspecific presentation of dyspnea and cough with pigment-laden macrophages seen on pathology; smoking cessation is first order of management followed by steroid suppression	Patchy bilateral centrilobular ground-glass infiltrates or fine nodules, with airway enlargement or thickening; minimal reticular or fibrotic findings
Acute interstitial pneumonia (AIP)	Acute presentation of hypoxemic respiratory failure with diffuse infiltrates, often indistinguishable from idiopathic acute respiratory distress syndrome with typical diffuse alveolar damage seen on pathology; equivocal response to steroid therapy with high inpatient mortality	Patchy ground-glass infiltrates and consolidation, absent of underlying fibrotic or chronic appearing interstitial process
Lymphoid interstitial pneumonia (LIP)	Rare, and now considered to be more associated with secondary disease (rheumatologic, immunodeficient or hematologic) rather than idiopathic; characterized by extensive interstitial polyclonal lymphoid cell infiltrates on pathology	Thin-walled cystic findings in the majority, with underlying patchy ground-glass or consolidative features with lower lobe predominance
Pleuroparenchymal fibroelastosis (PPFE)	Pleural elastosis seen on pathology when biopsy is obtained, though clinical presentation nonspecific and often associated with underlying parenchymal disease of which UIP is most common; prognosis poor based on limited case series	Upper-lobe-predominant bilateral pleural thickening, often associated with underlying parenchymal interstitial process and varying degrees of fibrosis (possibly UIP vs. NSIP-like)