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Pathology and radiology correlation of idiopathic interstitial pneumonias☆,☆☆

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Summary By nature, idiopathic interstitial pneumonias have been diagnosed in a multidisciplinary manner. As classifications have been subject to significant refinement over the last decade, the importance of correlating clinical, radiologic, and pathologic information to arrive at a diagnosis, which will predict prognosis in any given patient, has become increasingly recognized. In 2013, the American Thoracic Society and European Respiratory Society updated the idiopathic interstitial pneumonias classification scheme, addressing the most recent updates in the field. The purpose of this review is to highlight the correlations between radiologic and pathologic findings in idiopathic interstitial pneumonias while using updated classification schemes and naming conventions.

1. Introduction

Among the major revisions to the American Thoracic Society/European Respiratory Society/Japanese Respiratory Society/Latin American Thoracic Association (ATS/ERS/JRS/ALAT) classification of idiopathic interstitial pneumonias (IIPs) is a reorganization of the categories, which, in our opinion, serves to better clarify and organize these often-confusing entities. The new classifications, summarized from the 2013 revision, are in Table 1 [1]. An important update is the grouping of these entities into major, rare, and unclassifiable categories.

The major IIP category is further subgrouped into 3 types: chronic fibrosing IIP, which includes idiopathic pulmonary fibrosis (IPF) and nonspecific interstitial pneumonia (NSIP); smoking-related IIP, which includes respiratory bronchiolitis–interstitial lung disease (RB-ILD) and desquamative interstitial pneumonia (DIP); and acute/subacute IIP, which includes cryptogenic organizing pneumonia (COP) and acute interstitial pneumonia (AIP) [1]. The second group, rare IIP, is composed of idiopathic lymphoid interstitial pneumonia (ILIP) and idiopathic pleuroparenchymal fibroelastosis (IPPFE). The final group, unclassifiable IIP, is characterized

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Table 1 ATS/ERS classification of IIP, 2013 [1]

Major IIP	Rare IIP
Chronic fibrosing	
IPF	ILIP
INSIP	IPPFE
Smoking related	
RB-ILD	
DIP	
Acute/subacute	
ILIP	
IPPFE	
Unclassifiable IIP	

by cases where, despite often-lengthy multidisciplinary discussions, a definitive diagnosis cannot be reached. Unclassifiable IIP include either cases with inadequate clinical, radiologic, or pathologic data or cases with major discordance between clinical, radiologic, or pathologic findings. Examples include evaluations of cases after previous therapy, new unrecognized entities, or unusual variants of recognized entities. This group represents a significant number of patients and in 1 interstitial lung disease cohort represented 10% of the total [2]. We will also briefly review important differential diagnoses and, when appropriate, will make reference to our own experience and prior published articles.

Thoracic high-resolution computed tomography (HRCT) has been shown to alter the clinical diagnosis, investigation, and management plans in patients with interstitial lung disease [3]. In the following review, we will summarize the radiologic and pathologic features of these entities with additional emphasis on radiologic/pathologic correlation. Although the current classification is an improvement over previous classifications, there continues to be areas that require clarification by evidence-based consensus. For instance, infrequent histologic patterns of IIP, such as acute fibrinous organizing pneumonias and a group of bronchiolocentric patterns, are currently unrecognized as distinct IIP by the ATS or ERS. It remains uncertain if these are variants of existing IIP or exist only in association with other known etiologies such as hypersensitivity pneumonitis (HP) or collagen vascular diseases (CVDs) [1].

Table 2 HRCT and histologic criteria for UIP pattern

UIP pattern	Possible UIP pattern	Inconsistent with UIP pattern
HRCT (all features required)	HRCT (all features required)	HRCT (any features)
Subpleural and basal predominant changes	Subpleural and basal predominant changes	Upper to mid-lung predominant changes
Reticular abnormalities	Reticular abnormalities	Peribronchovascular predominant involvement
Honeycombing with or without traction bronchiectasis	Absence of features listed as inconsistent with UIP pattern	Extensive ground glass abnormality (extent greater than reticular changes)
Absence of features listed as inconsistent with UIP pattern	Histology (probable UIP)	Profuse micronodules (bilateral, predominantly upper lobes)
Histology (all features required)	Marked predominantly subpleural/paraseptal fibrosis with associated architectural distortion ± honeycombing	Discrete cysts (multiple, bilateral, distant from honeycomb areas)
Marked, predominantly subpleural/paraseptal fibrosis with associated architectural distortion ± honeycombing	Absence of heterogeneous involvement of lung parenchyma OR absence of fibroblastic foci	Diffuse mosaic attenuation/air trapping (bilateral, in 3 or more lung lobes)
Heterogeneous involvement of lung parenchyma with interspersed areas of normal lung sharply demarcated with fibrotic changes	Absence of features leading to an alternate diagnosis	Bronchopulmonary segment(s)/lobe(s) consolidation
Fibroblastic foci	Honeycomb changes only [a]	Histology (any features)
Absence of features leading to an alternate diagnosis	Histology (possible UIP) (all features required)	Bronchiolocentric (airway) predominant changes
	Patchy/diffuse lung parenchymal fibrosis with or without interstitial inflammation [b]	Features leading to an alternate diagnosis
	Absence of other criteria for UIP	DAD without underlying changes of UIP [c]
	Absence of features leading to an alternate diagnosis	Organizing pneumonia without underlying changes of UIP [d]
		Granulomas [e]

[a] A lung biopsy demonstrating exclusively honeycomb change in the lung is not diagnostic of UIP and can be seen in a number of other conditions including chronic HP. If a biopsy demonstrates exclusively honeycomb change, no definitive diagnosis can be made.

[b] Inflammation should be minimal.

[c] DAD, acute and organizing, can be seen in acute exacerbation of UIP.

[d] Patchy organizing pneumonia can be seen in conjunction with UIP.

[e] Rare granulomas can be seen in patients with UIP. If they are a predominant feature, other diagnoses such as HP, infection, and drug reactions must be considered [3].