Radiologic Evaluation of Idiopathic Interstitial Pneumonias

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KEYWORDS

• Idiopathic interstitial pneumonia • Interstitial lung disease • High-resolution CT

KEY POINTS

- Many known secondary causes can produce changes identical those seen in the idiopathic interstitial pneumonias, most commonly collagen vascular diseases, hypersensitivity pneumonitis, and drug reactions.
- Findings required for a confident radiologic diagnosis of usual interstitial pneumonia (UIP) are predominant distribution of fibrosis in the peripheral and posterior-basilar lung with associated honeycombing.
- Lung cancer in the setting of pulmonary fibrosis is often subtle, and careful inspection is mandatory to exclude early malignancy.
- Mild UIP and mild nonspecific interstitial pneumonia (NSIP) may seem similar; given the worse prognosis of UIP versus NSIP, these mild patterns are frequently grouped into the possible UIP category.
- A surgical biopsy may be required.

INTRODUCTION

The idiopathic interstitial pneumonias (IIPs) are a group of fibrosing and inflammatory pulmonary conditions that share many similar clinical features.[1] The classification of IIPs is based on specific histologic changes, each of which is related to a specific idiopathic condition. It is important to remember that many known secondary causes can produce histologic changes identical to those seen in the IIPs, most commonly collagen vascular diseases, hypersensitivity pneumonitis (HP), and drug reactions.[2,3] The IIPs are grouped into the chronic fibrotic conditions (usual interstitial pneumonia [UIP] and nonspecific interstitial pneumonia [NSIP]), the subacute and acute conditions (cryptogenic organizing pneumonia [COP] and acute interstitial pneumonia [AIP]), the smoking-related conditions (respiratory bronchiolitis interstitial lung disease [RB-ILD] and desquamative interstitial pneumonia [DIP]), and last, the rare conditions (lymphocytic interstitial pneumonia [LIP] and idiopathic pleuroparenchymal fibroelastosis).[2]

Given the many similarities between these conditions in terms of clinical presentation and radiologic/histologic findings, a collaborative multidisciplinary approach between the clinician, radiologist, and pathologist is paramount to achieve an accurate diagnosis. A collaborative diagnostic approach may not only lead to obtaining a more confident diagnosis in a shorter time, but also may often preclude the need for surgical lung biopsy.[4–6] Additionally, a collaborative approach is essential to exclude a known cause of lung disease.[2,7,8] The following is a discussion of the radiologic contribution to the multidisciplinary

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evaluation, with description of the typical radiologic findings seen in each of the IIPs and important associated caveats.

FIBROSING INTERSTITIAL PNEUMONIAS

Usual Interstitial Pneumonia

UIP is the most common of the IIPs and statistically carries the worst long term prognosis.[9,10] It is the histologic and radiologic correlate for idiopathic pulmonary fibrosis (IPF). A UIP-type pattern of pulmonary fibrosis can also be secondary to other conditions, including collagen vascular disease, chronic HP, drug reaction, and asbestosis. The UIP-type pattern induced by other underlying conditions may be indistinguishable from truly idiopathic UIP/IPF. Careful clinical evaluation for potential underlying causes of UIP is essential, because patients with a known secondary cause have a much better prognosis compared with patients with idiopathic UIP, where secondary causes have been excluded.[2,11]

The diagnosis of UIP can frequently be made solely by high-resolution CT (HRCT) evaluation. Multiple studies have shown that expert thoracic radiologists confidently diagnosing UIP have a positive predictive value of 95% to 100%. However, a less confident diagnosis of UIP drops the positive predictive value to as low as 70%.[4,12–14] A typical UIP pattern on HRCT with the appropriate clinical picture can often eliminate a surgical biopsy.[5,15] The guideline-based criteria for radiologic diagnosis of UIP now reflect the level of confidence in the diagnosis and should help to guide further diagnostic steps.[2]

On HRCT, the hallmark features of UIP are a peripheral predominant reticulation in a predominantly posterior and basilar lung distribution, with associated honeycombing (**Fig. 1**). Regions of reticulation represent fine fibrosis on histology.[16] Ground-glass opacity may also be seen, but is less prominent than the associated reticulation and represents microscopic pulmonary fibrosis beyond the resolution of HRCT.[17] The fibrotic features of UIP may be asymmetric compared with NSIP, which is usually symmetric.[18,19] Traction bronchiectasis and bronchiolectasis may develop in areas of reticulation, indicating that reticulation reflects fibrosis.[20,21] Unfortunately, in a substantial proportion of biopsy-proven cases of UIP/IPF, imaging findings are not specific for UIP.[22]

As UIP progresses, honeycombing develops in the subpleural lungs, representing end-stage fibrosis.[16] Honeycombing in a basal and peripheral distribution of fibrosis is highly supportive of a UIP diagnosis and should be considered definite UIP based on recently released guidelines (**Tables 1** and **2**).[22] Differentiating paraseptal emphysema from honeycombing, although usually straightforward, can be challenging and is a not uncommon dilemma given that smoking is associated with both entities. Honeycombing usually manifests as a regular pattern of thin-walled cysts, often in the lower lungs, whereas paraseptal emphysema most often manifests as several longer cysts often in the upper lungs and may contain subtle internal septations. However, confident differentiation of honeycombing from paraseptal emphysema may not be possible in a few cases.[23–25]

Although typical HRCT features may obviate the need for biopsy, the lack of typical findings does not rule out this diagnosis. It is recognized that in a substantial minority of cases the diagnosis of UIP cannot be made solely on CT. Up to 30% to 50% of UIP cases diagnosed by histology do not carry a confident radiologic diagnosis of UIP.[4,26] Equivocal radiologic features or clinical uncertainty should prompt consideration of surgical biopsy.[20,27–29]

Imaging is helpful in detecting complications of UIP/IPF. Acute respiratory decline in IPF patients are often a result of accelerated deterioration or opportunistic infections. Both of these conditions may have an overlapping appearance, demonstrating new regions of prominent ground-glass opacification, possibly with associated

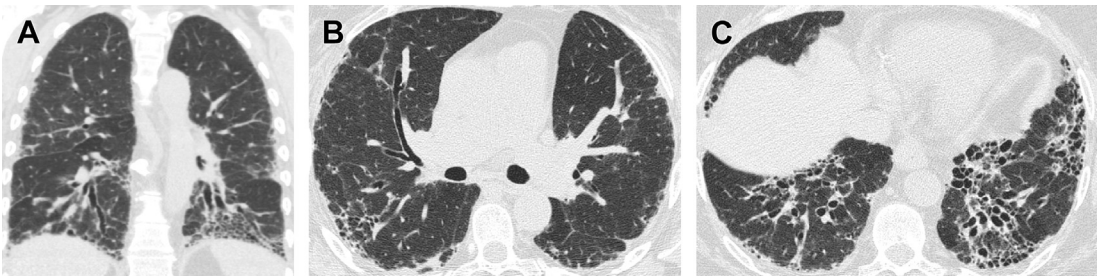

Fig. 1. Usual interstitial pneumonia (UIP) pattern of fibrosis. Coronal (*A*), axial mid lung (*B*), and axial lower lung (*C*) images showing peripheral and basilar predominant reticulation, honeycombing and traction bronchiectasis. Note the asymmetric pattern of fibrosis which is more common in UIP than nonspecific interstitial pneumonia.