Imaging features of Idiopathic Pulmonary Fibrosis: A pictorial review

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Learning objectives

1. To review the spectrum of imaging characteristics of idiopathic pulmonary fibrosis (IPF) with respect to the revised ATS/ERS/JRS/ALAT (American Thoracic Society, the European Respiratory Society, the Japanese Respiratory Society and the Latin American Thoracic Association) imaging criteria.
2. To emphasize the radiological and clinical key points necessary to correctly diagnose IPF and exclude other interstitial pneumonias.
Background

The interstitial pneumonias are a heterogeneous group of diseases affecting the lung interstitium and can be divided into idiopathic or secondary to a variety of causes such as collagen vascular diseases, pneumoconiosis, infections, smoking, among others [1].

Since they exhibit diverse pulmonary manifestations and some demonstrate overlapping HRCT features, the diagnosis of interstitial pneumonias is not always straightforward.

IPF is the most prevalent idiopathic interstitial pneumonia and is associated with a histologic and / or radiologic pattern of usual interstitial pneumonia (UIP) [2]. It is a chronic, progressive fibrotic lung disease of unknown cause, with a very poor prognostic (the overall median survival time is approximately 3 years) and is slightly more prevalent in men, usually over 50 years old [3].

Despite the important role of imaging in suggesting the diagnosis, the definite diagnosis always needs correlation with clinical manifestations and, frequently, also with pathology [3].
Imaging findings OR Procedure details

Since interstitial pneumonias are a varied group of diseases with very distinct courses and prognosis, it is vital to make a correct diagnosis in order to provide proper management. HRCT has a fundamental role not only in diagnosing but also in monitoring treatment results and progression of the disease. As earlier mentioned, pulmonary manifestations not always assume the typical imaging characteristics and they may overlap other diseases, so evidence-based guidelines were necessary to reach a confident diagnosis.

In 2000 the ATS/ERS/JRS/ALAT criteria was developed but it was mainly based in clinical aspects and the included CT findings were not very specific for IPF.

In 2011, the revised diagnostic criteria clearly divided the HRCT findings into "UIP", "possible UIP" and "inconsistent with UIP", the last two needing biopsy and clinical correlation for a definite diagnosis [3].

The panel also concluded that in the correct clinical context, the HRCT findings of a UIP pattern translated by the presence of honeycombing in an appropriated distribution was sufficient to reach a diagnosis of IPF without the need to perform lung biopsy.

Histologic hallmark feature of "UIP pattern" is the presence of scattered fibroblastic foci [2].

According to the revised criteria, imaging characteristics consistent with "UIP pattern" must include the presence of bilateral, predominantly subpleural, reticular abnormalities with an apicobasal gradient and honeycombing (clustered, mainly subpleural, cystic air space with thick well-defined walls of comparable diameters, usually 3-10 mm) with or without traction bronchiectasis in the absence of features inconsistent with UIP. It’s important to note that ground glass opacities are present in the majority of patients with UIP but are usually limited in extent [2].

"Possible UIP pattern" refers to the same characteristics of "UIP pattern" but without the presence of honeycombing.

The presence of upper or mid-lung predominance, peribronchovascular predominance, extensive ground glass opacities, profuse micronodules, discrete cysts (multiple, bilateral, away from the areas of honeycombing), diffuse mosaic attenuation or
consolidation in bronchopulmonary segments are "inconsistent with UIP pattern" and favor other diagnosis. [4]

In the presence of an UIP pattern, the diagnosis of IPF is an exclusion diagnosis once other causes of UIP pattern such as collagen vascular disease, drug toxicity and environmental exposures have been eliminated.

As with the HRCT, lung biopsy is not always diagnostic; it is important to notice that the lung is not uniformly involved in IPF, especially in early stages, so CT must be performed before biopsy for planning which segment should be sampled. Also, biopsy samples should be obtained from more than one lobe of the lungs, particularly because several subtypes of interstitial pneumonias can coexist.

The possible histopathological results are: findings compatible with UIP, not UIP, probable/possible UIP and non-classifiable fibrosis; in the last three situations, a multidisciplinary group should discuss the probability of IPF as the diagnosis and the best management in that particular case [4].

**Differential diagnosis:**

Although the advent of the revised diagnostic criteria of IFP is a major advance, there are still some key problems to be addressed, including lack of specific recommendations in the scenario of probable/possible IPF and the HRCT misdiagnosis of IPF by less experienced radiologists. [4] One of the problems is related to distinguishing honeycombing from pulmonary emphysema and traction bronchiectasis; in general, honeycombing is more thick-walled, subpleural and parallel to the chest wall than emphysema and in the second case a dilated airway can be identified by scrolling through contiguous thin-section axial images or by evaluating coronal reformats. [5] Other situation that can lead to misdiagnosing IPF is the presence of atypical imaging findings, such as extensive ground-glass opacities, nodules or microcystic honeycombing (typical of NSIP) or findings typical of COP like consolidation or a predominantly peribronchovascular distribution. [2]
Images for this section:

Fig. 1: 64 year-old man with IPF. Axial (a) and coronal (b) high-resolution CT images show typical findings of UIP pattern, with bilateral honeycombing (consisting of clustered, mainly subpleural, cystic air space with thick well-defined walls of comparable diameters, between 3-10 mm - green arrows) and marked reticular opacities (orange arrows). Note the asymmetrical but bilateral involvement of the lungs and the presence of an apicobasal gradient, well documented in (b).

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**Fig. 2:** 65 year-old woman with end-stage IPF and right lung transplant. Coronal high-resolution CT image shows typical findings of UIP pattern, with diffuse honeycombing and reticular opacities affecting the left lung, predominantly subpleural and with apicobasal gradient.

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**Fig. 3:** 62 year-old woman with early stage biopsy-proven IPF. Axial high-resolution CT image shows findings of possible UIP pattern, characterized by basal reticular opacities with subpleural and posterior predominance (orange arrows) and some traction bronchiolectasis (blue arrow).

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**Fig. 4:** 75 year-old man with rapidly progressive IPF. (a), (b) and (c): High-resolution CT images obtained at presentation shows reticular opacities (orange arrows), honeycombing (green arrows) and focal ground-glass opacity (red arrow). Moderate traction bronchiectasis are also present (blue arrow). These findings are consistent with the UIP pattern. (d), (e) and (f): Follow-up CT images (at the same levels) obtained 3 months later shows marked progression of the honeycombing, ground-glass densities and traction bronchiectasis, in particular at lung bases.

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**Fig. 5:** Secondary UIP pattern in a 55 year-old woman with SLE (Systemic Lupus Erythematosus). Axial high-resolution CT image shows areas of reticular density with subpleural predominance (orange arrow) associated with traction bronchiolectasis (blue arrow) and some ground-glass densities (red arrow). These features are compatible with possible UIP; biopsy findings were also in agreement with the CT characteristics.

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Fig. 6: Possible UIP pattern in a 67 year-old woman with no risk factors for lung interstitial disease. (a) Axial high-resolution CT image shows areas of reticular density with subpleural and basal predominance (orange arrows) associated with incipient ground-glass densities (red arrows). These features are compatible with possible UIP; biopsy findings were concordant with the CT characteristics and a confident diagnosis of IPF was achieved.

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Fig. 7: 56 year-old man, heavy smoker. (a) Axial high-resolution CT image shows marked centrilobular and paraseptal emphysema with superior lobes predominance. (b) Images of the inferior lobes show subpleural reticular opacities, honeycombing and traction bronchiectasis, findings compatible with UIP. (c) Images of the middle of the lungs show the difficulty in distinguishing emphysematous spaces from cysts associated with honeycombing. In general, honeycombing is more thick-walled, subpleural and parallel to the chest wall than emphysema. This patient has severe emphysema and concomitant IPF.

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**Fig. 8:** CT pattern inconsistent with UIP in a 55 year-old man. Axial high-resolution CT images shows areas of reticular density with subpleural predominance associated with ground-glass densities (red arrow) and traction bronchiectasis (blue arrow). Although these findings can suggest UIP, the presence of upper and mid-lung predominance (image b) and some peribronchovascular distribution of ground-glass opacities (yellow arrow) are inconsistent with UIP. The relative sparing of costophrenic angles (image c) is another finding unusual in IPF (more characteristic of secondary UIP). Biopsy proved to be Hypersensitivity Pneumonitis.

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Fig. 9: 51 year-old man, heavy smoker. CT findings inconsistent with UIP. Although axial high-resolution CT image shows incipient bilateral and basal reticular densities, the patchy ground-glass opacities is the most obvious finding. Multiple small cystic spaces in the posterior regions are also present but note these are away from the areas of typical honeycombing. Emphysema and bronchial wall thickening are also seen. These findings, in conjunction with the background of the patient, are consistent with the diagnosis of DIP (Descamative Interstitial Pneumonia).

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Fig. 10: 89 year-old man, with history of exposure to asbestos fibers. (a) Coronal high-resolution CT image shows calcified pleural plaques (yellow arrows). (b) Reticular opacities with subpleural and basal predominance (orange arrows) and traction bronchiectasis (blue arrow) are findings that are usually present in UIP pattern. In this case, the presence of pleural plaques, clinches the diagnosis of asbestosis.

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**Fig. 11:** 90 year-old woman with COP (Cryptogenic Organising Pneumonia) with associated fibrosis. (a), (b) and (c) Axial and coronal high-resolution CT images at different levels shows subpleural areas of reticular densities (orange arrows) and subtle honeycombing (green arrows); although these findings could suggest IPF, the presence of bilateral patchy, peripheral areas of consolidation (yellow arrows) suggests the diagnosis of COP with associated fibrosis.

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Fig. 12: 67 year-old woman with fibrotic NSIP. (a) and (b) High-resolution CT image shows extensive ground-glass opacities with an apicobasal gradient associated with basal reticular pattern (orange arrows) and traction bronchiolectasis (blue arrows), findings suggestive of fibrotic NSIP.

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Conclusion

IPF is associated with a very poor prognosis and an accurate diagnosis is essential for optimal management. Since HRCT is such a fundamental tool, the radiologist should be acquainted with the revised imaging criteria of IPF in order to avoid misdiagnosing this pathology.
References


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