Interstitial Pulmonary Fibrosis: How to classify honeycombing/reticular abnormalities without basal predominance in absence of features of inconsistent UIP following ATS 2011 guidelines?

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Learning objectives

The aim of the study was to assess the no-corrispondence of some Computed Tomography (CT) UIP pattern with the updated evidence-based guidelines for diagnosis and management of IPF devised in 2011 by the American Thoracic Society, the European Respiratory Society, the Japanese Respiratory Society, and the Latin American Thoracic Association.
Background

Idiopathic pulmonary fibrosis (IPF) is defined as a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause, occurring primarily in older adults, and limited to the lungs. It is characterized by progressive worsening of dyspnea and lung function and is associated with a poor prognosis [1].

Nonspecific interstitial pneumonia presents as a chronic or subacute process that fits into clinical spectrum of IPF; this form of interstitial pneumonia has a more favorable prognosis and need to be distinguished from IPF but that also differ from DIP, AIP, and COP [2].

Currently, HRCT scans are an integral part of the evaluation of patients with diffuse lung diseases [3] because of its usefulness in the diagnosis of pulmonary lesions and in determining prognosis and monitoring the progression of lesions [4].

HRCT scanning of the lung has become an indispensable tool to identify the presence of interstitial lung disease (ILD) and an organized approach to HRCT interpretation in these patients is required for proper diagnosis and management[5].

The multiple different ways in which physicians approach, the variability in the natural history of disease and in HRCT appearances, the lack of a validated algorithm for excluding known causes of lung fibrosis all contribute to the inherent confusion surrounding the diagnostic uncertainties characterizing IPF [6].

International evidence-based guideline on the diagnosis and management of IPF, with the purpose to analyze the additional evidence accumulated since the publication of the 2000 ATS/ERS consensus statement and to provide evidence-based recommendations for management, with an emphasis on diagnosis and treatment, have been published by the American Thoracic Society in 2011[7].
Findings and procedure details

127 patients were retrospectively selected from the archive of the Department of Lung Pathology at the Tor Vergata University of Rome; one case was excluded after an initial pathologic review; 103 cases presented a diagnosis of Idiopathic Interstitial Pneumonia (IPF/UIP), 23 patients presented a diagnosis of Non Specific Interstitial Pneumonia (NSIP) [diagnosis of Idiopathic Interstitial Pneumonia (IPF/UIP) and Non Specific Interstitial Pneumonia (NSIP) drawn on in our centre by multidisciplinary discussion with pulmonologists, radiologists and pathologists (if available lung biopsy].

Biopsy were available for six patients with a diagnosis of IPF/UIP and four patients with a diagnosis of Non Specific Interstitial Pneumonia (NSIP).

Of all the patients were available at least one high resolution computer tomography performed between 2008 and 2013; the C.T. chest of all patients were examined by two radiologists with great experience of Interstitial Pneumonia, Diffuse Pulmonary Lung Diseases.

Diagnosis were reassessed on the basis of radiological pattern found by the radiologists according to the criteria of ATS 2011 guidelines.
Conclusion

In general 18 of the 126 assessed patients (14.3%) don’t exhibit apical-basal gradient of the characteristic lesions of the disease (Fig.1-4).

Among 11 patients with an IPF/UIP, without apical-basal gradient, 7 (63.6%) presented honeycombing lung as much in the apical lung segments as in the basal lung segments. The remaining IPF/UIP patients (4 of 11 - 36.4%) presented reticular abnormalities as in the apical lung segments as in the basal lung segments. All the NSIP patients without apical-basal gradient presented reticular abnormalities as in the apical lung segments as in the basal lung segments.

The ATS 2011 HRCT criteria for UIP or "possible" UIP pattern request the presence of subpleural, basal predominance. Our work highlights a number of cases that reflect the characteristic aspects of the IPF/UIP (honeycombing lung, reticular abnormalities, subpleural predominance) in the absence of basal predominance. These cases do not meet criteria for UIP pattern neither for "possible" pattern resulting "inconsistent with" UIP radiological pattern. The parameters used to define the HRCT pattern result perhaps too selective, at least for the "possible" UIP HRCT pattern.

A number of cases that reflect all the features (except basal predominance) in order to be classified as IPF are excluded from this new radiological classification, while not presenting radiological features suggestive of other diffuse parenchymal lung disease (Fig.4). At this point, how should be classified these cases from the radiological point of view?
Fig. 1: Upper lung lobes axial CT scan showing reticular, subpleural abnormalities.

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Fig. 2: Mid thorax axial CT scan showing reticular, subpleural abnormalities more evident at the right lung.

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Fig. 3: Basal lung CT scan showing reticular, subpleural abnormalities without absence of features listed as inconsistent with UIP pattern (ATS 2011 guidelines).

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**Fig. 4:** Para-sagittal MPR reconstruction of the right lung showing reticular, subpleural abnormalities without a basal predominance with absence of features listed as inconsistent with UIP pattern (ATS 2011 guidelines).

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References


