UPDATE ON THE CLASSIFICATION OF INTERSTITIAL LUNG DISEASE

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Aims:
- To review recognised patterns of interstitial pneumonias nearly a decade on from the ATS/ERS consensus classification.
- To review a decade’s change in biopsy practices, biopsy types and how we perceive the ILD spectrum overall.
- To review critically some new patterns of ILD.

Since 2002, the seven histological patterns were recommended within the consensus classification for idiopathic interstitial pneumonias have undergone varying degrees of critical review. In relation to usual interstitial pneumonia (UIP) and a diagnosis of idiopathic pulmonary fibrosis, updated management recommendations are due to be published later this year, approved by the American, European and Japanese respiratory societies. Within these, there are revised histological criteria for a pattern of UIP, although these are not significantly different to those in the 2002 classification. The most significant change is recommending varying degrees of confidence in relation to the pattern can be integrated into multidisciplinary review, in particular the HRCT data. There has also been increasing interest in the epithelial-mesenchymal interface in relation to aetiology with the fibroblastic focus being a centre of attention as evidence suggests, though not universally, that their extent relates to prognosis. A pattern of UIP is also being increasingly recognised in patients with chronic hypersensitivity pneumonia.

In relation to non-specific interstitial pneumonia, particularly the fibrotic variant (F-NSIP) the 2002 document set provisional status in relation to a defined idiopathic clinicopathological entity. Subsequent work, in particularly an ATS-sponsored workshop, suggests that true idiopathic cases exist, albeit rarely, but that a cause is often found on multidisciplinary review, sometimes only becoming apparent on follow-up. In relation to respiratory bronchiolitis (RB) and desquamative interstitial pneumonia (DIP), there is general agreement that these histological patterns overlap and are often associated/caused by exposure to cigarette smoke. However, there are differences in the histological features and, whilst RB is nearly always is due to exposure to cigarette smoke, some cases of DIP are idiopathic. As such, although smoking-related interstitial lung disease has become a term used in diagnosis, it is not currently recommended as a unifying histopathologic term. Little has changed in relation to diffuse alveolar damage (DAD), other than there has been publication of a definition for acute exacerbation of IPF, histologically when DAD is superimposed upon a pattern of UIP. In relation to organising pneumonia, the definition of this histologic pattern has not changed, although there is increasing recognition of patients who progress to established interstitial fibrosis.

Finally, lymphoid interstitial pneumonia (LIP) has become increasingly rare over the past decade, likely due to preferential classification as cellular NSIP. A few patients still fit the clinicopathologic definition, although it is exceptionally rare for these to be in the idiopathic setting.

Outside of adults, these histological patterns are recognised in children, with a proposed more complex classification of childhood interstitial lung disease published in 2007. Its contents are outside the scope of this talk, although it is worth noting that
surfactant protein gene mutations are being increasingly identified and these may yet impact on adult classifications, especially familial cases and in younger patients.

There have also been advances in relation to the process of biopsing. Most institutions now sample at least two sites, ideally using preoperative targeting of areas showing active but not end-stage disease.\textsuperscript{10} This can lead to a decrease in inadequate biopsies. Controversy remains over whether transbronchial biopsies can/should be used to diagnose a pattern of UIP, although the upcoming document on the management of IPF recommends against this practice.\textsuperscript{11, 12} The decision to biopsy also appears to be involving over time. As an example, biopsies for RB-ILD have become increasingly rare over the past decade whilst there appears to be a slight increase in biopsies showing a pattern of usual interstitial pneumonia likely reflecting the importance of accurate diagnosis in relation to recent and ongoing drug trials.

Finally, the last decade has seen several new proposed histological patterns of interstitial lung disease being presented or published. These include pleuroparenchymal fibroelastosis,\textsuperscript{13, 14} a variety of bronchiocentric interstitial pneumonias/fibroses\textsuperscript{15-17} and acute fibrinous and organising pneumonia,\textsuperscript{18} together with unusual overlap patterns of interstitial lung disease such as hypersensitivity pneumonia and alveolar proteinosis.\textsuperscript{19} I have also seen an unusual cohort of patients with unexplained diffuse cystic lung disease and coexistent small airways disease that may be important in relation to the differential diagnosis of lymphangioleiomyomatosis.

Reference List


