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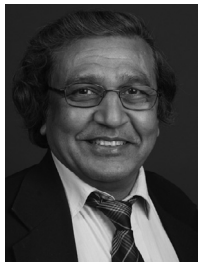
Foreword



Ganesh Raghu, MD, FCCP, FACP

Director CENTER for Interstitial Lung Disease (ILD), UW Medicine, ILD, Sarcoid and Pulmonary Fibrosis Program Co-Director, Scleroderma Clinic, UW Medicine Seattle, WA 98195

In the absence of infection, several acute and chronic lung disorders with variable intensity and extent of pulmonary inflammation and fibrosis are commonly referred to as interstitial lung disease (ILD). Over 150 agents and clinical situations have been listed as ILD. This list is enormous if one sums the several pharmaceutical and environmental agents and clinical situations as individual cause or clinical conditions. One simple way to group these includes ILD associated with (1) occupational and environmental factors (inhalation cause); (2) connective tissue diseases; (3) granulomatous lung disease; (4) some inherited diseases (metabolic storage diseases, Hermansky–Pudlak syndrome, neurofibromatosis etc), familial ILDs, and associated genetic mutations including telomere shortening, TERT/TERC, surfactant proteins; (5) certain specific entities (eg, pulmonary Langerhans cell granulomatosis, lymphangioleiomyomatosis); and (6) idiopathic interstitial pneumonias (IIP).

Ever since the term "IPF" got formalized by a consensus of opinions of international experts representing the American Thoracic Society and European Respiratory society in the year 2000, idiopathic pulmonary fibrosis (IPF), a subgroup of IIP has received a lot of worldwide attention by stakeholders that include patients, clinicians, investigators, patient advocacy groups, regulating agencies, donors, pharmaceutical industries, investors, basic and clinical scientists over the last 2 decades. The subgroup/entities of IIP have been classified and recently reclassified/clarified in international consensus statements.

While, IPF is clearly acknowledged as a disease with fatal prognosis, significant progress has been made in the understanding of the natural course and management of IPF over the last 25 years .We now have 2 drugs classed as "antifibrotic" agents approved for treatment of IPF.

With the advent of high-resolution computed tomography (HRCT) scans of the chest, distinctive patterns and distribution of the abnormalities of the lung parenchyma have been recognized. In this regard, substantial progress has been made in the field of individual interstitial pneumonias—specific HRCT patterns of usual interstitial pneumonia (UIP), nonspecific interstitial pneumonia, organizing pneumonia, respiratory bronchiolitis-desquamative interstitial pneumonia, lymphocytic interstitial pneumonia, pleuroparenchymal fibroelastosis, and combined pulmonary fibrosis with emphysema as well as alveolar filling disorders such as alveolar proteinosis, alveolar hemorrhage have helped the clinician confronted with the patient with new onset ILD to narrow down the differential diagnosis with the key input from the radiologist.

The radiologists and clinicians of this era must familiarize themselves and become aware of the several distinguishing features of ILD and IIP and recognize that IPF is a distinct clinical entity when confronted with a patient with ILD. It is essential to make an accurate diagnosis for appropriate therapeutic interventions and discuss prognosis with the patient. The pattern of UIP has been recently refined by international experts in documents published by major respiratory (the American Thoracic Society; the European Respiratory Society; the Japanese Respiratory Society, and the Asociacion Latinoamericana de Torax) and radiological (Fleischner) societies; the presence of UIP pattern in the HRCT scans eliminates the need for lung biopsy. However, the UIP pattern, despite it being the hallmark feature of IPF, the recognition of the UIP pattern in the HRCT does not equate to the diagnosis of IPF as UIP pattern is nonspecific and is seen in other fibrotic lung diseases, including connective tissue diseases, chronic hypersensitivity pneumonitis, asbestosis, use of certain prescribed drugs (eg, nitrofurantoin), and in some patients with advanced stages of pulmonary sarcoidosis.

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While the recent (2018) evidence-based clinical practice guideline (the American Thoracic Society—the European Respiratory Society—the Japanese Respiratory Society—the Asociacion Latinoamericana de Torax) empasizes the need for multidisciplinary discussions for the diagnosis of IPF and has made a conditional recommendation to obtain surgical lung biopsy (SLB) in patients whose HRCT images demonstrate the pattern as "probable for UIP" (the criteria of this pattern has been recently clarified) to ascertain the diagnosis of IPF, it is hoped that the recognition of the probable UIP pattern in the HRCT images and the recommended multidisciplinary discussions among experts in ILD will minimize the need for subjecting patients to SLB to ascertain the diagnosis of IPF by histopathology confirmation or elimination of UIP pattern in the SLB.

Despite the recognition of the specific HRCT image patterns that distinguishes the individual subgroups of IIP, specifically the UIP pattern, the physician must realize that a very thorough clinical evaluation and assessment is the key diagnosis procedure that might eliminate the need for subjecting some patients to surgery for obtaining that relatively larger lung specimen for diagnostic purposes. Without the clinical information gathered by the clinician, the histologic and radiologic features of the patterns in the lung are nonspecific. The diagnostic process should therefore start with the elicitation of a very thorough and extensive medical history that must include family medical history, environmental exposures, medications; there is no place for casual history-taking when evaluating a patient with ILD.

In essence, HRCT of the chest is an essential component of diagnostic evaluation of patients suspected to have ILD. It is imperative for the clinician confronted with the patient manifesting ILD currently to eliminate connective tissue diseases, environmental exposures known to cause ILD and it is only in the appropriate clinical setting, a diagnosis of IPF can be ascertained with the HRCT features of UIP without the need to clarify/confirm the histopathology in the lung biopsy.

It is hoped that in future, studies will demonstrate that clinical and/or imaging biomarkers, have very high specificity and sensitivity to ascertain an accurate diagnosis of IIP/ILD. Studies with machine learning for image patterns and/or molecular signatures in biological samples may in future reveal convincing data to eliminate the subjective variability in one's ability to elicit the medical history and perform physical examination as well as interpretation of the UIP patterns in HRCT scans or histopathology that have been in longstanding clinical use.

This issue of the Seminars in Roentgenology gives the readership a good insight and understanding of the importance and significance of recognition of patterns on the HRCT scan of the chest and for the need of multidisciplinary discussions (that includes the radiologist) to ascertain a specific diagnosis/In addition, the contributions by experts provoke new questions, new ideas, and perspectives for the future investigator and this includes machine learning for diagnosis and assessment of treatment response by quantification of the extent of fibrosis by imaging techniques.

I am hopeful that the reader will find this special on imaging ILD, edited by Dr Sudhakar Pipavath as a guest editor, useful and provoke useful clinical studies incorporating evolving knowledge of patterns in ILD and enhance the clinical understanding and management.

Ganesh Raghu, MD, FCCP, FACP
Division of Pulmonary, Sleep & Critical Care Medicine
University of Washington (UW)
Seattle, WA
E-mail address: graghu@uw.edu