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Preface

Interstitial Lung Disease: Idiopathic Interstitial Pneumonia

Ganesh Raghu, MD
Guest Editor

In the absence of infection, several acute and chronic lung disorders with variable degrees of pulmonary inflammation and fibrosis are commonly referred to as interstitial lung disease (ILD). Diffuse infiltrative pulmonary disease is perhaps a more appropriate descriptive term for this heterogeneous group of lung diseases, because the term *interstitium* actually refers to the microscopic anatomic space bounded by the basement membranes of epithelial and endothelial cells. It is in this interstitial space that the fibroblast-like cells (eg, connective tissue cells, myofibroblasts, mesenchymal cells) and extracellular matrix (eg, collagens, elastin, proteoglycans) are present. ILD clearly involves epithelial, endothelial, and mesenchymal cells, macrophages and recruited inflammatory cells, secreted proteins, and matrix components within the alveolar walls, and the disease process extends into the alveolar space, acini, bronchiolar lumen, and bronchioles. Thus the entire pulmonary parenchyma is involved in "interstitial" lung disease. Despite this misnomer, it seems appropriate to continue to use the term *interstitial lung disease* as such.

Over 150 agents and clinical situations have been associated with ILD, and this list continues to grow. In essence, the list becomes enormous if one considers the several pharmaceutical and environmental agents and clinical situations as individual causes or clinical conditions. One simple way to remember this list is to divide it into seven main entities: ILD associated with (1) occupational and environmental factors (inhalation cause), (2) collagen vascular diseases, (3) granulomatous lung disease of known and unknown causes (eg, hypersensitivity pneumonitis, sarcoidosis), (4) inherited diseases, (5) iatrogenic/drug induced, (6) certain specific entities (eg, pulmonary Langerhans cell granulomatosis, lymphangioleiomyomatosis), and (7) idiopathic interstitial pneumonia (IIP). Idiopathic pulmonary fibrosis (IPF) is a subgroup of IIP. Recent international consensus statements have clarified these entities, and IPF is clearly acknoweledged as a disease with fatal prognosis with an effective regimen yet to be determined. In the era of acquired immunosuppression (drugs or virus induced) and transplantation, diseases involving the pulmonary parenchyma in this clinical setting is generally not included in the category of ILD per se; the opportunistic lung infections, neoplasm, transplant-related immunologic problems, and so forth are confounding factors that influence pathogenesis in the management of lung disease occurring in the immunosuppressed host.

The clinicians of this era must be aware of the several distinguishing features of ILD and IIP and recognize that IPF is a distinct clinical entity when confronted with a patient who has ILD. It is essential to make an accurate diagnosis for appropriate therapeutic interventions and discuss prognosis with

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the patient. The physician must realize that thorough clinical evaluation and assessment is the key diagnostic 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 features in the lung biopsy is meaningless, because it is nonspecific. The diagnostic process should start with the elicitation of a thorough and extensive medical history that must include family medical history; there is no place for casual history-taking when evaluating a patient who has ILD. This is emphasized and discussed in the first article of this issue.

In this and the previous issue of the *Clinics in Chest Medicine*, ILD is discussed in a series of articles written by experts who are actively working in the forefront of this field. Articles in the previous issue discussed the clinical spectrum of ILD other than the IIPs.

This issue focuses on the IIPs, which are separately discussed by experts: "Imaging of the Chest" (Pipavath and Godwin), "Bronchoalveolar Lavage" (Meyer), "Pathology of Interstitial Lung Diseases" (Leslie), "Idiopathic Pulmonary Fibrosis" (Raghu and Chang), "Nonspecific Interstitial Pneumonia" (Nagai, et al), "Respiratory Bronchiolitis Associated with Interstitial Lung Disease and Desquamative Interstitial Pneumonia" (DuBois and Wells), "Cryptogenic Organizing Pnuemonia" (Cordier), "Acute Interstitial Pneumonia" (Vourlekis), "Pathogenesis of Pulmonary Fibrosis" (Noble and Homer), "New Antifibrotic Therapies" (Brown and Raghu), and "Lung Transplantation for Interstitial Lung Disease" (Bhorade and Lu).

Overall, an international blend of expertise, ideas, and current status of our understanding of ILD and IIP is provided for the reader. In addition, differences of approach and opinion are given to provoke new questions, fresh ideas, and perspectives for the future investigator. Acknowledging that the articles are written by different experts in the field of ILD/IIP, some overlap of the subject matter is inevitable. Rather, the overlap along with the differences in the individuals' style makes the reading of this issue, it is hoped, a particularly enjoyable one.

As Guest Editor, I am most grateful to all the authors for gladly contributing without hesitation. I am deeply indebted to their cooperation and patience. I sincerely hope that readers will find this issue useful in their clinical practice.

Ganesh Raghu, MD
Chief, Chest Clinic
Director, Interstitial Lung Disease
Sarcoid and Pulmonary Fibrosis Program
Medical Director, Lung Transplant Program
Professor of Medicine
University of Washington Medical Center
1959 East Pacific, Campus Box 356522
Seattle, WA 98195-6522, USA
E-mail address: graghu@u.washington.edu