

REVIEW

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Diagnosis and management of interstitial lung disease

Keith C Meyer

Abstract

The complex tasks of making a confident diagnosis of a specific form of interstitial lung disease (ILD) and formulating a patient-centered, personalized management plan in an attempt to achieve remission or stabilization of the disease process can pose formidable challenges to clinicians. When patients are evaluated for suspected ILD, an accurate diagnosis of the specific form of ILD that a patient has developed must be made to provide the patient with useful prognostic information and to formulate an appropriate management plan that can relieve symptoms and restore or significantly improve quality of life. A well-performed patient history and physical examination provides invaluable information that can be combined with appropriate laboratory testing, imaging, and, if needed, tissue biopsy to reach a confident ILD diagnosis, and high-resolution computed tomography (HRCT) of the thorax is usually a key component of the diagnostic evaluation. If treatment is indicated, many forms of ILD can respond significantly to immunosuppressive anti-inflammatory therapies. However, ILD accompanied by extensive fibrosis may be difficult to treat, and the identification of an effective pharmacologic therapy for idiopathic pulmonary fibrosis (IPF) has remained elusive despite the completion of many phase 3 clinical trials over the past decade. Nonetheless, patients with IPF or advanced forms of non-IPF ILD can benefit significantly from detection and treatment of various co-morbid conditions that are often found in patients (especially the elderly patient), and supportive care (oxygen therapy, pulmonary rehabilitation) can have a beneficial impact on quality of life and symptom palliation. Finally, lung transplantation is an option for patients with progressive, advanced disease that does not respond to other therapies, but only a relatively small subset of patients with end-stage ILD are able to meet wait listing requirements and eventually undergo successful lung transplantation.

Keywords: Interstitial lung disease; Idiopathic pulmonary fibrosis; Therapeutics; Diagnosis

Introduction

Well over one hundred different forms of interstitial lung disease (ILD) have been described (see Table 1 for major categories). These diffuse infiltrative lung disorders are typically characterized by the presence of inflammation and altered lung interstitium, and specific forms of ILD can be differentiated from one another when clinical data, radiologic imaging, and pathologic findings (if lung biopsy is needed) are combined to reach a confident diagnosis [1,2]. The histopathologic changes in the lungs of patients with ILD can range from granulomatous inflammation without parenchymal fibrosis in patients with sarcoidosis to extensive pulmonary fibrosis with architectural distortion of the lung in patients with

idiopathic pulmonary fibrosis (IPF). Some forms of ILD have been linked to specific genetic abnormalities (e.g. Hermansky-Pudlak syndrome, familial pulmonary fibrosis), and a number of gene variants have been associated with an increased risk to develop ILD disorders such as IPF, sarcoidosis, or chronic beryllium disease (CBD). Interstitial lung disease can also complicate connective tissue disorders (CTD), and lung histopathologic changes can have features of usual interstitial pneumonia (UIP) or non-specific interstitial pneumonia (NSIP) patterns in CTD-associated ILD [3].

Successful management of patients with ILD is dependent upon establishing an accurate and specific diagnosis [1,2]. Because various forms of ILD such as IPF, non-IPF forms of idiopathic interstitial pneumonia (IIP), CTD-ILD, and hypersensitivity pneumonitis (HP) can have similar clinical presentations, patients with

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