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REVIEW

Idiopathic pulmonary fibrosis: Early detection and referral

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Summary
Idiopathic pulmonary fibrosis (IPF), a devastating progressive interstitial lung disease (ILD) with no known cause or cure, is the most common and deadly of the idiopathic interstitial pneumonias. With a median survival of 3–5 years following diagnosis, IPF is characterized by a progressive decline in lung function and quality of life in most patients. Vigilance among clinicians in recognizing IPF early in the disease course remains critical to properly caring for these patients, as this provides the widest range of management options. When IPF is suspected, a multidisciplinary evaluation (MDE) by a clinician, radiologist and pathologist with ILD expertise should occur, as this improves diagnostic agreement in both community and academic settings. When community MDE is not possible, or diagnostic doubt exists, referral to an ILD center should be considered. ILD center referral may also provide access specialized care, including clinical trials and lung transplantation, and should be considered for any patient with an established diagnosis of IPF.

Contents

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Introduction

The interstitial lung diseases (ILDs) are a heterogeneous group of diffuse parenchymal lung disorders with similar clinical, radiographic, physiologic or pathologic features [1]. ILDs include disorders of known cause, and those with unknown etiology, referred to as the idiopathic interstitial pneumonias (IIPs) [2]. The most common and deadly of the IIPs is idiopathic pulmonary fibrosis (IPF) [3], defined as a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause, occurring primarily in older adults, and limited to the lungs [4]. IPF has a median survival time of 3—5 years after diagnosis, and is characterized by progressive decline in lung function and quality of life in most patients [3,5—7]. Despite an evolving understanding of the underlying disease process, the pathogenesis of IPF remains unknown.

Epidemiology

The global prevalence of IPF is difficult to establish and reflects the diagnostic complexity of the disease and the substantial resources needed to establish the diagnosis. These issues, along with heterogeneity in study design and case finding methodology, have resulted in variable epidemiologic data over the last several decades [4,8]. Two recent population studies in the US reported an IPF incidence of 6.8—8.8 and 16.3—17.4 cases per 100,000 person years using narrow and broad case definitions, respectively. These studies found the prevalence of IPF to be 14.0—27.9 (narrow case definitions) and 42.7—63.0 (broad case definitions) per 100,000 persons [9,10]. Recent epidemiologic studies from the UK demonstrate that the incidence of IPF has been rising over the last decade [11,12]. IPF has been described in all ethnic groups and in both rural and urban settings [7,13]. Affecting males predominantly, most cases are diagnosed after age 60 and incidence increases with age. [7,9—12].

Early diagnosis

Evolution of the diagnostic criteria for IPF has resulted in abandonment of the major and minor criteria set by the 2000 ATS/ERS consensus statement [14]. The latest international guidelines require exclusion of other known causes of ILD and the presence of a usual interstitial pneumonia (UIP) pattern on high-resolution computed tomography (HRCT) in patients for whom a surgical lung biopsy (SLB) has not been performed. In patients failing to demonstrate a typical UIP pattern on HRCT, IPF may still be diagnosed based on specific combinations of HRCT and SLB patterns (Table 1) [4]. This diagnostic complexity makes misdiagnosis and delayed diagnosis common [15]. A recent survey showed that a majority of patients with IPF reported seeing several physicians, and waiting over a year, before receiving the correct diagnosis [16]. Careful attention to historical clues, physical exam and other diagnostics can help establish the diagnosis early and provide those with IPF the full breadth of options available to them.

History

Any adult over age 50 presenting with unexplained dyspnea on exertion and cough should prompt consideration of IPF [4]. Dyspnea in this disease tends to arise insidiously, often over 6 months or more, and progresses steadily [17]. Over 80% of patients report a non-productive, often intractable cough that can be debilitating and refractory to antitussive medications [18—20]. Chest pain is uncommon in IPF and because the disease is limited to the lungs, the presence of systemic inflammatory symptoms, including fever, rash, weight loss, myalgia or arthralgia, makes IPF unlikely and should prompt consideration of an alternative diagnosis.

Cigarette smoking has been strongly linked to IPF, especially in those with greater than 20 pack-year history [21—23]. Hiatal hernia and gastroesophageal reflux disease (GERD) have also been linked to IPF [24—26]. Environmental and industrial exposures associated with IPF include