Imaging of Idiopathic Pulmonary Fibrosis

Joanna E. Kusmirek, MD[a,*], Maria Daniela Martin, MD[b], Jeffrey P. Kanne, MD[b]

KEYWORDS

• Idiopathic interstitial pneumonia • Idiopathic pulmonary fibrosis • Usual interstitial pneumonia • HRCT

KEY POINTS

- High-resolution computed tomography (HRCT) findings are crucial in the multidisciplinary diagnosis of idiopathic pulmonary fibrosis (IPF).
- If the HRCT pattern is consistent with usual interstitial pneumonia (UIP) and the clinical presentation is concordant, lung biopsy is not indicated.
- Most patients with a UIP pattern on HRCT have IPF, but the pattern can also occur with connective tissue disease, asbestos exposure, and drug toxicity, so thorough clinical evaluation is necessary.
- Mimics of UIP on HRCT include nonspecific interstitial pneumonia, fibrotic hypersensitivity pneumonitis, fibrosing sarcoid, asbestosis, and drug reaction.
- HRCT carries prognostic value in IPF and plays an important role in longitudinal monitoring as well as detection of complications such as infection, acute exacerbation, and cancer.

INTRODUCTION

Idiopathic interstitial pneumonias (IIPs) include a group of diffuse parenchymal lung diseases with variety of clinicopathologic presentations. The recent classification by the American Thoracic Society (ATS)/European Respiratory Society (ERS) from 2013,[1] divides IIPs into (**Box 1**): (1) chronic fibrosing IIPs, including idiopathic pulmonary fibrosis (IPF) and idiopathic nonspecific interstitial pneumonia (NSIP); (2) smoking-related IIPs, including respiratory bronchiolitis–associated interstitial lung disease (RB-ILD) and desquamative interstitial pneumonia (DIP); (3) acute or subacute IIPs, including cryptogenic organizing pneumonia (COP) and acute interstitial pneumonia; and (4) rare IIPs, including lymphoid interstitial pneumonia and idiopathic pleuroparenchymal fibroelastosis.

The ATS/ERS 2013 update on IIPs also proposed a classification based on disease behavior because the IIPs represent a heterogeneous group of diseases with different prognoses[1] (**Table 1**).

IPF is the most common of the IIPs and is a chronic, progressive, fibrosing lung disease of unknown cause characterized by the histopathologic pattern of usual interstitial pneumonia (UIP).[2]

The prognosis is poor in most cases, with median survival ranging from 2.5 to 3.5 years.[3–5] However, progression and prognosis can be variable, and although most patients experience rapid progression, some patients remain fairly stable.[6–8] The prevalence of IPF is estimated to range from 14 to 42.7 per 100,000 in the United States, and from 1.25 to 23.4 per 100,000 in Europe; it is higher among men than women. The annual incidence is estimated at be 6.8 to 16.3 per 100,000 in the United States and 0.22 to 7.94 per 100,000 in Europe.[9–11]

Cigarette smoking is strongly associated with IPF, with up to two-thirds of patients with IPF being

Disclosures: Consultant for Parexel International, Genentech (J.P. Kanne).
[a] Department of Radiology, Virginia Commonwealth University, 1250 East Marshall Street, Richmond, VA 23298, USA; [b] Department of Radiology, University of Wisconsin, 600 Highland Avenue, Madison, WI 53792-3252, USA
* Corresponding author.
E-mail address: joanna.kusmirek@vcuhealth.org

Radiol Clin N Am 54 (2016) 997–1014
http://dx.doi.org/10.1016/j.rcl.2016.05.004
0033-8389/16/$ – see front matter

Box 1
Classification of IIPs according to the official ATS/ERS (2013)

Chronic fibrosing IIPs

Idiopathic pulmonary fibrosis

Idiopathic nonspecific interstitial pneumonia

Smoking-related IIPs

Respiratory bronchiolitis–associated interstitial lung disease

Desquamative interstitial pneumonia

Acute or subacute IIPs

Cryptogenic organizing pneumonia

Acute interstitial pneumonia

Rare IIPs

Idiopathic lymphoid interstitial pneumonia

Idiopathic pleuroparenchymal fibroelastosis

Data from Travis WD, Costabel U, Hansell DM, et al. An official American Thoracic Society/European Respiratory Society statement: update of the international multidisciplinary classification of the idiopathic interstitial pneumonias. Am J Respir Crit Care Med 2013;188(6):733–48.

current or former smokers. No evidence of direct causation has been established; however, the highest risk of developing IPF exists for patients who have most recently quit.[12] Smoking also adversely affects survival in IPF.[13]

Table 1
Classification of IIP based on disease behavior according to the American Thoracic Society/European Respiratory Society 2013 update.

Reversible and self-limited disease	Many cases of RB-ILD
Reversible disease with risk of progression	Cellular NSIP and some fibrotic NSIP, DIP, COP
Stable with residual disease	Some fibrotic NSIP
Progressive irreversible disease with potential for stabilization	Some fibrotic NSIP
Progressive irreversible disease despite therapy	IPF, some fibrotic NSIP

Data from Travis WD, Costabel U, Hansell DM, et al. An official American Thoracic Society/European Respiratory Society statement: update of the international multidisciplinary classification of the idiopathic interstitial pneumonias. Am J Respir Crit Care Med 2013;188(6):733–48.

CLINICAL PRESENTATION

IPF should be considered in the differential diagnosis of unexplained chronic exertional dyspnea, dry cough, or both in adults, especially older patients, because the typical age of presentation is the sixth and seventh decades of life.[2] Physical examination may reveal inspiratory crackles and digital clubbing. Pulmonary function tests typically show low lung volumes with restrictive physiology and reduced carbon monoxide diffusion capacity in the lung (DLco). There are several issues that should be addressed by clinicians while evaluating patients for IPF, as listed in **Table 2**.

Gastroesophageal reflux disease (GERD) and aspiration occur frequently in patients with IPF[15,16] and are often asymptomatic.[17] Survival and functional advantage are seen in those treated for GERD as opposed to those who are not.[18] It is debated whether or not GERD contributes to the development of IPF or is an effect (possibly by altered mechanics).[17] GERD was also proposed as a causative/contributing factor to the development of acute exacerbations in IPF.[19]

IMAGING TECHNIQUE/PROTOCOLS

Pulmonary fibrosis on chest radiography most commonly manifests as reticulation and linear opacities[20] along with decreased lung volumes (**Fig. 1**). Linear opacities are nonspecific and can be seen in other conditions, including emphysema, pulmonary Langerhans cell histiocytosis, and lymphangioleiomyomatosis; however, lung volumes are preserved or increased with these diseases. When pulmonary fibrosis is suspected clinically or on chest radiographs, high-resolution computed tomography (HRCT) of the chest should be obtained (**Table 3**). Because of advances in computed tomography (CT) technology, the distinction between conventional CT and HRCT have become blurred. Current multidetector CT scanners acquire volumetric data compatible with HRCT images and allow multiplanar reformats,[21] which greatly help in evaluation, especially in assessment of honeycombing (**Fig. 2**). Volumetric CT acquisition with multidetector CT is generally preferred to noncontiguous imaging, despite slightly higher radiation exposure.[22–24] The effective radiation dose from chest CT is usually less than 5 mSv,[23] with doses of approximately 2 mSv or less readily achievable on current scanners. In the lungs, the natural high contrast between air and tissue allows low-dose HRCT imaging. Using low-dose technique and newer reconstruction algorithms, particularly iterative reconstruction (IR), can reduce the dose by up to