

Idiopathic Pulmonary Fibrosis (IPF) Diagnosis and Workup

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Abstract

Idiopathic pulmonary fibrosis (IPF), is a rare and often fatal lung disease. The clinical presentation is nonspecific, after questioning the patient about dusty jobs, or medication intake and after discarding collagen diseases by laboratory tests and discarding sarcoidosis and histiocytosis. We should think about the possibility of IPF.

The Gold Standard for diagnosis is high resolution computerized tomography. When it presents with honeycombing at the basis of the lungs, it is typical of Usual interstitial pneumonia (UIP) and non-need for lung biopsy. If not we need to do biopsy.

Keywords: *Idiopathic Pulmonary Fibrosis (IPF); Usual Interstitial Pneumonia (UIP)*

Introduction

Idiopathic pulmonary fibrosis (IPF), is a rare and often fatal lung disease whose cause is unknown and clinical symptoms are nonspecific. Normally patients present with breathless during exercise and dry cough. Such symptoms are not specific to IPF and could be present in other cardio-respiratory diseases. We should obtain a comprehensive clinical history, including medication intake, occupation, hobbies, and environmental exposure history. Questioning about other systems like cardiac or kidney, are indispensable aiming at excluding other entities of interstitial lung disease.

The diagnosis of idiopathic pulmonary fibrosis now requires the following:

- The exclusion of occupational disease through clinical history, lung functions and pulmonary imaging (e.g. Asbestosis or silicosis).
- The exclusion of drug toxicity e.g. Methotrexate.
- Exclude collagen diseases, vasculitis, sarcoidosis.... etc. This needs performing the appropriate tests.
- High resolution CT scan HRCT: There are four diagnostic categories based on high-resolution tomography (HRCT) of the lung: usual interstitial pneumonia (UIP) pattern, probable UIP pattern, indeterminate pattern and alternative diagnosis.
- For patients with UIP pattern on CT scan there is no need to confirm the diagnosis with lung biopsy.
- If any category other than UIP pattern is present lung biopsy is required to differentiate IPF from other idiopathic interstitial pneumonias.

Imaging studies

Chest radiography

There is nonspecific radiological pattern for idiopathic pulmonary fibrosis. The typical findings are peripheral reticular opacities and Honeycombing especially at the basis of the lungs. Although lower lobe volume loss can also be seen.

High-resolution CT scan

High-resolution computed tomography (HRCT) findings are an essential component of the diagnostic pathway of idiopathic pulmonary fibrosis. On HRCT images, idiopathic pulmonary fibrosis is characterized by sub pleural patchy, and bibasilar reticular opacities, and baso-posterior honey combing. If all that is present, diagnostic for IPF could be retained without biopsy. Practically, there are four diagnostic categories based on high-resolution tomography (HRCT) of the lung: usual interstitial pneumonia (UIP) pattern, probable UIP pattern, indeterminate pattern and alternative diagnosis.

UIP pattern is as follows:

- Subpleural and basal predominant distribution as described in the introduction.
- Honeycombing with or without peripheral traction bronchiectasis or bronchiolectasis.

Probable UIP pattern is as follows:

- Subpleural and basal predominant; distribution is often heterogeneous.
- Reticular pattern with peripheral traction bronchiectasis or bronchiolectasis.
- May have mild ground-glass opacities.

Indeterminate pattern for UIP is as follows:

- Subpleural and basal predominant.
- Subtle reticulation; may have mild ground-glass opacities or distortion („early UIP pattern”).
- CT features and/or distribution of lung fibrosis that do not suggest any specific etiology („truly indeterminate”).

Alternative diagnosis pattern is findings suggestive of another diagnosis based on [1] CT features and predominant distribution.

Surgical lung biopsy

A surgical lung biopsy specimen can be obtained through either an open lung biopsy or video-assisted thoracoscopic surgery (VATS). A surgical lung biopsy provides the best approach which to distinguish usual interstitial pneumonia from other interstitial pneumonias. VATS is preferred because it is associated with less post-surgical complications and less hospital stay than with open lung biopsy.

Given specificity of HRCT for the recognition of histopathologic UIP pattern, surgical lung biopsy is not recommended In patients with UIP pattern on HRCT.

However, for patients with newly diagnosed interstitial lung disease of unknown cause who are clinically suspected of having idiopathic pulmonary fibrosis and have an HRCT pattern of probable UIP, indeterminate, We should perform surgical biopsy.

Other nonspecific tests

Laboratory studies

A routine laboratory evaluation is not helpful except for its role in ruling out other causes of diffuse parenchymal lung disease. Serological testing is important to exclude connective tissue disease. Polycythemia is a rare finding despite the frequency of chronic hypoxemia. Elevation of systemic inflammatory markers (i.e. erythrocyte sedimentation rate or C-reactive protein level) or the presence of hypergammaglobulinemia is found in IPF yet such findings are non-diagnostic.

There is non need to measure serum biomarkers such as matrix metalloproteinase (MMP)-7, surfactant protein D (SPD), chemokine ligand (CCL)-18, or Krebs von den Lungen-6 to confirm Or discard The diagnosis of IPF.

Pulmonary function testing

The characteristic findings when performing pulmonary function tests in patients with idiopathic pulmonary fibrosis are a restrictive ventilatory defect and a reduced diffusion capacity for carbon monoxide.

6-Minute walk testing

The 6MWT assess exercise capacity, and severity of the disease.

Bronchoscopy

Bronchoscopy with BAL and/or transbronchial biopsy can be used for differential diagnosis. BAL fluid analysis can be useful to exclude other alternative diagnoses. Consequently, cellular analysis of BAL fluid is suggested for patients with HRCT pattern of probable UIP. Increased numbers of neutrophils in BAL fluid are found in 70 - 90% of all patients with idiopathic pulmonary fibrosis, and less than 30% lymphocytosis in BAL fluid is important in distinguishing idiopathic pulmonary fibrosis from non-idiopathic pulmonary fibrosis diagnoses [2-4].

Conclusion

Idiopathic pulmonary fibrosis is confirmed clinically and by CT scan with honey combing. If the radiologic presentation is not typical we need biopsy.

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