Idiopathic Pulmonary Fibrosis

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Interstitial Restrictive Lung Disease
Interstate-sign and Lungs Restricted by a Belt
Idiopathic Pulmonary Fibrosis (IPF) is a chronic, progressive fibrosing interstitial lung disease of unknown cause, primarily affecting adults over 50 years old. It is characterized by the histopathologic and radiologic pattern of usual interstitial pneumonia (UIP), leading to a decline in lung function. IPF involves repeated epithelial injury and dysregulated repair mechanisms, leading to fibroblast proliferation and extracellular matrix deposition. This results in progressive scarring, loss of alveolar architecture, decreased lung compliance, and impaired gas exchange.

Age over 50 years, Males, Tobacco
50-Cent Man and Tobacco
Although the trigger for the disease is unknown, there are three known risk factors:

1. Age > 50 years
2. Male Gender
3. Tobacco Use

CLINICAL FEATURES

Dyspnea and Cough
Disc-P-lungs and Coughing-coffee-pot
One of the features of restrictive lung diseases and, therefore, idiopathic pulmonary fibrosis is dyspnea that increases with exercise and cough.

Fatigue
Sleepy-guy
As the disease progresses and dyspnea increases, the patient begins to feel fatigued. Fatigue in IPF results from hypoxemia, increased breathing effort, inflammation, and poor sleep. It worsens as dyspnea progresses, reducing physical activity and quality of life. Early intervention with oxygen therapy, pulmonary rehab, and psychological support can help manage fatigue effectively.

Crackles
Crackers
On auscultation, patients with IPF may have crackles, heard at the lung bases bilaterally.

Digital Clubbing
Club causing Clubbing
A clinical feature in various pulmonary diseases, such as idiopathic pulmonary fibrosis, is digital clubbing.

DIAGNOSIS

Bronchiectasis
Broccoli-with-tassels
Another feature that can be found in the CT of patients with idiopathic pulmonary fibrosis is traction bronchiectasis, which appears as the lung displays more fibrosis. Those zones pull on normal lung areas.

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Honeycomb Pattern

Beehive Honeycomb

Diagnosis of idiopathic pulmonary fibrosis is done with various studies, such as a CT, after other causes of pulmonary fibrosis have been ruled out.

A distinctive, but not exclusive, finding in CT of patients with idiopathic pulmonary fibrosis are basal and subpleural heterogenous infiltrates in a honeycomb pattern. Similarly, in CXR, patients display reticular or reticulonodular infiltrates in the bases (honeycomb lung).

Restrictive Pattern

Restrictive-belt on Lungs

In spirometry, the pattern seen is restrictive, with decreased total lung capacity, forced vital capacity, and forced expiratory volume in the first second. Since all volumes decrease, it is important to remember that the FEV1/CVF ratio is usually normal, which differentiates this pattern from obstructive lung disease.

Another feature is a decrease in diffusing capacity for carbon monoxide.

COMPLICATIONS

Respiratory Failure

Dead Lungs

As fibrosis progresses, there is less functional lung tissue, and capillary resistance increases. This is further promoted by hypoxia, which leads to vasoconstriction in pulmonary capillaries. All of this gives rise to pulmonary hypertension and, eventually, respiratory failure.

Right Heart Failure

Right Dead Heart

Pulmonary hypertension and fibrosis lead to an increase in the force that the right ventricle has to exert to pump lungs towards the lungs. This increase causes the right heart to adapt and dilate, but as the disease progresses, right heart failure ensues. This failure, in turn, can lead to arrhythmias such as atrial fibrillation and atrial flutter, which can further worsen right heart function.

TREATMENT

Oxygen

O2-tank

Idiopathic pulmonary fibrosis is a progressive disease that eventually causes respiratory failure and death. The only known treatment today is a lung transplant. As patients deteriorate, supplemental oxygen is used to alleviate symptoms.

Antifibrotic Medications

Stop-sign Med-bottle Fibrous-Sack

Patients with IPF are treated with antifibrotic medications, such as Pirfenidone and Nintedanib, to slow disease progression and improve quality of life. These drugs do not cure IPF but help delay lung function decline.

Lung Transplant

Lungs Train-plant

Ultimately, patients with idiopathic pulmonary fibrosis end up in respiratory failure, which makes lung transplant the only definitive therapeutic option.

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