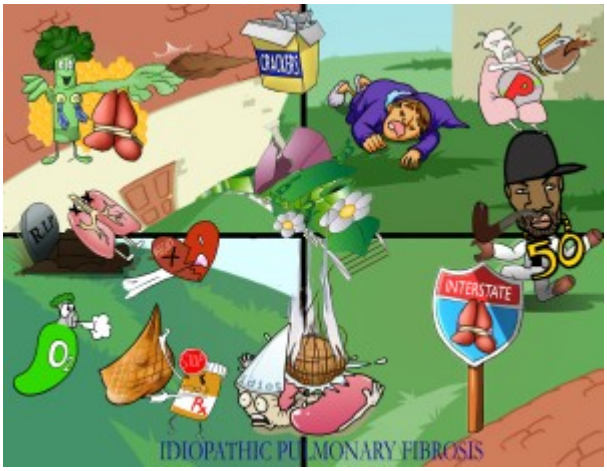
Idiopathic Pulmonary Fibrosis



PLAY PICMONIC

Interstitial Restrictive Lung Disease

Interstate-sign and Lungs Restricted by a Belt

Idiopathic pulmonary fibrosis is an interstitial restrictive lung disease without a known cause.

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.

Crackles

Crackers

On auscultation, patients with IPF may have crackles.

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.

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

Antifibrotic medications, like Pirfenidone and Nintedanib, are used to help slow down disease progression.

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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