

# Pleuroparenchymal Fibroelastosis

- PPFE = Rare form of interstitial lung disease characterized by
- Fibrosis that predominantly affects the upper lobes of the lungs
- Involving both the pleura and the subpleural lung parenchyma.
- Increasingly recognized as a distinct entity among idiopathic interstitial pneumonias,

# Etiology

- **Idiopathic:** Many cases are of unknown cause.
- **Secondary Associations:** PPFE has been linked to previous bone marrow, lung, or other organ transplantation, prior radiotherapy, some chemotherapies, recurrent lung infections, and occasionally autoimmune or connective tissue diseases.
- **Other Factors:** Occupational exposures (e.g., asbestos, aluminum) and genetic predispositions have also been reported, but smoking is not considered a risk factor

# High-Resolution CT (HRCT)

- **Bilateral apical pleural thickening:** This is the hallmark finding, often extending caudally from the lung apex.
- **Subpleural fibrosis:** Dense, well-demarcated subpleural consolidation with coarse reticulation, predominantly in the upper lobes.
- **Traction bronchiectasis/bronchiolectasis:** Commonly seen within fibrotic areas.
- **Architectural distortion:** Manifested as upper lobe volume loss and retraction of the hilar structures.
- **Peripheral consolidation:** May be present in the affected regions.

# High-Resolution CT (HRCT)

- **Reticular abnormalities:** Including thickening of interlobular septa.
- **Pneumothorax:** Spontaneous pneumothorax is a recognized complication, more likely in advanced disease.
- **Platithorax:** Flattening of the chest wall with deep suprasternal notch and/or posterior tracheal retraction.
- **Sharp demarcation:** There is often a clear boundary between affected and unaffected lung tissue.
- **Lower lobe sparing:** PPFE rarely extends into the lung bases; any lower lobe fibrosis usually suggests a coexisting interstitial lung disease such as UIP or NSIP

# Additional Imaging Features

- **Lymphadenopathy:** Mediastinal and/or axillary lymphadenopathy may occasionally be seen.
- **Honeycombing and bullae:** These may develop in advanced cases, contributing to the risk of pneumothorax.
- **Nuclear medicine:** Rarely, PPFE can present as a hypermetabolic nodule on FDG-PET









