Interstitial Lung Disease ILD

- The interstitium of the lung is the area between the alveolar sacs and the blood vessels
- the interstitium has fibroblasts and collagen and elastin fibers in addition to lymphatics
- this area is very thin and cannot be identified in a CT scan but when we have ILD the membrane become thickened and it will appear on the CT
- ILD results from Repeated exposure to inflammatory agents or imperfect repair of damaged tissue
- it is not an airway disease (not associated with sputum)
- ILD are characterized clinically by respiratory symptoms associated with :
    - Radiologically diffused infiltrates
    - Histologically by distortion of the gas exchanging units
    - Physiologically by restriction of lung volumes and impaired oxygenation (patients are usually hypoxic)
- Secondary pulmonary lobule : group of alveoli surrounded by interstitium enclosed by lymphatic vessels with an artery passing from the center and a vein on the side
- in ILD the lymphatic surrounding become full of fibroblasts and collagen from the interstitium leading to the fibrous thickening seen on CT
- Symptoms
    - mainly Age > 70 years
    - Exertional dyspnea
    - nonproductive cough (Rarely sputum production, hemoptysis, or wheezing)
    - symptoms of pulmonary hypertension like dizziness or palpitations
- Duration
    - Acute : symptoms less than a week
        - Infection
        - Acute Hypersensitivity Pneumonitis
        - Acute Eosinophilic Pneumonia
        - Drug reaction
        - Acute lupus pneumonitis
        - Diffuse Alveolar Hemorrhage
    - Chronic : more than 4 weeks : usually Idiopathic Pulmonary Fibrosis
- Idiopathic Interstitial Pneumonia is the main type & it includes
    - Acute Interstitial Pneumonitis : acute / poor response to steroids / complete recovery is possible
    - Idiopathic Pulmonary Fibrosis : insidious / poor response to steroids / complete recovery is not possible
- other types include granulomatous ILD (sarcoidosis), drug induced or rheumatoid ILD

- Occupational exposures : asbestos associated with construction workers and those who build ships and planes / silica / biomass fuels / birds especially pigeons / cotton factories
- Most common from of ILD is Idiopathic Pulmonary Fibrosis
    - characterized histologically by usual interstitial pneumonia UIP
    - associated with finger clubbing
    - mainly in smokers and old men
    - HRCT shows Subpleural, reticular infiltrates with basilar predominance and areas of honeycombing
- Hypersensitivity pneumonitis HP is highly associated with the birds and it affects the upper part of the lungs
- Sarcoidosis
    - Pathologic hallmark is noncaseating granulomas
    - mainly in age 20-50
    - Constitutional symptoms include weight loss, fatigue, fever and malaise
    - respiratory symptoms : productive cough, dyspnea, hemoptysis, hoarseness
    - Eyes : Uveitis, keratoconjunctivitis Sicca, Uveoparotid fever
    - Heart : arrhythmias
    - Skin : Lofgren Syndrome : erythema nodosum with hilar adenopathy in the lungs, arthralgias and fever
    - 75% of patients have Hepatic granulomas
    - serum ACE might be elevated
    - chest xrays show hilar nodes - reticular opacities - pulmonary fibrosis
    - treatment : Corticosteroids
- Physical examination
    - General : Cyanosis
    - Chest : Increased tactile and vocal fremitus
    - Dull percussion note
    - Bronchial breath sounds
    - Velcro Crackles usually fine end inspiratory
    - Pleural friction rub
    - Whispered pectoriloquy
    - V/Q mismatch
    - shunting
    - decreased DLCO
    - Decreased lung compliance
    - low FVC and normal FEV1/FVC ratio
    - reduced TLC

    - might see elevated JVP
- clinical findings
    - signs of lupus like malar rash and raynaud
    - signs of scleroderma like calcinosis and fish mouth
    - signs of rheumatoid arthritis like ulnar deviation
    - signs of dermatomyositis
- Lab test
    - Rheumatoid factor
    - ANA
    - Anti dsDNA
    - ENA
- Gold standard for diagnosis is High Resolution CT scan which shows the paraseptal thickening and sometimes nodules (in sarcoid)
- CXR shows bilateral Hilar enlargement and in a later stages shows fibrosis and infiltrates
- in asbestosis the particles usually accumulate in the pleura or the diaphragm which can cause pleural effusion, asymptomatic plaques or fibrosis (lower part of the lungs)
- asbestosis leads to high risk of cancers especially Mesothelioma
- Honeycombing appearance is suggestive of severe fibrosis resulting in Secondary Traction Bronchiectasis (mainly with IPF)
- Smoking related ILDs are associated with upper part of the lung and have Pulmonary Langerhan Histiocytosis (pulmonary nodules appearing on the CT)
- Drug Induced ILD : pleomycin / amiodarone / nitrofurantoin (for UTIs) / methotrexate
- ILDS
    - affecting lower part of lungs : IPF / Rheumatoid / scleroderma / asbestosis
    - affecting upper part of lungs : sarcoidosis / hypersensitivity pneumonitis / ankylosing spondylitis
- Management
    - Supplemental oxygen if patient is hypoxic
    - Pulmonary rehabilitation (physiotherapy)
    - Measures to relieve symptoms like dry irritant cough
    - Treatment of comorbid conditions like anemia, OSA, GERD, pulmonary hypertension, infectious complications
    - Vaccinations for pneumococcal infections
- We ALWAYS give patients drugs for GERD with or without symptoms like omeprazole or lanzoprozole
- treatment for IPF and Scleroderma : Antifibrotics like pirfenidone & nintedanib (they stop the progression not fix it) - giving steroids is lethal

- for patients with rheumatologic disorders or vascultic disorders like Wegner and any other form we use immunosuppressants and systemic steroids (can be used with sarcoidosis)
- asbestosis has no management
- Lung transplant
    - Lung transplantation is the only form of therapy that may improve quality of life and survival for patients with IPF
    - 5 year survival following lung transplantation for IPF or other forms of pulmonary fibrosis is approximately 50%