Interstitial Lung Disease

Primary

Idiopathic Interstitial Pneumonia

Subtypes Based on Pathologic Pattern

(UIP) Usual Interstitial Pneumonia - fibrosis, with relatively little inflammation, most common

(NSIP) Non-specific Interstitial Pneumonia

(DIP) Desquamative Intersitial Pneumonia

(LIP) Lymphoid Interstitial Pneumonia
slowly progressive dyspnea and cough, "velcro" crackles, clubbing , worse at the bases and periphery, Poor Prognosis

Granulomatous

Sarcoidosis
(Debate if this should be 1° or 2°)
Multisystem characterized by granulomatous inflammation

20-40 year olds, common in African Americans, Northern Europeans

Unknown stimulus activates T-helper lymphocytes, resulting in formation of non-caseating granulomas
Stage 0: Normal
Stage 1: Hilar lymphadenopathy
Stage 2: Hilar lymphadenopathy + lung infiltrates
Stage 3: Lung infiltrates (without lymphadenopathy)
Stage 4: Fibrosis

Lofgren's Syndrome
Lymphadenopathy, erythema nodosum and polyarthritis
Many patients also have fever, malaise and uveitis, very good prognosis

Other (Rare)

Lymphangiolei omyomatosis (LAM),

Pulmonary Langerhans cell histiocytosis (PLCH)

Eosinophilic pneumonia

Secondary

Congestive Heart Failure

Impaired cardiac function resulting in fluid leaking out of pulmonary capillaries into intersitial space

Infection

"Atypical" Pneumonia

Acute onset (<4 weeks) but some more insidious

Fever
Chills
Pleuritic Chest pain

Malignancy

Metastases to lung as nodules

Some present as ILD (Obstruction of Pulmonary Lymphatics vessels)

Most Adenocarcinomas (breast, lung, colon, stomach)

Ask:
History of Malignancy, Risk Factors, weight loss)

Hypersensitivity Pneumonia

Immunologic (hypersensitivity) reaction to inhaled antigen

Requires recurrent exposures over time, > 200 different organic antigens associated with HP

Presentation may be acute (resemble "atypical" pneumonia), subacute or chronic (may be indistinguishable from idiopathic ILDs)

Classic Examples
"Farmer's Lung" (thermophillic actinomycetes in moldy hay)

"Bird Fancier's Lung" (proteins from bird droppings)

Treatment (remove from antigen, prednisome may hasten resolution)

Connective Tissue Disease

Rheumatoid Arthritis, Systemic Lupus and Scleroderma

ILD may precede musculoskeletal symptoms or be an incidental finding

Pneumonconiosis

Lung disease (non-neoplastic) resulting from inhalation of inorganic and organic dusts

Intensity and duration of exposure
Individual sysceptibility
Nature and properties of the dust (size, toxicity, etc.)

Asbestos

ILD caused by asbestos exposure

Silicosis

ILD caused by inhalation of silicon dioxide

Coal

Drugs/Radiaition

Drugs:
rheumatoid
Amiiodarone
Nitrofurantoin
Methotrexate
Beomycin

Acute or Chronic Radiation Pneumonitis
4-12 weeks after exposure, 6-12 after. Affects lung within radiation field

Interstitial Lung Disease

- Interstitium: connective tissue surrounding vessels, airways, lymphatics and pleura
- Intersitial Lung Disease:
- Originate in the interstitium
- Infiltration with fluid, cells and/or connective tissue
  - Can cause
    - Impairment of lung mechanics (↓ complicance, ↑ WOB, ↓ lung volume)
    - Impairment of gas exchange (↓ lung volumes = ↓ SA for gas exchange, extra stuff impedes transfer of O2 from alveoli to capillaries)
- Can involve other lung tissues as well as diffuse parenchymal lung disease may be a more appropriate term
- Usually involves multiple parts of both lungs
- Pathologically: disease of the interstitium
- Radiologicallly: extra lines/dots (↑ interstitial markings)
- Clinically:
  - Progressive SOB
  - Cough (may be present)
  - Chest Pain
  - Wheezing not usually present
  - Physical Exam: crackles, O2, desat