INTERSTITIAL LUNG DISEASES

- Diffuse parenchymal lung diseases, often collectively referred to as the interstitial lung diseases (ILDs

Causes

IDIOPATHIC.?CAUSE

Connective tissue disease–associated (for example, rheumatoid arthritis,SLE)

environmental and occupational inorganic or organic dusts

Hypersensitivity pneumonitis (for example, farmer's lung,, bird fancier's lung)

Pneumoconioses (for example, silicosis)

Drug-induced (for example, chemotherapeutic agents,)

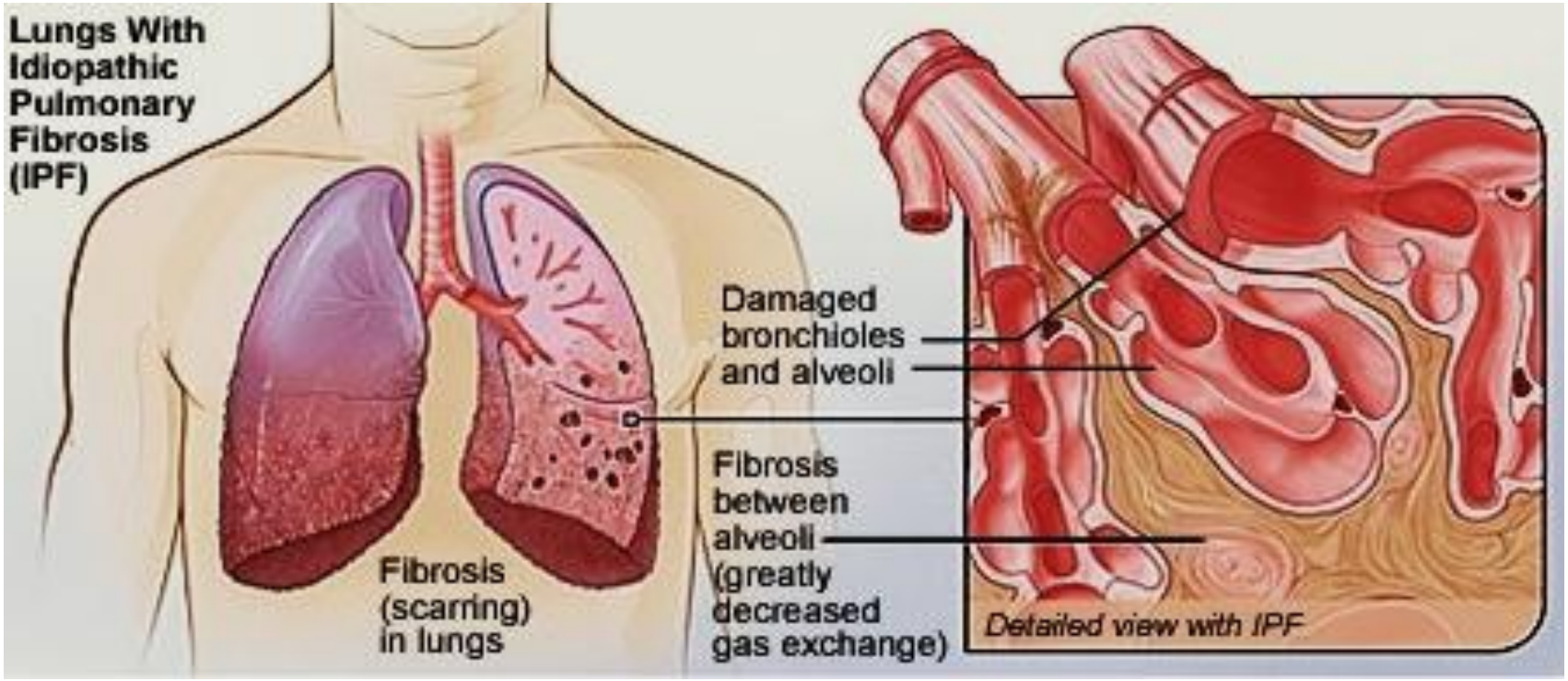
Radiation-induced

idiopathic interstitial pneumonias

They are a subset of diffuse interstitial lung diseases of unknown etiology characterized by expansion of the interstitial compartment (ie, that portion of the lung parenchyma sandwiched between the epithelial and endothelial basement membranes) with an infiltrate of inflammatory cells. The inflammatory infiltrate is sometimes accompanied by fibrosis,

Also known as CFA(Chronic fibrosing alleveolitis.

- Progressive dyspnea
- Restrictive pulmonary physiology
- Radiographically diffuse lung disease

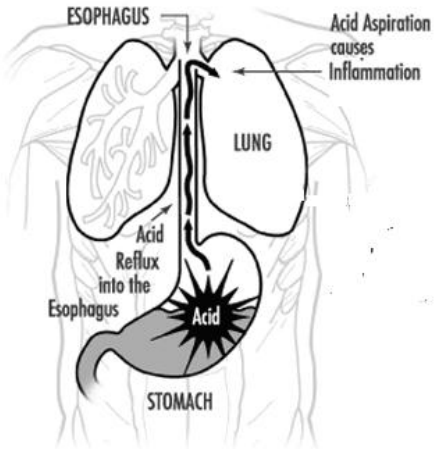


Risk factors

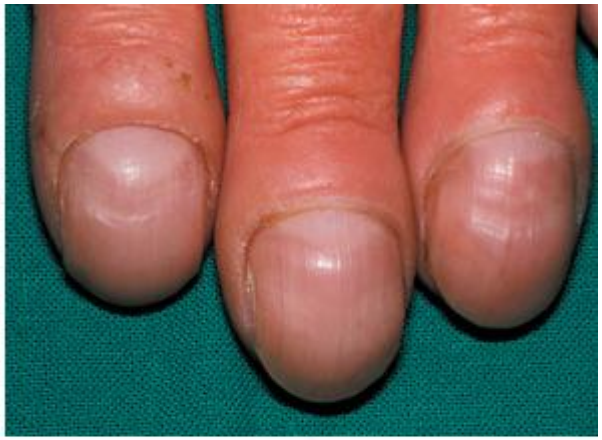
Familial IPF- rare.

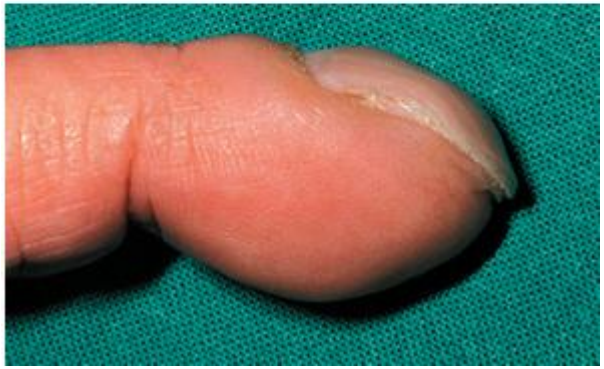
Environmental factors:

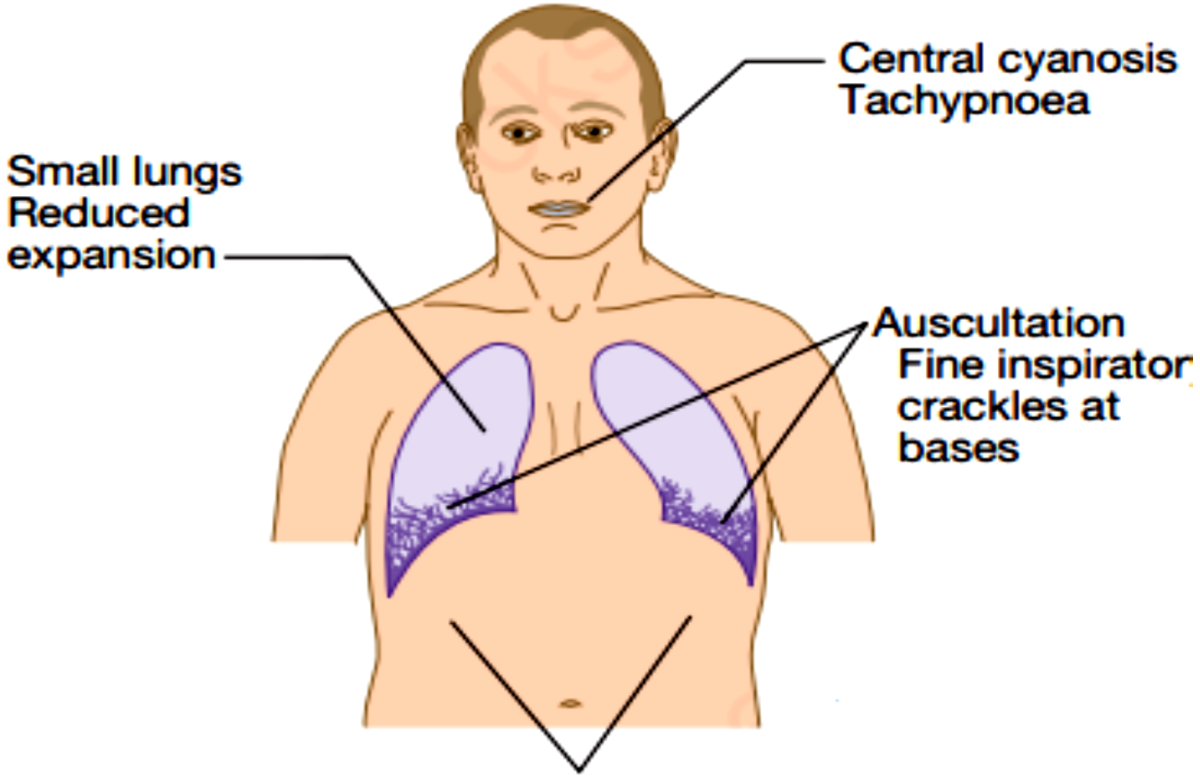
1. • Smoking
2. • Inhalation of organic and inorganic dust
3. • Gastroesophageal reflux



Pulmonary fibrosis







Also: finger clubbing common in idiopathic pulmonary fibrosis; raised JVP and peripheral oedema if cor pulmonale

Natural history of IPF

progressive impairment of lung compliance and gas exchange

Complications:

death because of

- hypoxic respiratory failure type one
- pulmonary hypertension & cor pulmonale
- Increased risk of bronchogenic carcinoma

assessment

Assessment of dyspnea as per history and clinical exam to rule out common ubderlying causes

Pulmonary function Lung volumes

Dlco

Resting ABG

Echocardiography.

Ct scan and biopsy

Treatment

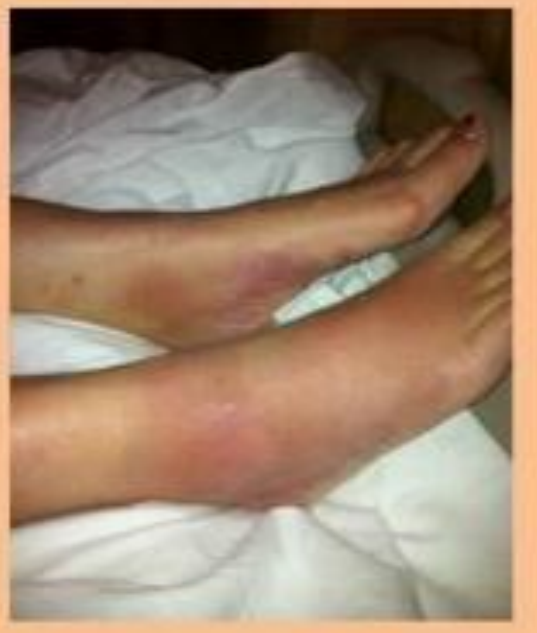
- Corticosteroids + immunosuppressives
- Low dose prednisone+ azathioprine or cyclophosphamide
- Pirfenidone Antifibrotic agent
- Immuno modulators like
- Interferon γ - inhibitory effect on fibroblasts
- Colchicine
- Lung transplantation.

sarcoidosis

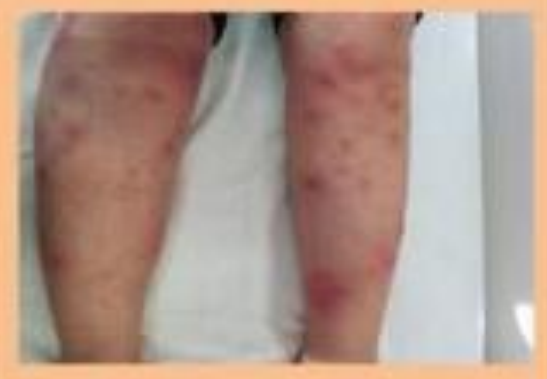
- multisystem, granulomatous, inflammatory condition of unknown cause that occurs in young adults of both sexes.
- can be asymptomatic.
- Or acute systemic presentations with fever, erythema nodosum, polyarthralgia, and hilar lymphadenopathy (Löfgren syndrome).
- An estimated 90% of patients have pulmonary involvement at the time of presentation.
- Others may involves the eyes and skin; the central nervous system, heart, and gastrointestinal tract are less commonly involved.

(Löfgren syndrome)

Hilar lymphadenopathy

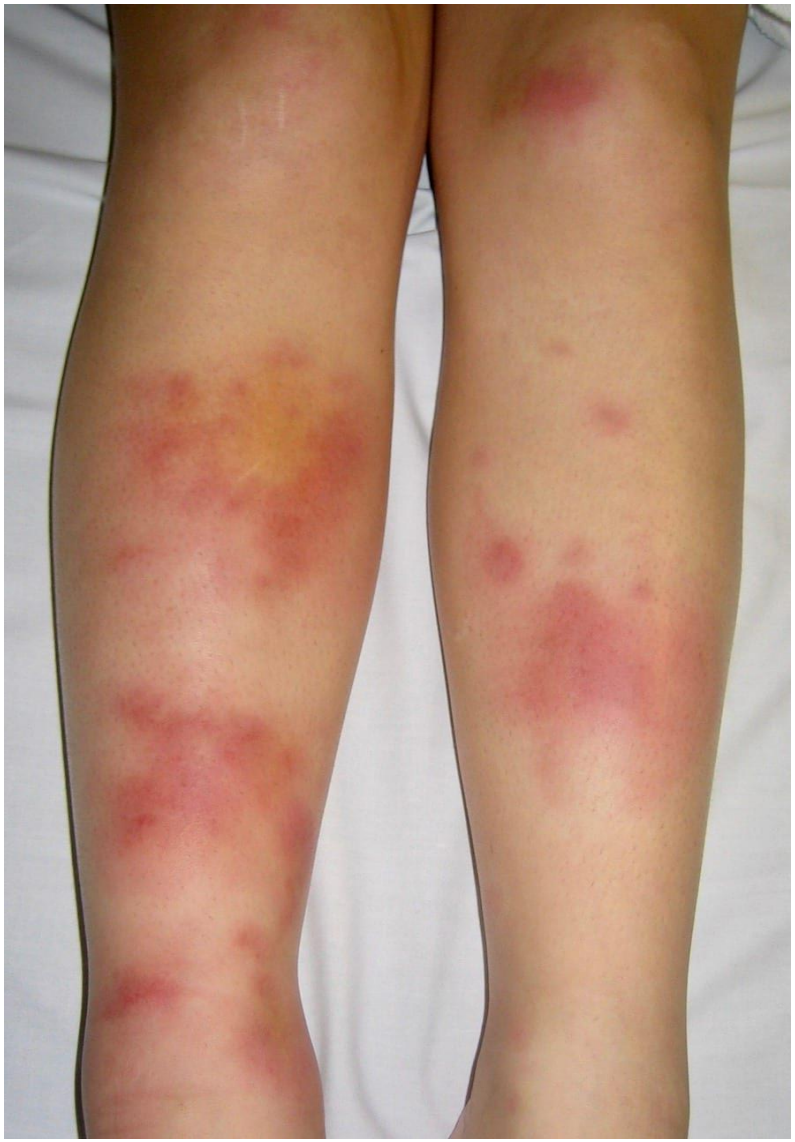


Acute polyarthritis (usually ankles)



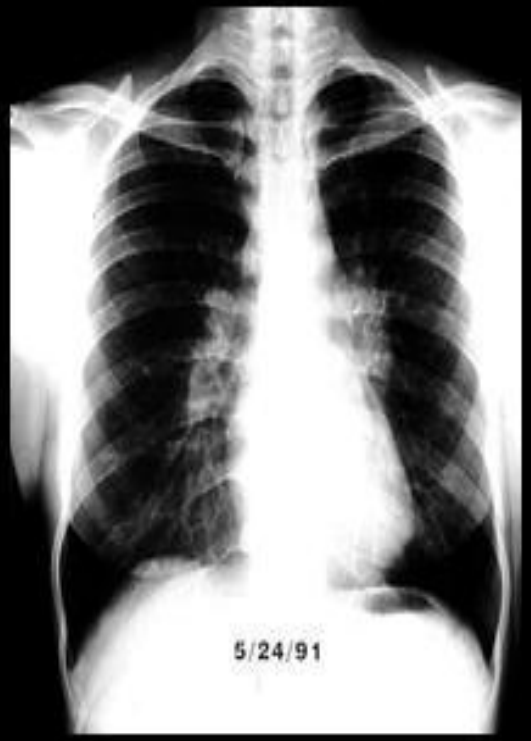
Erythema nodosum

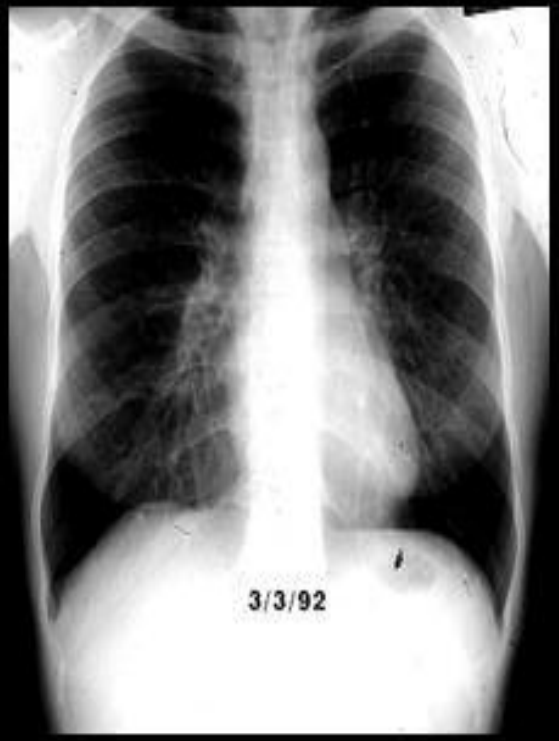
- Erythema nodosum

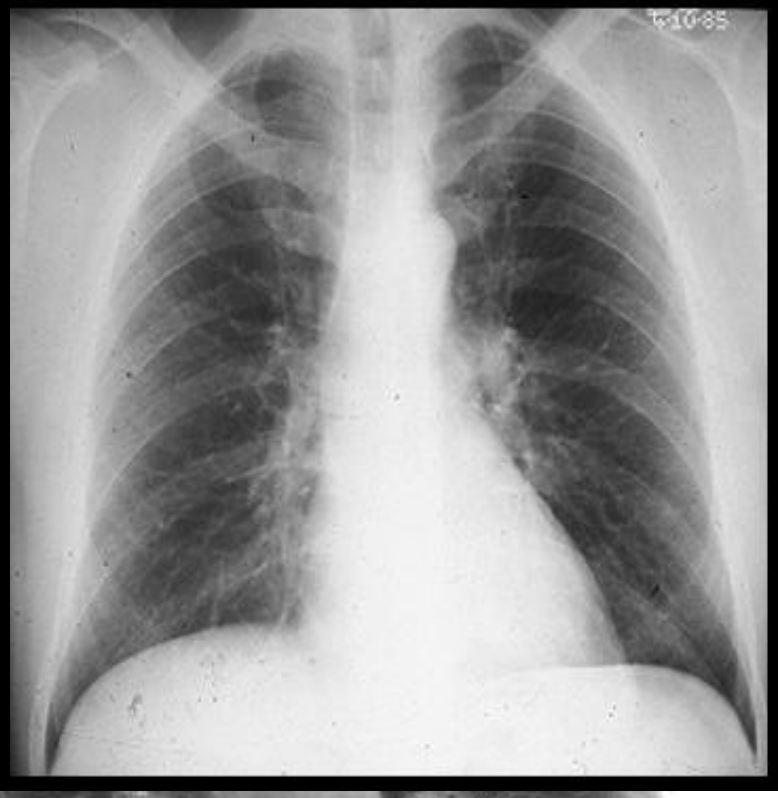


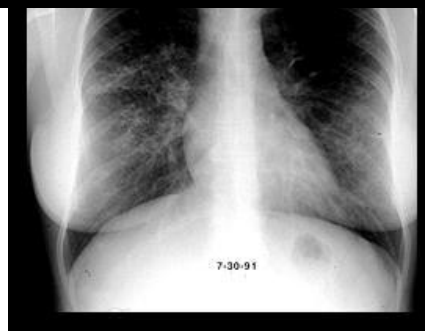
Staging by x ray

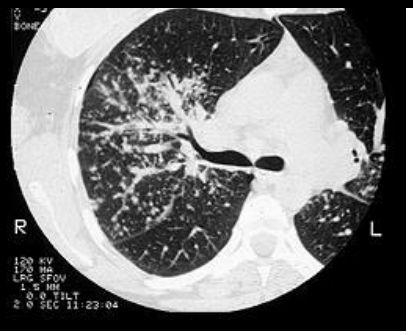
Stage	Radiographic Pattern
0	Normal
I	Hilar lymphadenopathy
II	Hilar lymphadenopathy; abnormal lung parenchyma
III	No lymphadenopathy; abnormal lung parenchyma
IV	Parenchymal fibrotic change with architectural distortion

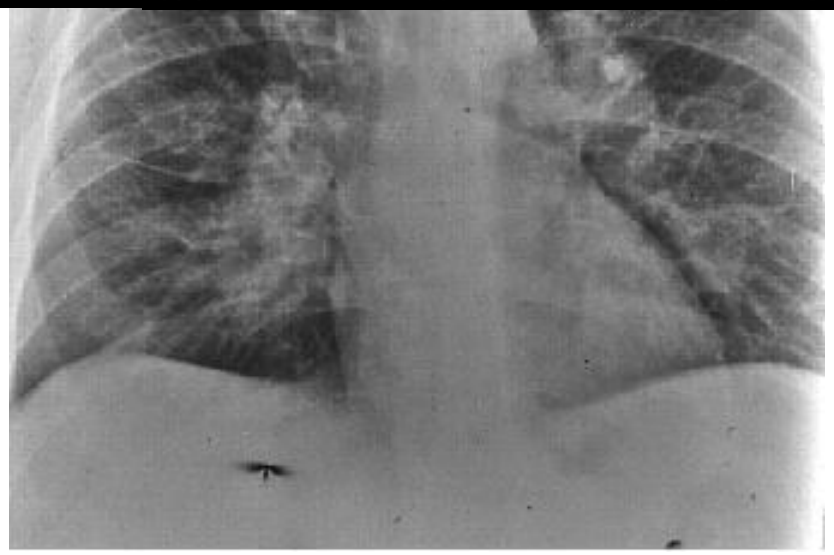

5/24/91


3/3/92



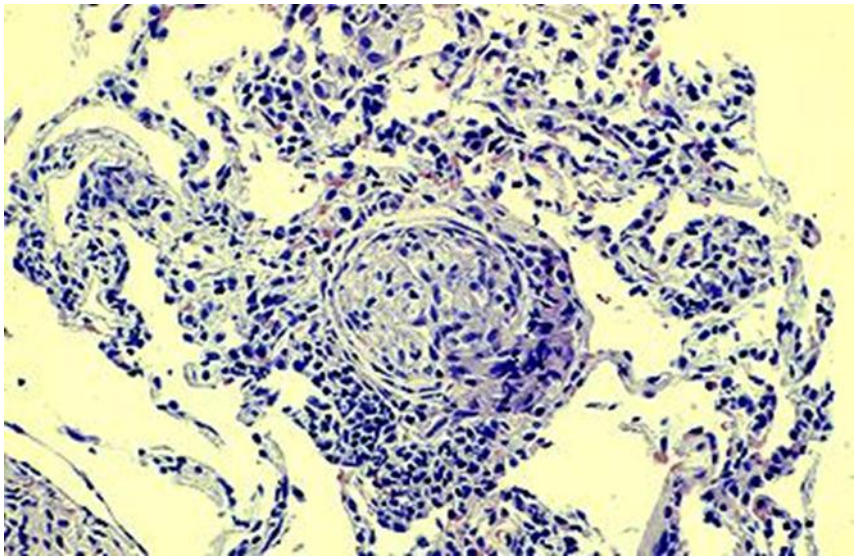

7-30-91


R
L



- The diagnosis of sarcoidosis requires biopsy of organ involvement showing noncaseating granulomatous inflammation with the appropriate clinical and radiographic findings and the exclusion of other diseases.
- .

Non caseating granuloma



- Sarcoidosis resolves spontaneously in up to two thirds of patients. Treatment not e indicated in patients with asymptomatic radiographic abnormalities
- When treatment is indicated, Prednisone, 0.5 mg/kg for 1 to 3 months and gradual tapering with alternate-day dosing of 10 to 20 mg to complete 9 to 12 months of therapy
- Patients who do not tolerate corticosteroids or who have resistant disease may be treated with methotrexate, hydroxychloroquine, or azathioprine.

Hypersensitivity Pneumonitis

allergic, inflammatory lung disease also called extrinsic allergic alveolitis.

It results from exposure to airborne allergens, which cause a cell-mediated immunologic sensitization.

, may produce parenchymal lung fibrosis.

Typical antigens that causes an allergic response are derived from bacterial and fungal elements in decaying organic matter, including moldy hay, grain dust, lumber, and stagnant water.

Antigen Source	Associated Disease
• Moldy hay, or grain	• Farmer's lung
• Bird feathers and droppings	• Bird fancier's lung
•	

Cl /features

- Dyspnea and dry cough .
- fever, weight loss, fatigue, and body aches.
- **Physical examination**
- lung crackles
- **Treatment**
- **eliminating exposure to the offending antigen ..**
- **Oral corticosteroids +_ cytotoxic agents**
- **mayr lung transplantation .**

Drug-induced and Radiation-induced Interstitial Lung Disease

- ✓Pt c/o fatigue, low-grade fever, and cough. Physical examination may reveal crackles
- ✓Radiographic findings include a pattern of reticular lines with scattered ground-glass opacification progressing to consolidation.
- ✓may resolve quickly after the offending agent is removed.
- ✓ Corticosteroids may be beneficial in selected patients with severe disease.

Drug-Induced Lung Diseases: Hypersensitivity vs. Fibrotic Interstitial

Acute/Cellular Infiltrate +/- Eosinophilia

Amiodarone

Bleomycin

Busulfan

Chronic/Interstitial Infiltrate

Amphotericin B

Bleomycin

Cyclophosphamide

Amiodarone

Methotrexate

Aproach

CLINICAL PRESENTATION OF ILD

- Exertional dyspnea.
- nonproductive cough.
- an abnormal chest radiograph.
- May symptoms associated with another disease, such as a connective tissue disease.
- With lung function abnormalities on simple office spirometry, particularly a restrictive ventilatory pattern.

Evaluation of patient with ILD

- History
- Clincal examination
- Basic initial investigations; CBC,RFT
- CXR
- ABG
- Pulmonary function tests- spirometry, lung volumes and diffusion capacity
- HRCT –chest
- Bronchoscopy and BAL
- Lung biopsy

SYMPTOMS

- Dyspnea — A sense of shortness of breath
- Grading the level of dyspnea is useful as a method to gauge the severity of the disease and to follow its course
- Cough
- Hemoptysis
- Wheezing — Wheezing is an uncommon Chest pain — Chest pain is also uncommon in most ILDs. However, pleuritic chest pain may occur in ILD associated with connective tissue disease

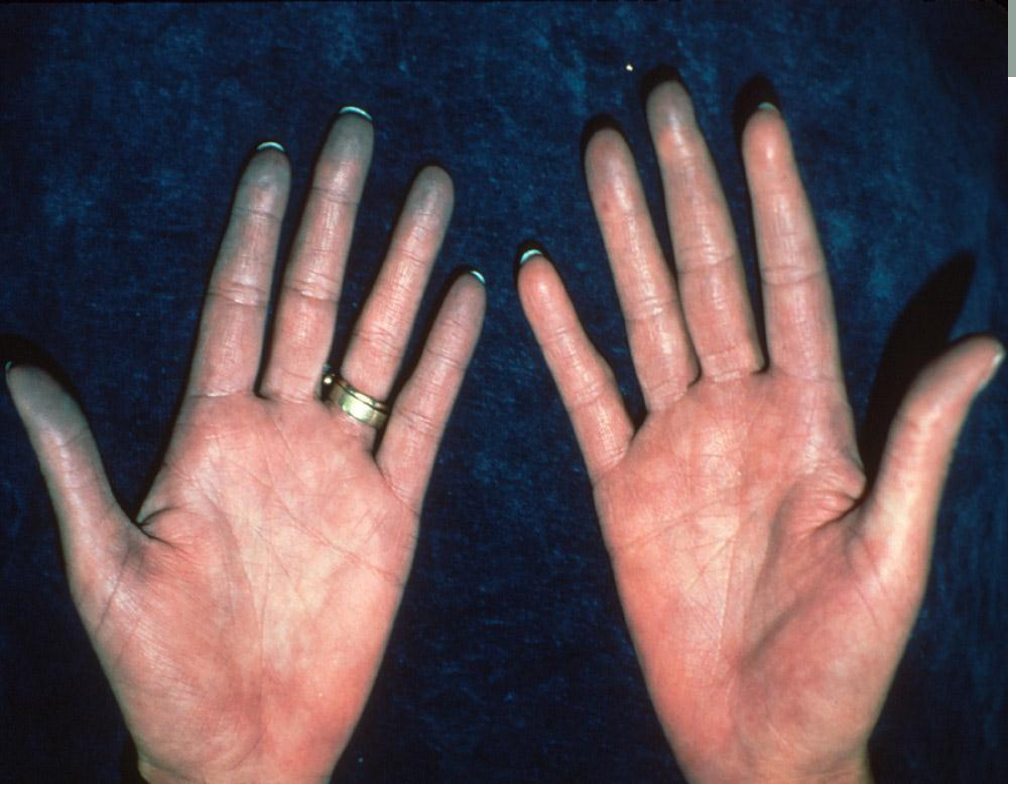
Other symptoms

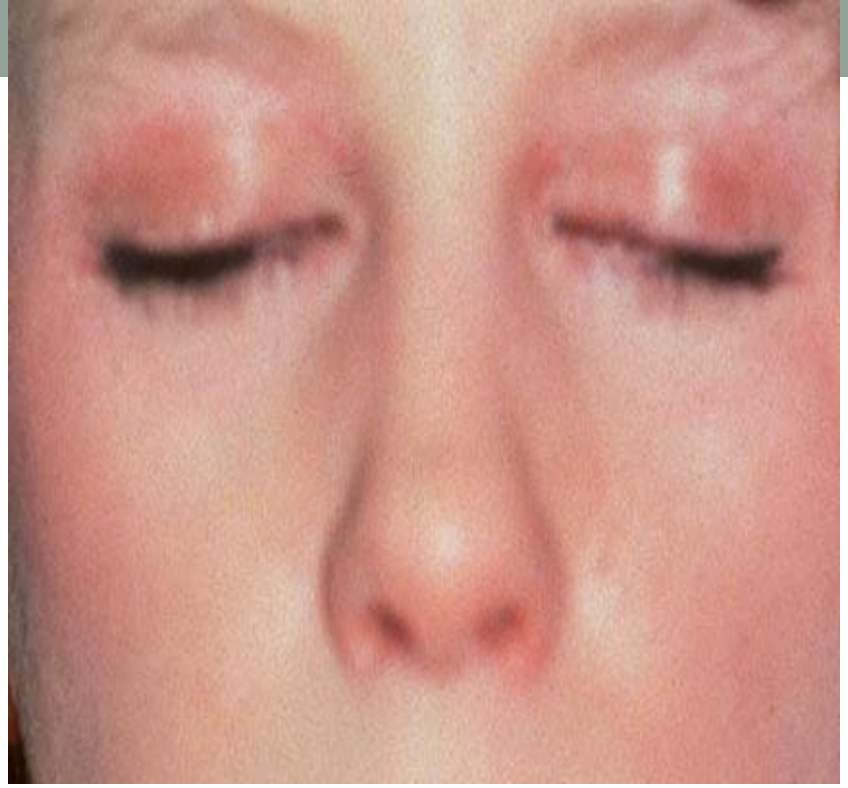
- — Clinical findings consistent with a connective tissue disease should be carefully recorded. These include musculoskeletal pain, weakness, fatigue, fever, joint pains or swelling, photosensitivity, Raynaud phenomenon, pleuritis, dry eyes, and dry mouth.

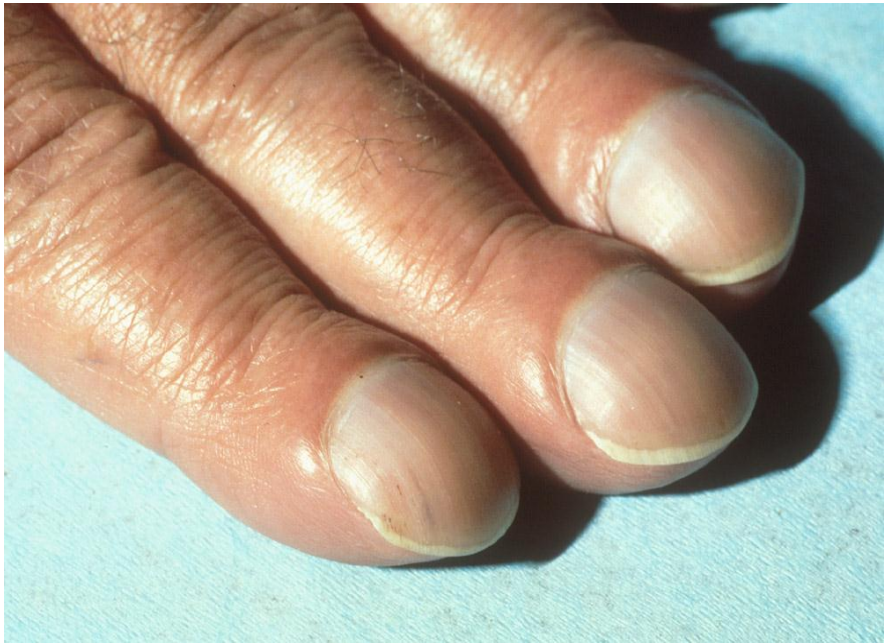
PHYSICAL EXAMINATION —

The physical examination in patients with ILD is frequently abnormal, but the findings are nonspecific.

- Crackles — common in most forms of ILD.
- Cor pulmonale — augmented P2, lower limb oedema
- clubbing is a late manifestation suggesting advanced fibrosis of the lung.
- Cyanosis — it is usually a late manifestation indicative of advanced disease.







Blood tests

Leukopenia connective tissue diseaseand drug-induced
Leukocytosis Hypersensitivity pneumonitis;
Eosinophilia Eosinophilic pneumonia; drug-induced
Thrombocytopenia connective tissue disease
Hemolytic anemia Connective tissue disease
Normocytic anemia; connective tissue disease

Hypercalcemia Sarcoidosis
Urinary sediment abnormalities Connective tissue disease; systemic vascultiis; drug-induced
Serum angiotensin-converting enzyme Sarcoidosis
Antibasement membrane antibody Goodpasture's syndrome
Antineutrophil cytoplasmic antibody Wegener's granulomatosis,
Serum precipitating antibodies Hypersensitivity pneumonitis

Serologic studies

- if clinically indicated by features suggestive of a connective tissue disease (antinuclear antibodies, rheumatoid factor, anti-synthetase antibodies), environmental exposure (hypersensitivity precipitin panel), or systemic vasculitis (antineutrophil cytoplasmic antibodies [ANCA], anti-glomerular basement membrane antibody)

Arterial blood gas

- The resting arterial blood gases may be normal or may reveal hypoxemia
-

radiography

- The most common radiographic abnormality on routine chest radiograph is a reticular pattern and may be nodular or mixed patterns.
- HRCT appears to provide greater diagnostic accuracy than plain films

Imprtance of site of involvment in xray

Lower zone predominance

Idiopathic pulmonary fibrosis

Rheumatoid arthritis (associated with usual interstitial pneumonia)

Asbestosis

Upper zone predominance

Sarcoidosis

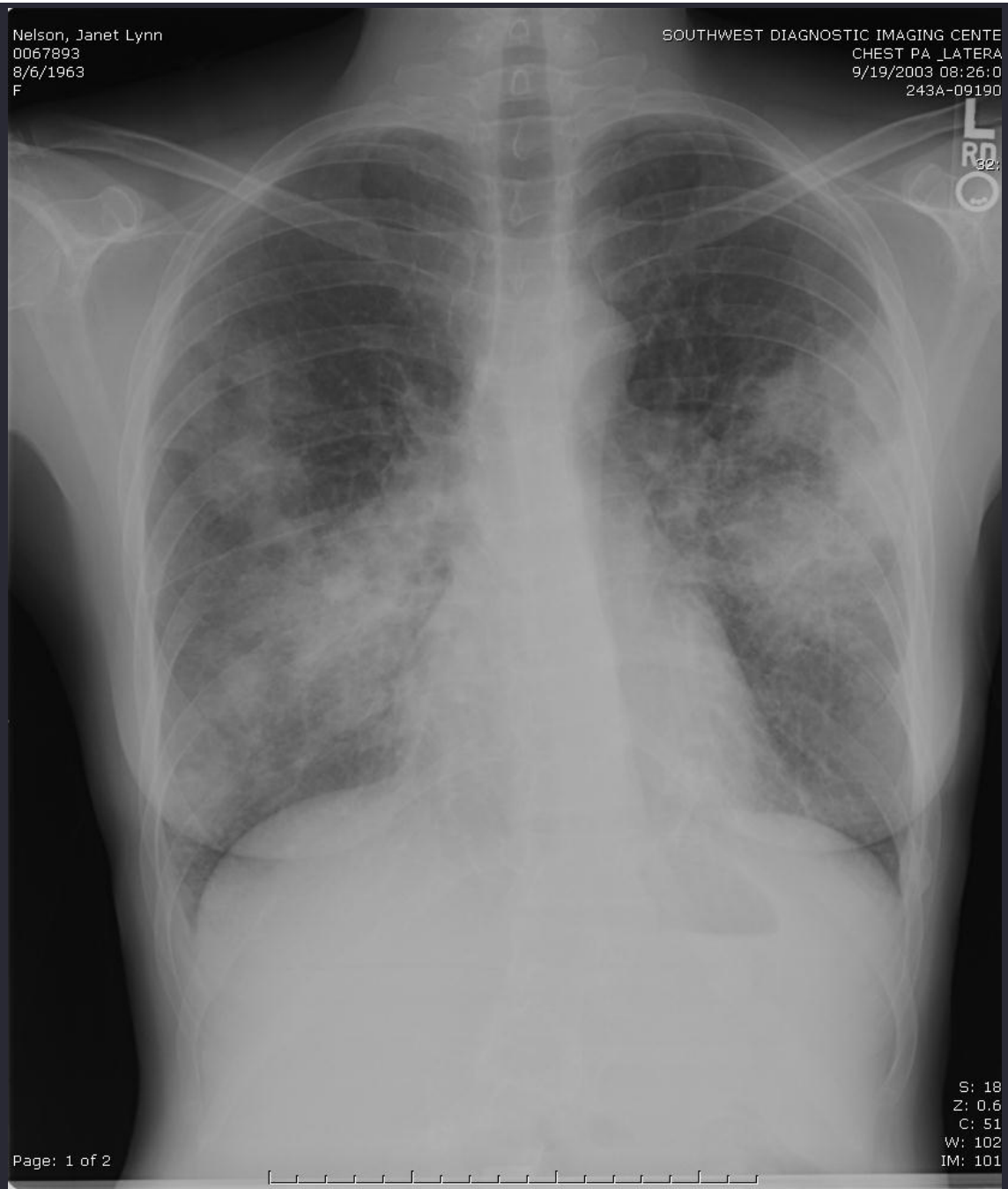
Chronic hypersensitivity pneumonitis

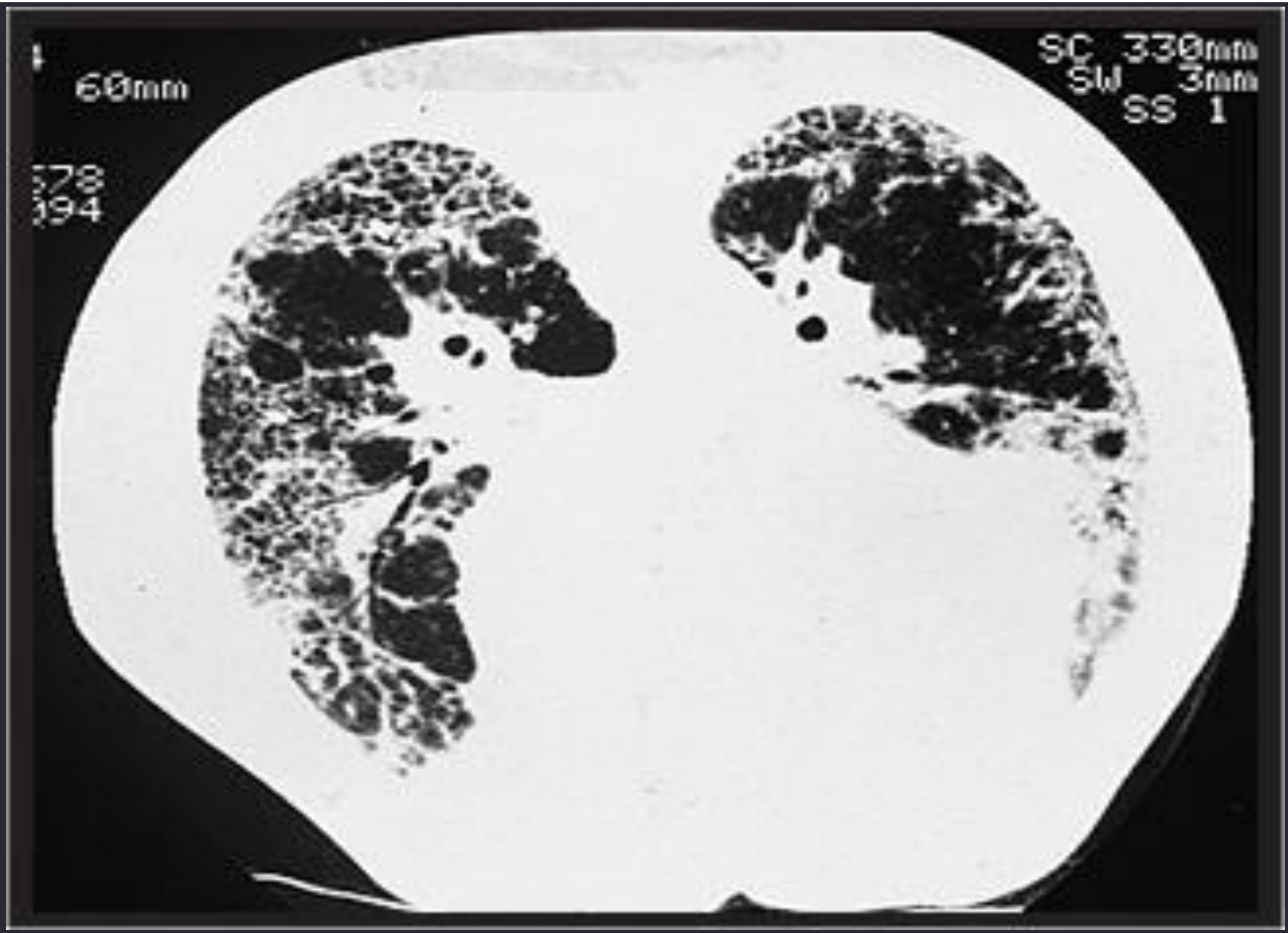
Chronic infectious diseases (eg, tuberculosis, histoplasmosis)

Ankylosing spondylitis

Silicosis

Radiation induced fibrosis


Nelson, Janet Lynn
0067893
8/6/1963
F
SOUTHWEST DIAGNOSTIC IMAGING CENTE
CHEST PA _LATERA
9/19/2003 08:26:0
243A-09190
L
RD
32,
S: 18
Z: 0.6
C: 51
W: 102
IM: 101
Page: 1 of 2

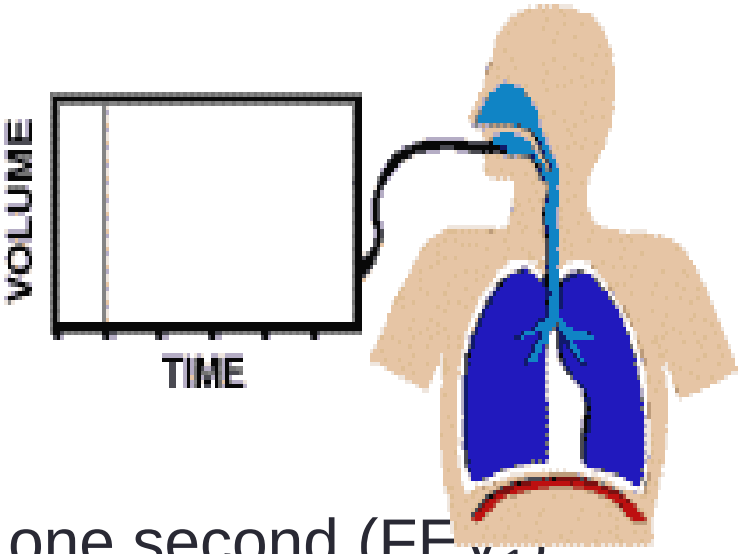

60mm
SC 330mm
SW 3mm
SS 1

Pulmonary function test PFT:

- Complete lung function testing (spirometry, lung volumes, diffusing capacity)
- Most of the interstitial disorders have a restrictive defect with reductions in total lung capacity (TLC), functional residual capacity (FRC), and residual volume (RV) .
- the FEV1/FVC ratio is usually normal or increased

Spirometry

- Simple, office-based
- Measures flow, volumes
- Volume vs. Time
- Can determine:
  - Forced expiratory volume in one second (FEV_1)
  - Forced vital capacity (FVC)
  - FEV_1/FVC
  - Forced expiratory flow 25%-75% (FEF_{25-75})



Restrictive Pattern

- Decreased FEV_1
- Decreased FVC
- FEV_1/FVC *normal* or *increased*

Bronchalveolar lavage

- BAL cellular analysis studies may be useful to narrow the differential diagnostic possibilities between various types of ILD.
- BAL is a minor extension of routine fiberoptic bronchoscopy and may help define the stage of disease and allow for the assessment of disease progression or response to therapy.

ROLE OF LUNG BIOPSY

- To provide a specific diagnosis.
- To assess disease activity.
- To exclude neoplastic and infectious processes

OCCUPATIONAL LUNG DISEASES

Occupational lung dis

- Exposure to dusts, gases, vapours and fumes at work can cause several different types of lung disease:
- Acute bronchitis and even pulmonary oedema from irritants such as sulphur dioxide, chlorine, ammonia or the oxides of nitrogen
- Pulmonary fibrosis due to mineral dust
- Occupational asthma (the commonest industrial lung disease).
- Hypersensitivity pneumonitis .
- Bronchial carcinoma due to industrial agents (e.g. polycyclic hydrocarbons.

- environmental/occupational history is vital . Systematically collecting information about a patient’s past employment, hobbies, and household exposures is practical. Taking a focused exposure history aimed at identifying occupational or environmental risk factors .h/o suggestiv of clustering.

When to Suspect Occupational Lung Disease

1. Patient concern.(c/0)
2. Patient report of a temporal pattern of signs and symptoms, with improvement over the weekend or during vacation.
3. Patient report that several coworkers are affected with a similar illness.
4. Patient report of known hazardous substances at work.
5. Lack of a therapeutic response to aggressive appropriate treatment.

Asbestos-associated Lung Disease

- Asbestos fibers are naturally occurring fibrous, hydrated silicate (cause asbestos-related lung diseases
- Pleural disease
- Pleural plaques (localized, often partially calcified)
- Benign pleural effusion
- Mesothelioma
- Parenchymal lung disease
- Asbestosis Lung cancer

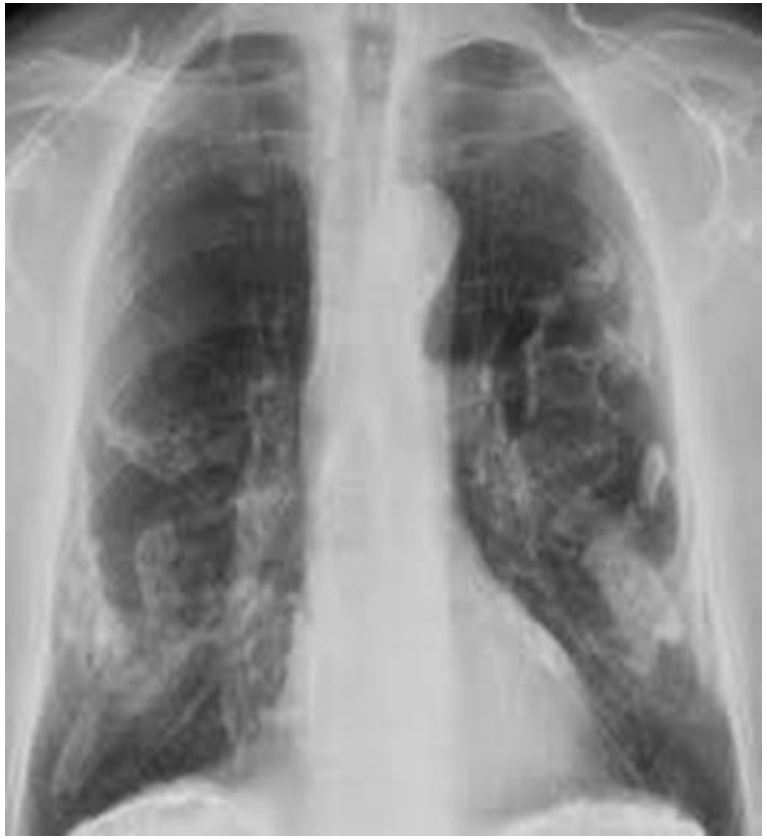

© picture alliance / Pressefoto Ulmer

Risk Factors

- risk increase withcumulative exposure to the asbestos fiber.
- The largest number of exposed workers was in the construction industry, and the shipbuilding.

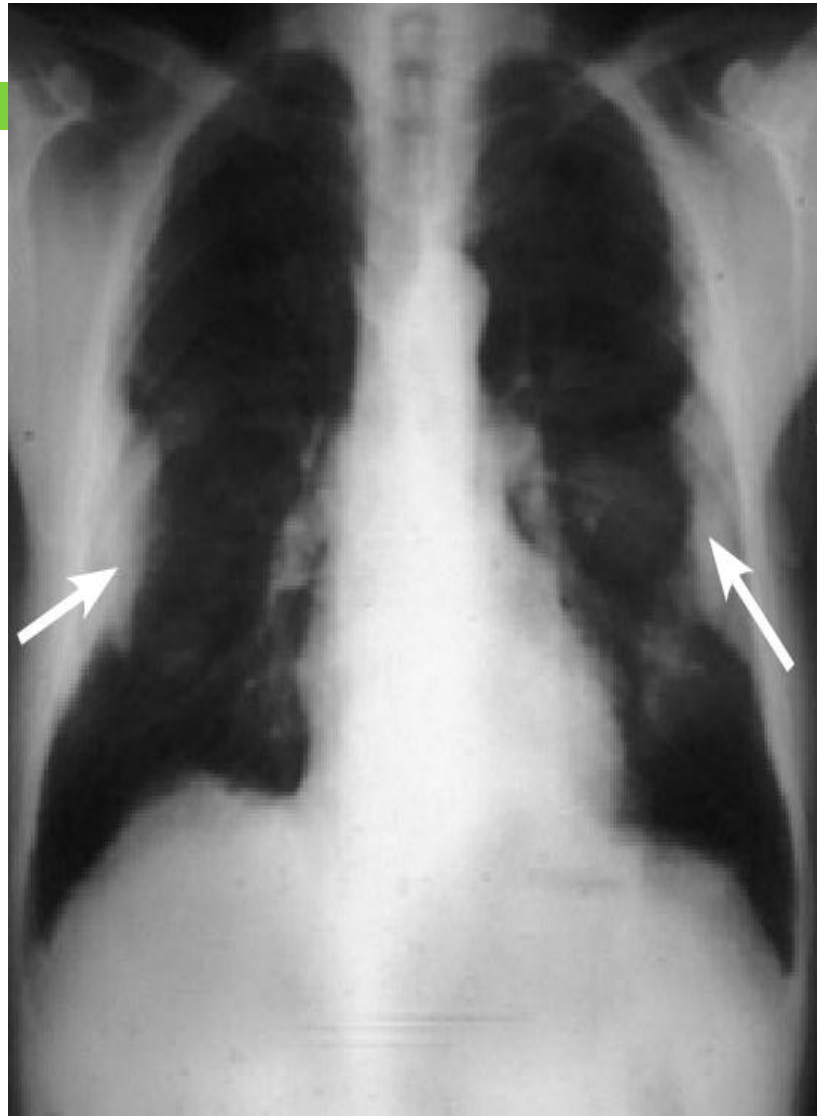
Asbestosis

- bilateral interstitial fibrosis of the lung parenchyma caused by inhalation of asbestos fibers
- Pt c/o breathlessness,
- o/e clubbing and bibasilar inspiratory crackles, and pulmonary function testing showing a restrictive pattern
- and decreased DLCO.



Pleural Plaques

- Pleural plaques are typically develop bilaterally, with a latency of more than 10 years. Patient should be monitored for the development of additional asbestos-related intrathoracic disease.



treatment

- There is no effective pharmacologic therapy for asbestosis.
- Steroids are of no values in treatment.
- Management includes supplemental oxygen if necessary ,influenza and pneumococcal vaccinations, smoking cessation if applicable, and aggressive therapy for intercurrent infections.

Asbestos-related Lung Cancer

- Cigarette smoke and asbestos have a synergistic (multiplicative) effect on the risk for lung cancer. but this is not true for mesothelioma.

Chronic Obstructive Pulmonary Disease

- Smoking is the most common risk factor for COPD.
- The most common occupational and environmental causative agents for COPD are
- coal, silica, and cadmium exposure.

Diagnosis and Management

- Dx and pharrmacologic management is similar to that of other forms of COPD, except that removal from exposure must also be considered.

- 38 year old male presents with 11 month history of rhinitis, followed by cough, shortness of breath and chest tightness.
- No environmental (seasonal) medication or aspirin allergy
- Ex-smoker: 5 pack years.
- Occupation: automobile painter for 3 years
- Otherwise well
- Symptoms: worse at end of the workday – improves on weekends and vacations
- Physical exam: un-remarkable. Cardiac and lung sounds were normal. There were no inspiratory or expiratory wheezes, nor a prolonged expiratory phase.

occupational asthma

- Occupational asthma (OA) is a disease characterized by variable airflow obstruction, airway and airway inflammation attributed to a particular occupational environment and not due to stimuli encountered outside the workplace
- Approximately 10 percent of asthmatic subjects identify workplace exposure factors.

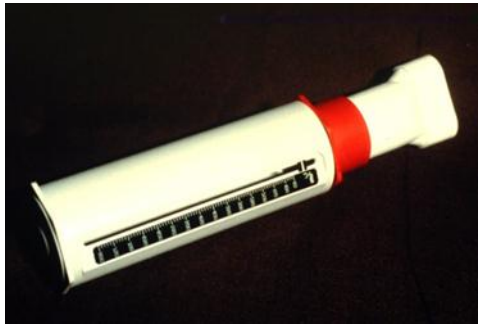
Risk factors

- Isocyanates (eg, toluene diisocyanate,) insulators, painters
- Metals (eg, chromic acid, vanadium, platinum salts)-> welders and chemical workers
- Animal proteins (eg, domestic and laboratory animals, fish and seafood)→ Farmers, veterinarians, poultry , fish and seafood processors.

Major causes of occupational asthma

	Occupation at risk
Low molecular weight chemicals	
Isocyanates (eg, toluene diisocyanate, diphenylmethane diisocyanate, hexamethylene diisocyanate, naphthalene diisocyanate)	Polyurethane workers, roofers, insulators, painters
Anhydrides (eg, trimellitic anhydride, phthalic anhydride)	Manufacturers of paint, plastics, epoxy resins
Metals (eg, chromic acid, potassium dichromate, nickel sulfate, vanadium, platinum salts)	Platers, welders, metal and chemical workers
Drugs (eg, beta lactam agents, opiates, other)	Pharmaceutical workers, farm workers, health professionals
Wood dust (eg, Western red cedar, maple, oak, exotic woods)	Carpenters, woodworkers
Dyes and bleaches (eg, anthraquinone, carmine, henna extract, persulfate, reactive dyes)	Fabric and fur dyers, hairdressers
Amines	Chemists, cleaners, plastic manufacturers
Glues and resins (eg, acrylates, epoxy)	Plastic manufacturers
Miscellaneous (eg, formaldehyde, glutaraldehyde, ethylene oxide, pyrethrin, polyvinyl chloride vapor)	Laboratory workers, textile workers, paint sprayers, health professionals
High molecular weight organic materials	
Animal proteins (eg, domestic and laboratory animals, fish and seafood)	Farmers, veterinarians, poultry processors, fish and seafood processors
Flours and cereals	Bakers, food processors, dock workers
Enzymes (eg, pancreatic extracts, papain, trypsin, Bacillus subtilis, bromelain, pectinase, amylase, lipase)	Bakers, food processors, pharmaceutical workers, plastic workers, detergent manufacturers
Plant proteins (eg, wheat, grain dust, coffee beans, tobacco dust, cotton, tea, latex, psyllium, various flours)	Bakers, farmers, food and plant processors, health professionals, textile workers

- Pre and post shift monitoring of lung function
- Spirometry
- PEF at and off work for period of several weeks - 4 times daily, preferable every 2 hours



rx

- Primary treatment: removal of exposure
- If not removed?
- Can exposure be controlled to control disease
- Personal protective measures
- Keep in position with medication control
- Desensitization?

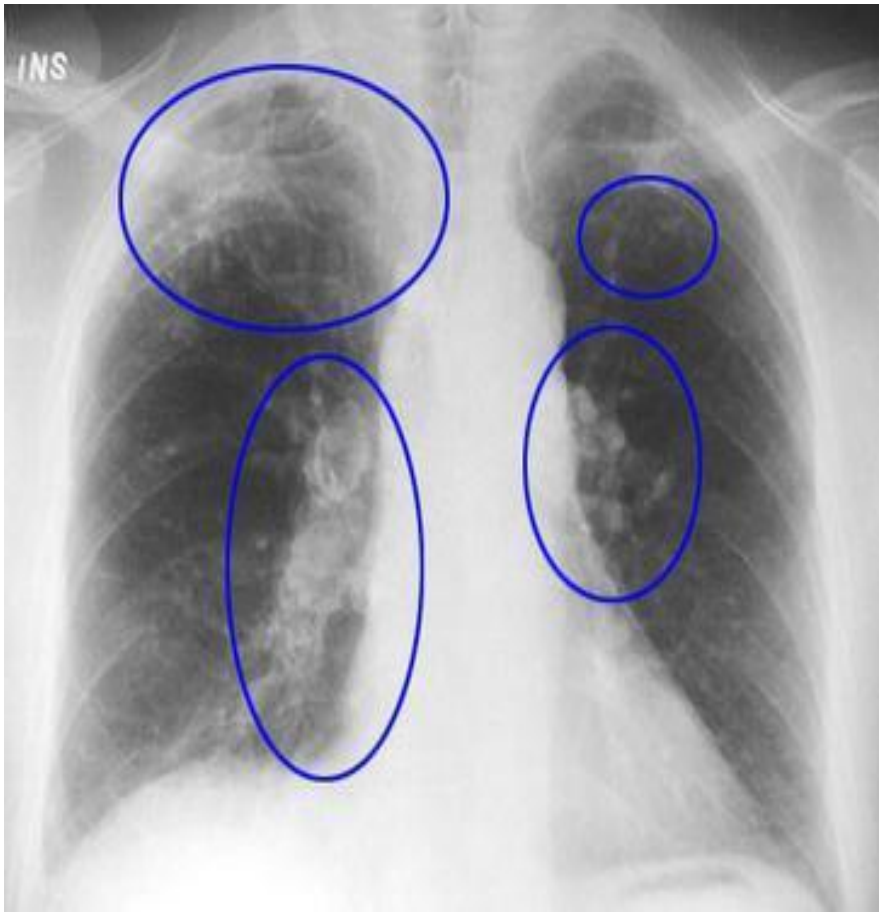
Silicosis

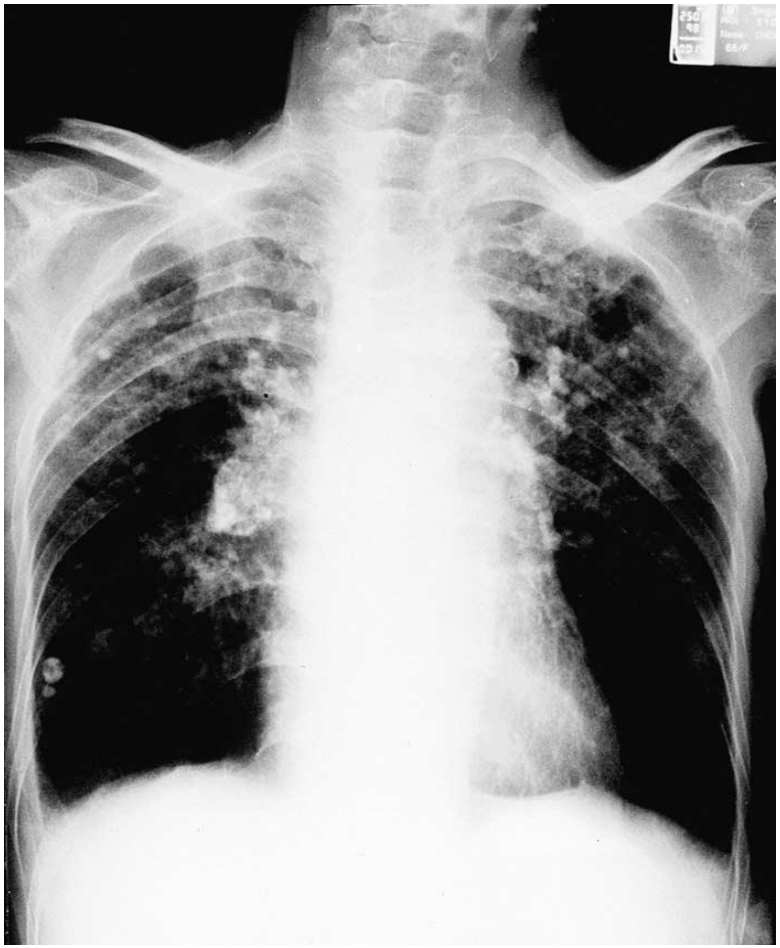
Silicosis refers to pulmonary diseases caused by inhalation of free crystalline silica (silicon dioxide)

<u>Occupations associated with silicosis</u>

1. coal mining
2. Hard rock mining
3. Tunneling
4. Quarrying and stone cutting
5. Steelworks
6. Sand blasting
7. Construction

egg-shell calcification plus the upper lobe nodules are typical of silicosis





- Chronic silicosis — simple slicosis slowly, 10 to 30 years after first exposure. to become radiographically apparent
- May many years after cessation of employment in a job associated with exposure .
- Acute silicosis is characterized by rapid onset of cough, weight loss, fatigue, and pleuritic pain.
- On physical examination, crackles are usually present.
- The prognosis is very poor. Patients rapidly develop cyanosis, cor pulmonale, and respiratory failure.

- In a minority of those with chronic disease, nodules coalesce resulting in progressive massive fibrosis (PMF).
- (PMF) is associated with more severe symptoms than simple silicosis. Physical examination frequently demonstrates decreased or other abnormal breath sounds. Signs of chronic respiratory failure and cor pulmonale..
- Patients with silicosis have increased risk of TB

  And lung cancer.

treatment

- Non specfic symptomatic therapy should include treatment of airflow limitation with bronchodilators.
- Rx of respiratory tract infection and use of supplemental oxygen (if indicated) to prevent complications of chronic hypoxemia.

Chronic beryllium disease (berylliosis)

- occur in metal , ceramics, and nuclear weapons manufacturing .
- A patient is considered to have CBD if he or she has all of the following:
  - A history of any beryllium exposure
  - A positive blood or bronchoalveolar lavage (BAL) beryllium lymphocyte proliferation test.
  - Non caseating pulmonary granulomas on lung biopsy

Cl/ f

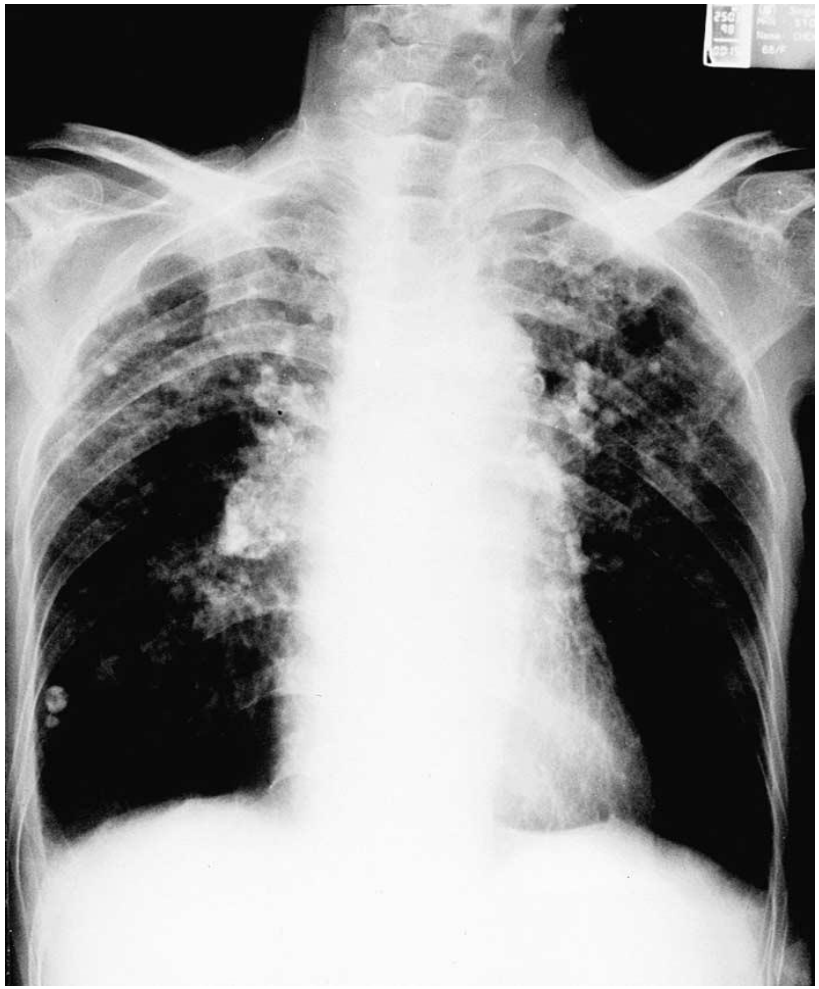
- dry cough, shortness of breath, night sweats, fatigue, and weight loss. .
- Pulmonary examination typically reveals bibasilar crepitations.
- advanced disease lead to cor pulmonale and digital clubbing.

- Chest radiographs may be normal or show hilar adenopathy with reticulonodular opacification
- computed tomography is more sensitive in identifying the presence of parenchymal nodules, ground glass opacities and hilar or mediastinal adenopathy
- Rx Steroids and Oxygen

Coal workers pneumoconiosis

- Prolonged inhalation of coal dust
- Simple pneumoconiosis- does not progress if miner leaves the industry.
- Progressive massive fibrosis.
- Caplan syndrome- massive fibrotic nodules in patients with RA.

This 80-year-old male used to work in a sand quarry. He was asymptomatic. What is the diagnosis?



silicosis

- bilateral infiltrates and egg-shell calcification of the hilar lymph nodes. The egg-shell calcification plus the upper lobe nodules are typical of silicosis.
- Differential diagnoses of upper lobe infiltrates include tuberculosis, ankylosing spondylitis and silicosis, .
- Differential diagnoses of egg-shell calcification include
- sarcoidosis, lymphoma following radiotherapy, and coal-worker's pneumoconiosis.

Thank u