A 55 year old man comes for evaluation of exercise intolerance over the past 6 months. He has no significant past medical history. He informs you that over the past week he cannot walk across the room without getting “short of breath.” He takes no medications and has never smoked. The physical exam is significant for a respiratory rate of 24/minute, jugular venous distension 8 cm, coarse crackles on auscultation, clubbing, and trace pedal edema on the legs. The chest x-ray reveals diffuse reticular disease.
What is the diagnosis?

- Idiopathic Pulmonary Fibrosis.
INTERSTITIAL & INFILTRATIVE PULMONARY DISEASES

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Holy Family Hospital
Rawalpindi Medical College
Definition.
Classification.
Features common to all DPLDs (clinical manifestations, physical findings, workup).
Idiopathic interstitial pneumonias.
Idiopathic Pulmonary Fibrosis.
Sarcoidosis
Occupational Lung Diseases/ Pneumoconiosis
A heterogeneous group of conditions affecting the pulmonary parenchyma (interstitium) and/or alveolar lumen, which are frequently considered collectively as they share a sufficient number of clinical, physiological and radiographic similarities.
Fig. 19.55 Classification of diffuse parenchymal lung disease.
FEATURES COMMON TO THE DIFFUSE PARENCHYMAL LUNG DISEASES

CLINICAL PRESENTATION:
- **Cough**: usually dry, persistent and distressing
- **Breathlessness**: usually slowly progressive; insidious onset; acute in some cases
EXAMINATION FINDINGS:

- **Crackles**: typically bilateral and basal.
- **Clubbing**: common in idiopathic pulmonary fibrosis but also seen in other types, e.g. asbestosis.
- **Central cyanosis and signs of right heart failure** in advanced disease.
Laboratory investigations

- **Full blood count**: lymphopenia in sarcoid; eosinophilia in pulmonary eosinophilias and drug reactions; neutrophilia in hypersensitivity pneumonitis.
- **Ca2+**: may be elevated in sarcoid.
- **Lactate dehydrogenase**: may be elevated in active alveolitis.
- **Serum angiotensin-converting enzyme**: non-specific indicator of disease activity in sarcoid.
- **ESR and CRP**: non-specifically raised.
- **Autoimmune screen**: anti-cyclic citrullinated peptide (anti-CCP) and other autoantibodies may suggest connective tissue disease.
FEATURES COMMON TO THE DIFFUSE PARENCHYMAL LUNG DISEASES

- RADIOLGOGY:
- Chest X-ray: typically small lung volumes with reticulonodular shadowing but may be normal in early or limited disease.
HRCT: combinations of ground glass changes, reticulonodular shadowing, honeycomb cysts and traction bronchiectasis, depending on stage of disease.
FEATURES COMMON TO THE DIFFUSE PARENCHYMAL LUNG DISEASES

- **PULMONARY FUNCTION:**
- Typically *restrictive ventilatory defect* with reduced lung volumes and impaired gas transfer; exercise tests assess exercise tolerance and exercise-related fall in SaO2.
Bronchoscopy
- Bronchoalveolar lavage.
- Transbronchial biopsy.
- Bronchial biopsy.

Video-assisted thorascopic lung biopsy (in selected cases).

Others
- Liver biopsy.
- Urinary calcium excretion.
A major subgroup of DPLDs.
Unknown aetiology.
They are often distinguished by the predominant histological pattern on tissue biopsy; hence they are frequently referred to by their pathological description - for example, usual interstitial pneumonia or non-specific interstitial pneumonia.
The most important of these is idiopathic pulmonary fibrosis.
Idiopathic Pulmonary Fibrosis

- A progressive fibrosing interstitial pneumonia of unknown cause, occurring in older adults and associated with the histological or radiological pattern of usual interstitial pneumonia (UIP).
CAUSES

- Workplace
- Medications
- Genetic
- Cigarette Smoking
- Drug Use
SYMPTOMS

- Occasional Chest Pain
- Rapid Weight Loss
- Fatigue and Weakness
- Chest Discomfort
- Loss of Appetite
- Cough (usually dry)
- Shortness of Breath
- Dyspnea on Exertion
- Decreased tolerance for activity
PATIENT ASSESSMENT

- Abnormal breath sounds on auscultation - Crackles (especially in the lower lung fields)

- **Cyanosis** around the mouth or fingernails (in advanced stages)

- **Digital Clubbing**
POSSIBLE COMPLICATIONS

- Chronic Hypoxemia
- Cor Pulmonale
- Polycythemia
- Pulmonary Hypertension
- Respiratory Failure
DIAGNOSING PULMONARY FIBROSIS

A series of tests will be performed to confirm diagnosis:

- Bronchoscopy

- Tests for connective tissue diseases such as Rheumatoid Arthritis and Scleroderma

- Bronchoalveolar Lavage

- Pulmonary Function Tests
DIAGNOSING PULMONARY FIBROSIS

- Chest X-Ray
DIAGNOSING PULMONARY FIBROSIS

- Lung Biopsy
TREATMENT

- No known cure exists for Pulmonary Fibrosis

- Medications such as corticosteroids reduce swelling and inflammation

- OXYGEN

- Lung Transplant in advanced stages

- Rehabilitation and Education programs
Research on medications is currently being performed:

- **Acetylcysteine** - may prevent further damage to the lungs
- **Interferon gamma-1b** - manmade version of a substance that your body normally produces to help fight infections
- **Pirfenidone** - may reduce scarring
- **Colchicine** - may slow scarring process
- **Penicillamine** - may improve lung function
Sarcoidosis

- Multisystem chronic inflammation characterized by non caseating granulomas.

Caseating granulomas

TB

Non caseating granulomas

Sarcoidosis
Sarcoidosis

- Can involve any organ.
- 90% thoracic (lung & LN).
- Thoracic involvement accounts for most of the morbidity & mortality associated with the disease.
Epidemiology

- **Age:** 3rd decade.
- **Sex:** F > M.
- **Race:** black Americans.
Sarcoidosis

Asymptomatic 50%

Respiratory symptoms
- Cough
- Dyspnea
- expectoration

Constitutional symptoms
- Fatigue
- Weight loss
- Night sweats

Erythema nodosum
Lacrimal gland enlargement
Parotid gland enlargement
Nasal cutaneous sarcoid lesions (lupus pernio)
Cranial nerve palsy
Interstitial lung disease
Granulomatous liver disease
Phalangeal bone cysts
Skin plaques and nodules Infiltration of scars
Mononeuritis multiplex Peripheral neuropathy
Pachymeningitis Space-occupying lesion Diabetes insipidus
Anterior uveitis Sicca syndrome
Lymphadenopathy Bilateral hilar lymphadenopathy (BHL)
Cardiac arrhythmia Heart block, sudden death
Splenomegaly
Nephrocalcinosis Hypercalciuria Renal stones
Erythema nodosum
Arthropathies Osteoporosis
ERYTHEMA NODOSUM
LUPUS PERNIO
LABORATORY FINDINGS

- Kviem test +ve.
- Elevated angiotensin converting enzyme (ACE) 60-80%. ACE levels tested regularly to check the severity of the disease and to monitor the response to therapy.
LAB ABNORMALITIES

- Lymphocytopenia.
- Mild eosinophilia.
- Increased E.S.R.
- Hyperglobulinemia,
- PFTs.
“Egg shell” calcification of hilar nodes.
Plural effusions.
Cavitations.
Atelectasis.
Pneumothorax.
Cardiomegaly.
Acute sarcoidosis characterized by triad of Lofgren syndrome:
- Bilateral hilar lymphadenopathy
- Arthritis
- Erythema nodosum
Acute sarcoidosis characterized by uveoparotitis.
HEERFORDT’S SYNDROME
PULMONARY SARCOIDOSIS
PULMONARY SARCOIDOSIS

Pulmonary sarcoidosis

- Lymphadenopathy
  - Typical
  - Atypical
- Parenchymal sarcoidosis
  - Typical
  - Atypical
- Tracheobronchial sarcoidosis
- Pleural effusion
### Staging of Sarcoidosis on the Basis of Chest Radiographs

<table>
<thead>
<tr>
<th>Stage</th>
<th>Abnormality Description</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No abnormalities</td>
<td>5%–10%</td>
</tr>
<tr>
<td>1</td>
<td>Lymphadenopathy (fig. A)</td>
<td>50%</td>
</tr>
<tr>
<td>2</td>
<td>Lymphadenopathy + pulmonary infiltration (fig. B)</td>
<td>25%–30%</td>
</tr>
<tr>
<td>3</td>
<td>Pulmonary infiltration (fig. C)</td>
<td>10%–12%</td>
</tr>
<tr>
<td>4</td>
<td>Fibrosis</td>
<td>5%</td>
</tr>
<tr>
<td></td>
<td>(up to 25% during the course of the disease)</td>
<td></td>
</tr>
</tbody>
</table>
One of the distinguishing features of sarcoidosis.
- As the lung disease worsens,
- the nodal enlargement usually regresses.
Prognosis of pulmonary sarcoidosis

- 3/4 Complete resolution of hilar lymphadenopathy
- 1/3 Complete resolution of parenchymal disease
- 1/5 Irreversible pulmonary fibrosis
No known cure.

**NSAIDs.**

**Corticosteroids**, primary treatment for inflammation and granuloma formation.

Prednisolone, 1 mg/kg for 4-6 weeks followed by slow taper over 2-3 months.

Abnormal cardiac/Neuro/Ocular /Hypercalcemia/Multi system - Sterioids must.
**TREATMENT**

- **Cutaneous Sarcoidosis**: Topical steroids/Hydroxychloroquine: 200-400mg/day.
- **Methotrexate**: Start with 10mg/week and maintain with 2.5 to 15mg/week.
- **Azathioprine**: 50-150mg/day.
- **TNF-alpha inhibitors**.
- **Lung transplantation**.
Pneumoconiosis is a generic name covering the group of lung disorders which result from the inhalation of “inorganic dusts.”
Depending upon the type of dust, the disease is given different names:

- **Coalworker's pneumoconiosis** (also known as miner's lung, black lung or **anthracosis**) — coal, carbon
- **Asbestosis** — asbestos
- **Silicosis** (also known as "grinder's disease" or Potter's rot) — silica
- **Bauxite fibrosis** — bauxite
- **Berylliosis** — beryllium
- **Siderosis** — iron
- **Byssinosis** — cotton
- **Silicosiderosis** — mixed dust containing silica and iron
- **Labrador lung** (found in miners in Labrador, Canada) — mixed dust containing iron, silica and **anthophyllite**, a type of asbestos
- **Stannosis** — tin oxide
Chest X-ray may show a characteristic patchy, subpleural, bibasilar interstitial infiltrates or small cystic radiolucencies called honeycombing.

Pneumoconiosis in combination with multiple pulmonary rheumatoid nodules in rheumatoid arthritis patients is known as Caplan's syndrome.
Figure 25-2. Chest X-ray of a patient with asbestosis.
Figure 25-3, Calcified pleural plaques on the superior border of the diaphragm (arrows) in a patient with asbestosis. Thickening of the pleural margins also is seen along the lower lateral borders of the chest. A, Anteroposterior view. B, Lateral view.
GENERAL MANAGEMENT OF PNEUMOCONIOSIS

Control of occupational diseases is the responsibility of the:
- Worker
- Management
- Community health department
- State and federal governments

Prevention is the key

After the disease is established, it has no effective cure
What are parenchymal lung diseases?
What is Idiopathic Pulmonary Fibrosis?
What is Sarcoidosis?
What are Pneumoconiosis?
THANK YOU