Sarcoidosis

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Definition

- Multi systemic inflammatory disease characterized by the presence of non caseating granulomas, of unknown cause.
- Occurs in young, otherwise healthy adults.
- Women appear to be slightly more susceptible than men.
- At least 5% of patients with sarcoidosis will have a family member with sarcoidosis.
pathology

- noncaseating granulomas are caused by increased local fibroblast stimulation and hence fibrosis
- Mediastinal & superficial lymphadenopathy.
- lung is most commonly affected. Other organs commonly affected are the liver, skin, and eye.
- recent studies show higher incidence of mycobacterial catalase-peroxidase protein in granuloma.
Clinical Manifestations

• 1/3 of patients are asymptomatic (discovered on screening CXR).

• Respiratory complaints including cough and dyspnea are the most common presenting symptoms.

• Nonspecific constitutional symptoms include fatigue, fever, night sweats, and weight loss.
Skin manifestations

• Skin involvement is eventually identified in over a third of patients with sarcoidosis.

• The classic lesions include Erythema nodosum, maculopapular lesions, hyper- & hypopigmentation, keloid formation & subcutaneous nodules.

• specific complex of involvement of the bridge of the nose, the area beneath the eyes, and the cheeks is referred to as lupus pernio (chronic sarcoidosis).
Eye manifestations

• Anterior uveitis, over 1\4 of patients.
• Red eye.
• photophobia, blurred vision, and increased tearing can occur.
• Posterior uveitis, choroidal nodule, papilledema.
Neurologic manifestations

- is reported in 5–10% of patients (Any part of the Central Nervous System or Peripheral Nervous System can be affected).
- Optic neuritis, peripheral neuropathy, bilateral facial palsy, parkinsonism & meningeal involvement.
Other manifestations

• 20–30% of patients will have liver involvement (portal hypertension)

• myalgias and arthralgias.

• Hypercalcemia &/or hypercalciuria occurs in 10% of patients leading to nephrocalcinosis & bone cyst.
Diagnosis

- FBC & ESR (anemia, lymphopenia, pancytopenia)
- Chest x-ray: staging:

<table>
<thead>
<tr>
<th>Stage 0</th>
<th>Normal</th>
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<tbody>
<tr>
<td>Stage I</td>
<td>Hilar lymphadenopathy</td>
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<tr>
<td>Stage II</td>
<td>Hilar lymphadenopathy and parenchymal infiltrate</td>
</tr>
<tr>
<td>Stage III</td>
<td>Parenchymal infiltrate</td>
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<tr>
<td>Stage IV</td>
<td>Fibrosis</td>
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- Increase serum Ca++ & urine Ca++.
- Elevated levels of ACE, but not specific.
Chest radiographs of pulmonary sarcoidosis. A. Stage II sarcoidosis pattern with prominent, discrete “standaway” hilar nodes, right paratracheal adenopathy, and fine reticulonodular infiltrates. B. Fibrocystic sarcoidosis with extensive scarring, bullous and cystic changes, hilar retraction, and parenchymal infiltrates.
Diagnosis

• Tuberculin test, almost always –ve.
• PFT show restrictive defect.
• CT scan show mediastinal lymphadenopathy.
• Kviem-Siltzbach procedure is a specific Diagnostic test for sarcoidosis. An intradermal injection of specially prepared tissue derived from the spleen of a known sarcoidosis patient is biopsied 4–6 weeks after injection. If noncaseating granulomas are seen, this is highly specific for the diagnosis of sarcoidosis.
Diagnosis

• Bronchoscopy & BAL: show lymphocytosis.
• Biopsy of the affected organ.
management

• Acute disease: observation, if no to minimal symptoms without cardiac, neurological, ocular, calcium abnormality.

• Chronic disease: systemic treatment with glucocorticoid e.g prednisone 20-40mg, taper dose within 6 months to 7.5-10mg & continue.

• If steroid toxicity or no response use methotrexate or azathioprine as steroid sparing agents

• Hydroxychloroquine 200-400mg for skin & musculoskeletal manifestations.
management

• ifliximab, thalidomide, cyclophosphamide if other treatments fail to cure the disease.
• Topical steroids for eye manifestations only.
Differential diagnosis

- Pulmonary Tuberculosis (caseating granuloma, +ve AFB).
- Lymphoma
- Berylliosis
- Fungal infection e.g: histoplasmosis
- Hypogammaglobulinaemia and recurrent infections.
COMPLICATIONS

• blindness, paraplegia, or renal failure.
• Heart failure & ventricular arrhythmia from diffuse cardiac muscle involvement.
• Mycetoma with massive hemoptysis.
• Pulmonary arterial hypertension is reported in at least 5% of sarcoidosis patients.