Sarcoidosis

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Epidemiology

• Female > Males
• 20-29 y and women over 50
• Sweden and Iceland ➔ 60/100,000
• High risk group in the US:
  • African Americans ladies
    • More severe
• Higher prevalence in non-smokers
Initial symptoms

• The most common → restrictive lung disease symptoms
• General symptoms → fatigue, night sweats, weight loss, and erythema nodosum
• Organ specific symptoms
• As many as 50% → Asymptomatic
Clinical course

• ~ 2/3rd → remain stable or experience a remission within a decade after diagnosis, with few or no consequences thereafter.
• ~ 20% → pulmonary fibrosis.
• <5% die due to lung fibrosis with respiratory failure or cardiac or neurologic involvement
Poor prognosis

• Stage 2-3 pulmonary disease at the time of diagnosis
• Onset after the age of 40 years
• Black race
• Hypercalcemia
• Splenomegaly
• Osseous involvement
• Chronic uveitis
• Lupus pernio
Good prognosis

• Common early-stage features:
  • fever
  • polyarthritis
  • erythema nodosum
  • bilateral hilar lymph node enlargement

(Löfgren syndrome)

85% remission rate
A

Brain complications
Eye problems (burning, itching, tearing, or pain)
Salivary glands
Enlarged lymph nodes in neck and chest
Heart complications
Granulomas (inflamed lumps) in lungs
Liver enlargement
Spleen enlargement

B

Lupus pernio (painful skin sores on face) and skin lesions on back, arms, neck, face, and scalp

Erythema nodosum (itchy and painful rashes) on the lower legs and ankles

Closeup view of lung cross-section

Enlarged lymph nodes in chest near windpipe and lungs
Scarring and granulomas in lung
Different subtypes

- Annular sarcoidosis
- Erythrodermic sarcoidosis
- Ichthyosiform sarcoidosis
- Hypopigmented sarcoidosis
- Löfgren syndrome
- Lupus pernio
- Morpheaform sarcoidosis
- Mucosal sarcoidosis
- Neurosarcoidosis
- Papular sarcoid
- Scar sarcoid
- Subcutaneous sarcoidosis
- Systemic sarcoidosis
- Ulcerative sarcoidosis
Pathophysiology

• Not well understood
• Accumulation of monocytes, macrophages and activated T-lymphocytes
• TNF-alpha, IFN-gamma, and IL-12, characteristic of a Th1 response
• Increased macrophage and CD4 helper T-cell activation → accelerated but ineffective response
• Anergy → increased risk of infections and cancer
Diagnosis

• Compatible clinical and radiologic findings
• Non-caseous granulomas and the absence of causative organisms
• Ruling out other potential etiologies
For every patient

- Histologic verification
- Assessment of the extent and severity of organ involvement
- Assessment of whether disease is stable or likely to progress
- Determination of whether a patient might benefit from treatment
Lungs

• Primarily an interstitial lung disease → alveoli, small bronchi, and small blood vessels
• 90% of patients
• 50% → permanent pulmonary abnormalities
• 5 -15% → progressive fibrosis
• HRCT vs. CT
# Staging of Sarcoidosis on the Basis of Chest Radiographs

<table>
<thead>
<tr>
<th>STAGE 0</th>
<th>No abnormalities</th>
<th>5%–10%</th>
</tr>
</thead>
<tbody>
<tr>
<td>STAGE 1</td>
<td>Lymphadenopathy (fig. A)</td>
<td>50%</td>
</tr>
<tr>
<td>STAGE 2</td>
<td>Lymphadenopathy + pulmonary infiltration (fig. B)</td>
<td>25%–30%</td>
</tr>
<tr>
<td>STAGE 3</td>
<td>Pulmonary infiltration (fig. C)</td>
<td>10%–12%</td>
</tr>
<tr>
<td>STAGE 4</td>
<td>Fibrosis</td>
<td>5% (up to 25% during the course of the disease)</td>
</tr>
</tbody>
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Typical lung findings

• Lymphadenopathy: bilateral hilar, right paratracheal
• Micro or macro nodules
• Lymphangitic spread: peribronchovascular, subpleural, interlobular septal
• Fibrotic changes
• Bilateral perihilar opacities
• **Upper- and middle-zone predominant**
Reversible vs. irreversible lung lesions

Reversible
• Micronodules, macronodules
• Consolidation
• Ground-glass opacities
• Interlobular septal thickening
• Intralobular linear opacities

Irreversible parenchymal abnormalities
• Honeycomb-like opacities, cysts, bullae, emphysema
• Architectural distortion
• Traction bronchiectasis, bronchiolectasis
• Volume loss in upper lobes, retraction of hila
• Mycetoma (in 10% of patients with end-stage sarcoidosis and a preexisting cavity)
Figure 3. Mediastinal adenopathy in a 60-year-old man.
Pulmonary sarcoidosis in a 26-year-old woman.

Koyama T et al. Radiographics 2004;24:87-104
Axial high-resolution CT scan (pulmonary parenchymal window) shows bilateral enlargement and peripheral calcification of mediastinal and hilar lymph nodes (arrows).
Ground-glass opacities in pulmonary sarcoidosis.

Criado E et al. Radiographics 2010;30:1567-1586
Ground-glass opacities in pulmonary sarcoidosis.

Criado E et al. Radiographics 2010;30:1567-1586
Axial high-resolution CT scan shows mediastinal lymph node enlargement and a reticular pattern produced by nodularity and thickening of interlobular septa, pleural surfaces, and fissures, features that are seen in lymphangitic carcinomatosis as well as.
Heart

• In 25% of patients, often conductive system
• Myocardial involvement ➔ 5%
• MR:
  • Increased signal intensity on T2
  • Enhancement on contrast-enhanced T1
  • Limited MR use due to pacemaker
• Gallium-67 scintigraphy
  • low sensitivity and specificity
  • helpful in monitoring disease activity
Cardiac sarcoidosis in a 59-year-old woman with abnormal electrocardiographic findings.
Cardiac sarcoidosis in a 60-year-old man who presented with complete atrioventricular blockage.

Koyama T et al. Radiographics 2004;24:87-104
CNS

- Up to 25% of patients
- Clinically recognizable in <10%
- Manifestations and prognosis are variable
- Imaging findings mimic infectious or metastatic disease
- CSF analysis might help:
  - increased ACE titer
  - Increased CD4:CD8 ratio
Neurosarcoidosis in a 24-year-old man who presented with diabetes insipidus.

Axial T2-weighted MR image demonstrates an isointense periventricular lesion (arrow) surrounded by minimal high-signal-intensity edema.

On a contrast-enhanced T1-weighted MR image, the lesion demonstrates enhancement (arrow).
Neurosarcoidosis.

Koyama T et al. Radiographics 2004;24:87-104
Leptomeningeal

• Commonly affecting the base of the brain → aseptic meningitis
• Contrast-enhanced T1-weighted imaging diagnostic.
• Less common → small enhancing nodules on the brain surface and in the perivascular spaces
• Brain and spinal cord involvement occur in the early stage of the disease and responds rapidly to steroid treatment.
Spinal cord

• Cervical and thoracic spinal regions
• T2-weighted $\rightarrow$ intramedullary lesion with decreased signal intensity.
• Enlarged spinal cord with high signal intensity due to associated edema.
• Sarcoid granuloma $\rightarrow$ enhancement on T1
Leptomeningeal involvement in a 23-year-old woman.
Leptomeningeal involvement in a 34-year-old man who presented with bilateral facial palsy.

Koyama T et al. Radiographics 2004;24:87-104
Spinal cord involvement in a 23-year-old man who presented with mild paresis of the hands.
Eyes

• In up to 80% of patients
• Uveitis ➔ the most common condition and is typically bilateral.
• Acute uveitis usually resolves spontaneously or responds to local corticosteroid therapy (eyedrops).
• Lacrimal glands involvement less common
Ocular involvement in the same patient as in Figure 1.
Ocular involvement in the same patient
Liver and spleen

- 50%–80% of autopsy specimens
- Dysfunction of these organs is uncommon.
- Isolated hepatosplenic sarcoidosis do not require treatment.
- Splenic nodules are larger and more common than hepatic
- Multiple nodules in hepatic sarcoidosis are easily mistaken for more common diseases, including metastases and lymphoma
Hepatosplenic involvement in a 49-year-old woman with pulmonary sarcoidosis.
Figure 28a. Bone sarcoidosis in a 28-year-old man who presented with left thumb pain.

Koyama T et al. Radiographics 2004;24:87-104
MSK

• Usually in patients with generalized disease
• Joint involvement is common
• Inflammatory arthralgia (up to 40% of cases) → knees, ankles, elbows, and wrists.
• Erythema nodosum, periarticular ankle inflammation, and mediastinal lymphadenopathy → Löfgren syndrome → self-limiting course with spontaneous resolution

• Skeletal involvement → phalanges in the hands and feet → 5%–10% of patients

• Radiologic features:
  • cystlike radiolucent areas
  • a lacelike honeycomb appearance
  • extensive bone erosion with pathologic fractures
  • subcutaneous soft-tissue mass or tenosynovitis may also be present. The combination of these radiologic features is virtually diagnostic.
Bone sarcoidosis in a 28-year-old man who presented with left thumb pain

Koyama T et al. Radiographics 2004;24:87-104
Bone sarcoidosis