Thoracic sarcoidosis: Pictoral review of typical and atypical findings

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Authors: A. Ferreira, J. Calha; Lisbon/PT
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Learning objectives

- To review and illustrate the manifestations of thoracic sarcoidosis in plain radiographs and in high resolution computed tomography (HRCT), including typical and atypical findings, in patients with biopsy proven pulmonary sarcoidosis.
- To highlight the more useful findings in the differential diagnosis with other entities.
- To distinguish reversible from irreversible disease.

Background

Sarcoidosis is a systemic disease of unknown etiology, characterized by the presence of noncaseating granulomas. It may involve almost any organ, but most morbidity and mortality result from pulmonary disease, which is present in 90% of patients. The pulmonary disease may resolve or, in approximately 20% to 25% of cases, evolve to irreversible fibrosis. Based on plain radiographic findings, sarcoidosis is classified in stages. These stages have some correlation with the course of the illness.

It is thought that sarcoidosis results from exposure of genetically susceptible individuals to environmental agents. Sarcoidosis is associated to a great variety of connective tissue diseases such as rheumatoid arthritis, ankylosing spondylitis, among others.

It is most common between 20 to 40 years old, with a second peak of incidence in females after 50 years old. Has a slight female predominance.

Mortality is 1-5% of the affected individuals.

Thirty to fifty percent of the patients are asymptomatic. Symptomatic patients present respiratory complaints in 50% of cases, which include dyspnea, cough and chest pain. May also present with constitutional symptoms such as weight loss, fatigue, weakness and malaise. Usually has an insidious onset and is associated to a multisystem involvement. Five percent of the patients develop pulmonary hypertension, which seems not be involved with the fibrosis grade.

Sarcoidosis results from the accumulation of activated macrophages and T-cell lymphocytes that ultimately result in the formation of noncaseating granulomas. In the early stage the granulomas are distributed along the bronchovascular bundles,
lymphatics and interlobular septa. Late in the illness the granulomas become confluent and undergo fibrosis.

**Imaging findings OR Procedure details**

In this educational exhibit we show the wide spectrum of chest imaging findings of sarcoidosis, including lymph node abnormalities and lung disease.

**Radiographic Findings**

Based on plain radiographic findings, sarcoidosis is classified in stages. These stages have some correlation with the course of the disease.

<table>
<thead>
<tr>
<th>Stage</th>
<th>Radiographic Findings</th>
<th>Resolution</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Normal</td>
<td></td>
<td>10%</td>
</tr>
<tr>
<td>I</td>
<td>Mediastinal and hilary lymphadenopathy</td>
<td>55-90%</td>
<td>40%</td>
</tr>
<tr>
<td>II</td>
<td>Lymphadenopathy + parenchymal disease</td>
<td>40-70%</td>
<td>35%</td>
</tr>
<tr>
<td>III</td>
<td>Parenchymal disease</td>
<td>10-20%</td>
<td>10%</td>
</tr>
<tr>
<td>IV</td>
<td>Fibrosis</td>
<td>0%</td>
<td>5%</td>
</tr>
</tbody>
</table>

- Lymphadenopathy
  - Bilateral and symmetrical
  - Bilateral hilar and right paratracheal lymph node enlargement - common
  - Less common locations - aorticopulmonary window, subcarinal region and anterior mediastinum
  - Hilary unilateral - rare
  - May compress the bronchi causing atelectasis
  - Calcification - 5% in the early stage; more than 20% after 10 years of disease
    - Eggshell calcification - rare
- Parenchymal disease

  - Bilateral and symmetrical
  - Predominate in upper lobes
  - Most frequent patterns - nodular and reticulonodular
  
  - Nodular pattern
    - Thirty to 60%
    - irregular margins
    - upper and middle predominance
    - 1-10 mm diameter
  
  - Reticulonodular pattern
    - Twenty-five to 50%
    - Nodules accompanied of interlobular septal thickening, or nodules and intralobular linear opacities
  
  - Parenchymal consolidation
    - Ten to 20%
    - Bilateral and symmetrical, involving mainly the upper and middle lung zones
  
  - Fibrosis
    - Five percent at presentation, 20-25% later in the course of the disease
    - Perihilar distribution with upper and middle predominance
    - Associated to superior retraction of the hila, distortion of the bronchovascular bundles, bulla formation, traction bronchiectasis and compensatory overinflation of the inferior lobes
  
  - Cavitation
    - Less frequent
    - Spontaneous resolution or superinfection and mycetoma formation (1-3%)

- Pleural Disease

  - Pleural effusion - 3% - resolution within 4 to 8 weeks or pleural thickening
  - Spontaneous pneumothorax - 1%

Computed Tomography

- Pulmonary Manifestations

  - CT provides better characterization of disease's distribution and extent
  - HRCT findings correlates with the histologic findings
  - Most common finding - small nodules in perilymphatic distribution
    - Are seen on HRCT at initial evaluation in 90-100%
    - Reflect confluence of microscopic granulomas
    - Smooth/irregular margins, well defined, 2-5mm in diameter
• Adjacent to bronchi and pulmonary vessels, along the interlobular septa, interlobular fissures, and costal subpleural regions - thickening of these structures
• Nodules may extend to the peribronchiolar interstitium - centrlobular nodules
• Conglomeration of subpleural granulomas - pleural pseudoplaques
• Confluence of granulomas - large nodules or masses
  • 1-4 cm in diameter
  • are seen in 15-25%
  • predominate in the upper lobes and the peribronchovascular regions
  • occasionally may cavitate
• Extensive microscopic interstitial granulomas - ground-glass opacities or areas of consolidation
• Regular and nodular bronchial wall thickening
• Bronchial stenosis - uncommon
• Mosaic attenuation pattern
• Air trapping in expiratory scans - 90% of patients
• Fibrosis
  • Irregular linear opacities
  • Irregular septal thickening
  • Traction bronchiectasis
  • Honeycombing

<table>
<thead>
<tr>
<th>Reversible Disease</th>
<th>Irreversible Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nodules</td>
<td>Irregular lines</td>
</tr>
<tr>
<td>Interlobular septal thickening</td>
<td>Reticular opacities</td>
</tr>
<tr>
<td>Ground-glass opacities</td>
<td>Architectural distortion</td>
</tr>
<tr>
<td>Consolidation</td>
<td>Honeycombing</td>
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</tbody>
</table>

- Pulmonary Arterial Hypertension
  • Usually results from extensive fibrosis
  • Other causes - extrinsic compression, pulmonary veno-occlusive disease, granulomatous vascular involvement and pulmonary vasoconstriction by vasoactive factors
  • Diameter of the main pulmonary artery > 29 mm or, diameter of the main pulmonary artery/diameter of the ascending aorta > 1

- Hilar and Mediastinal Lymphadenopathy
  • Are seen in approximately 90% of patients
• Most common enlarged lymph nodes - paratracheal and hilar (virtually all patients)
• Also common - aorticopulmonary window (90%); subcarinal (60%); anterior mediastinal (50%); posterior mediastinal (15%)
• Axilla, internal mammary chain and retrocrural region - rare
• Calcification - 40-50%
  • Most common bilateral, hilar, paratracheal, and subcarinal
  • Mostly focal, but may be diffuse or eggshell

Images for this section:

Fig. 1
LYMPHADENOPATHY

Calcified lymphadenopathy
- 40-50%
- Hilar, right paratracheal and subcarinal - common
- May be focal, but may be diffuse or eggshell

Differential Diagnosis
- Tuberculosis
- Silicosis

Fig. 2
PARENCHYMAL DISEASE

Nodular/Reticulonodular opacities
- Most common finding - small nodules in perilymphatic distribution
- Are seen on HRCT at initial evaluation in 90-100%
- Smooth/irregular margins, well defined, 2-5mm in diameter
- Distributed along the bronchovascular bundles, lymphatics and interlobular septa.
- Involve mainly the upper and middle lung zones

Differential Diagnosis
- Lymphangitic carcinomatosis
- Lymphoproliferative disorder
- Silicosis

Fig. 3
PARENCHYMAL DISEASE

Centrilobular nodules
• Less frequent finding
• Nodules extend to the peribronchiolar interstitium

Differential Diagnosis
- Hypersensitivity Pneumonitis
- RB-ILD

Fig. 4
PARENCHYMAL DISEASE

Ground-glass attenuation
- Present in 40% of the patients
- More frequent in smokers
- Extensive microscopic interstitial granulomas

Differential Diagnosis
- PCP
- Hypersensitivity Pneumonitis

Fig. 5
PARENCHYMAL DISEASE

Consolidation
- 10-20%
- Are seen on HRCT at initial evaluation in 90-100%
- Bilateral consolidation with air bronchogram
- Involve mainly the middle lung zones

Differential Diagnosis
- Tuberculosis
- COP
- Lymphoma

Fig. 6
PARENCHYMAL DISEASE

Large nodules or masses
- Are seen in 15-25%
- 1-4 cm in diameter
- Predominate in the upper lobes and the peribronchovascular region
- Occasionally may cavitate

Differential Diagnosis
- Silicosis
- Lymphoma

Fig. 7
**Fig. 8**

**PARENCHYMAL DISEASE**

**Fibrosis**
- Irregular linear opacities
- Irregular septal thickening
- Traction bronchiectasis
- Honeycombing rare
- Perihilar distribution with upper and middle predominance

**Differential Diagnosis**
- UIP
- Chronic Hypersensitivity Pneumonitis
Conclusion

Sarcoidosis has a wide spectrum of manifestations, hence many different diseases must be considered in the differential diagnosis. HRCT plays an important role in the presumptive diagnosis of sarcoidosis, and enables differentiation of reversible from irreversible disease.

Personal Information

Ana Ferreira, Hospital Pulido Valente, CHLN
anacavalo@netcabo.pt

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