The Radiology of Pulmonary Sarcoidosis

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Outline

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Introduction

- What is sarcoidosis?
  - Sarcoidosis is a chronic noncaseating granulomatous disease of unknown etiology that affects many organs and tissues.
- The organs most affected are as follows:
  - Lungs 90%
  - Lymphatics 75%
  - Skin/Eyes/Joint 25%
  - Bone marrow/Spleen 30-40%
  - Liver 60-90%
  - CNS/MSK/Heart 5%
- Prevalence
  - Estimated at 10-40 per 100,000 with slight female preponderance
  - Higher among U.S. blacks than whites by at least 10:1
  - Individuals aged 20-40 years are at highest risk.
Introduction cont’d

- **Disease Presentation**
  - 50% of cases are discovered incidentally on CXR in asymptomatic individuals.
  - Most common presenting sx are cough, dyspnea and chest pain.
  - Clinicians beware: MANY other presentations are possible.
  - 40% acute onset & constitutional vs. 60% chronic & pulmonary

- **Diagnosis (* indicates required criteria)**
  - *Characteristic clinical and radiologic findings*
  - *Tissue biopsy evidence of noncaseating granuloma(s)*
  - *Negative bacterial and fungal cultures (R/O infection, especially TB)*
  - Pulmonary Function Tests: low FVC, nl FEV1/FVC, low DLCO
  - Elevated serum ACE

- Treatment of symptomatic individuals is with steroids & immunosuppressives \(\rightarrow\) lung transplant for severe disease.

- Prognosis: Two thirds of cases resolve spontaneously, one third are long-term, and 5% result in fatality.
Anatomy

- Normal hilar markings
- Normal interstitial markings
- Right paratracheal area
- Aortopulmonary window

http://www.vh.org/adult/provider/radiology/LungAnatomy/

www.uptodate.com
Plain Film Chest Radiography

- CXR (PA & lateral) is the initial diagnostic study of choice.
- CXR is also useful in monitoring disease progression.

**Advantages**
- Fast
- Cheap
- Widely available
- Relative ease of interpretation

**Limitations**
- CXR is a useful anatomical guide to lung involvement but it cannot stage the biological activity of the disease process and cannot assess functional defects.
- CXR is not as sensitive for sarcoidosis as CT.
CXR: Sarcoid Stage I

- Enlarged **bilateral hilar**, right paratracheal (arrow), and aortopulmonary window (arrowhead) **nodes**
- Normal parenchyma
- Ddx
  - Sarcoidosis
  - Lymphoma/Leukemia
  - Tuberculosis
  - CA Mets
  - Fungal Infection

www.meddean.luc.edu/lumen/MedEd/Radio/sarc/sarc.htm
CXR: Sarcoid Stage II

- Bilateral hilar adenopathy
- Fine linear and reticular opacities in perihilar lung parenchyma

www.uptodate.com
CXR: Sarcoid Stage III

- Interstitial disease, now *without* hilar LAN
- Ddx:
  - CHF
  - lymphangitic spread of CA
  - infection (viral, mycoplasma)
  - pneumoconiosis
  - collagen vascular disease
  - Sarcoidosis
CXR: Sarcoid Stage IV

- Interstitial opacities with upper zone predominance, volume loss, and advanced fibrosis.
- Ddx
  - Collagen vascular disease (RA, scleroderma)
  - Sarcoid stage IV
  - Silicosis
  - Asbestosis
  - Hypersensitivity pneumonitis
  - Idiopathic fibrosis
  - Drug/radiation toxicity

www.meddean.luc.edu/lumen/MedEd/Radio/sarc/sarc.htm
High Resolution CT Scan

- HRCT uses very thin slices (1mm) to achieve better spatial resolution & precision.
- HRCT is indicated after normal CXR in a symptomatic patient - the setting of high clinical suspicion of disease.

Advantages
- High sensitivity for adenopathy, infiltrates, and architectural distortion.
- HRCT can identify areas of reversible vs. irreversible lung damage.

Disadvantage
- Expensive
CT: Sarcoid Stage I

- Bilateral hilar adenopathy
- **Ddx**
  - Sarcoidosis
  - Tuberculosis
  - CA Mets
  - Fungal Infection
  - Lymphoma/Leukemia

www.uhrad.com
CT: Sarcoid Stage II

- Bilateral hilar lymph node enlargement
- Multiple miliary peribronchiolar nodules scattered diffusely throughout both lungs.
CT: Sarcoid Stage III

- Beaded or irregular thickening of bronchovascular bundles with nodules along bronchi, vessels, and subpleural regions.
- Normal hilar lymph nodes
- Ddx:
  - CHF
  - Lymphangitic spread of CA
  - Infection (viral, mycoplasma)
  - Pneumoconiosis
  - Collagen vascular disease
  - Sarcoidosis

www.uptodate.com
CT: Sarcoid Stage IV

- Broad bands of fibrosis in the upper lobes.
- Ddx
  - Collagen vascular disease (RA, scleroderma)
  - Sarcoid stage IV
  - Silicosis
  - Asbestosis
  - Hypersensitivity pneumonitis
  - Idiopathic fibrosis
  - Drug/radiation toxicity

www.meddean.luc.edu/lumen/MedEd/Radio/sarc/sarc.htm
Gallium 67 Scintigraphy

- A nuclear medicine test used largely to support diagnosis
- Radioactive gallium is injected intravenously and imaged 1-2 days later.

Advantages
- Non-invasive
- Serial exams monitor disease progression
- Gallium distinguishes areas of fibrosis from areas of inflammation
- Full-body scan detects extra-pulmonary spread of disease

Limitations
- low specificity
- difficult interpretation.
Whole Body Gallium Scan

www.acta-clinica.kbsm.hr/Acta5/KOVACIC.PDF
1/26/01 – EM is a 40 yo previously healthy female p/w fever, cervical and posterior auricular adenopathy, intermittent HA and neck pain/stiffness, intermittent right upper extremity weakness.

- ESR was elevated. She was seropositive (IgM) to both EBV & CMV.

- Radiologic studies were done...
**Patient Presentation: MR**

- **Rt paraspinal soft tissue mass**
  - Infiltrates epidural space at T1/T2
  - Bony erosion of posterior vertebral elements
  - No cord compression or disc herniation apparent

- **Ddx:**
  - Infection (CMV, EBV, other)
  - Neoplasm (lymphoma)
  - The mass is found to be a cryptococcal abscess. EM’s sx resolve with medication.

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[B1 DMC] [B1 DMC] [B1 DMC]
Patient Presentation: CXR

- 3/26/02 – EM returns now c/o adenopathy and discomfort spreading down to chest and back. She describes “heaviness in her lungs” without frank dyspnea or cough. She also has a new skin lesion on the right forearm.
- Initial CXR is unremarkable.
- Further radiologic evaluation is performed…
Patient Presentation: CT

- Bilateral upper lobe infiltrates with nodular septal thickening
- Axillary adenopathy
Patient Presentation: CT

- Normal hila
- Normal bases
- Ddx
  - CHF
  - lymphangitic spread of CA
  - infection (cryptococcus)
  - pneumoconiosis
  - collagen vascular disease
  - Sarcoidosis, Stage III
Patient Presentation: Dx & Mgmt

- **5/13/02** - EM is diagnosed with sarcoidosis:
  - Radiographic evidence as discussed
  - Cultures are negative.
  - Skin lesion bx reveals non-caseating granulomas.
  - Serum ACE is elevated.

- Because she is largely asymptomatic, she decides – in concert with her physician – to conservatively monitor her disease and forego treatment for now.

- **1/26/03** – repeat chest CT is unchanged. EM continues to be asymptomatic and clinically stable.
Summary

- Sarcoidosis is a chronic non-caseating granulomatous disease, typically affecting the lungs with or without extra-pulmonary involvement.

- Stage-specific findings are recognizable on CXR & CT allowing us to track progression of disease over time.
  - Stage I: bilateral hilar adenopathy
  - Stage II: “along with diffuse reticular and nodular interstitial markings with upper zone predominance
  - Stage III: interstitial disease, now without hilar adenopathy
  - Stage IV: fibrosis

- Ddx includes infection (esp. TB), lymphoma & CA mets: these must be ruled out in order to make the diagnosis of sarcoidosis. Other diagnostic criteria are radiographic and pathologic evidence consistent with sarcoidosis.

- The prognosis is usually favorable.
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