Sarcoidosis

Pathophysiology, Diagnosis, and Treatment

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Fever of Unknown Etiology

• M. S. is a 52 yo white F with a history of scleroderma and cholelithiasis who presented with dry cough, shaking chills, and fevers to 102 F despite being put on amp/gent/flagyl
Chest film

- There were no abnormalities identified
In view of her significant presentation, a chest and abdominal CT were ordered.

**Chest CT**

- A chest CT showed mediastinal lymphadenopathy, and an interstitial reticulo nodular pattern predominantly in the upper lobes.
Ddx Upper Lobe Interstitial Process

- Silicosis
- Sarcoid
- Atypical PCP
- Hypersensitivity Pneumonitis
- Ankylosing spondylitis
- Langerhans cell histiocytosis
Patient’s Abdominal CT

Abdominal CT showed:

- 1.8 cm gallstone without signs of cholecystitis.
- Multiple low-attenuation splenic lesions

8mm hepatic cyst (incidental finding)

Low-attenuated splenic lesions
Lung and spleen findings suggest Sarcoidosis. Splenic and lung biopsies were obtained and showed non caseating granulomata confirming the diagnosis of Sarcoidosis.
The patient underwent a laparoscopic cholecystectomy and splenectomy. Her fever and night sweats abated and she was discharged without further treatment.
Follow up Chest X-ray

- 4 mo later she came back for outpatient f/u and her CXR showed she had developed hilar adenopathy.
Repeat Chest CT

- Mediastinal windows showed:
  - Hilar and mediastinal lymph node enlargement
  - Hilar adenopathy
  - Mediastinal (subcarinal) adenopathy
Repeat Chest CT

Lung windows showed:
Numerous small sub-cm lung nodules bilaterally
Chest findings are compatible with progression of stage 11 sarcoidosis
Sarcoidosis

- F > M, 20-40 yo
- United States: black > white 10:1 – 17:1
- Europe: disease affects mostly whites
- Chronic multisystem disorder of unknown etiology
- Noncaseating granulomas in affected organs: T lymphocytes and macrophages
- Most frequently symptomatic organs:
  - Lung 90%
  - Lymph nodes 75-90%
  - Skin/eye/liver 25% each
  - Bone marrow/spleen 15-40%
  - CNS/MS/heart 5% each
Disease Presentation

• Most patients have some respiratory symptoms
• Subacute sarcoidosis: 20-40%
  – Fever, malaise, anorexia, or weight loss
  – +/- Dry cough, dyspnea, retrosternal chest discomfort
  – Often self-limited
• Chronic sarcoidosis: 40-70%
  – Dry cough, dyspnea, or retrosternal chest discomfort
  – +/- Fever, malaise, anorexia, weight loss
  – May progress to permanent lung and secondary organ damage
Diagnosis

Required for diagnosis:

• History / Physical exam
• Negative blood tests and positive pulmonary function tests
• CXR indicative of sarcoidosis
• Biopsy evidence of noncaseating granulomatous process
  – lung parenchyma (usual site, via bronchoscopy)
  – hilar nodes, skin, conjunctiva, lip
Presentation of Pulmonary Sarcoid on Chest XRay
CXR: Sarcoid Stage I

- In stage I disease, there is hilar (± mediastinal) adenopathy without interstitial infiltrate.
Differential for Hilar Adenopathy

- Tb - usually unilateral
- Sarcoid - usually bilateral, symmetric
- Fungal infection
- Metastatic disease - especially renal, thyroid
- Lymphoma/leukemia
- Benign lymph node hyperplasia (Castleman’s disease)
CXR: Sarcoid Stage II

- In stage II disease, hilar adenopathy is accompanied by interstitial infiltrate, which tends to be more prominent at the apices as in this patient.
CXR: Sarcoid Stage III

- In stage III disease, hilar LAN has disappeared and diffuse interstitial infiltrates remain
- Differential includes: CHF, lymphangitic spread of CA, infection (viral, mycoplasma), sarcoid, pneumoconiosis, collagen vascular disease

From: Teaching file of Dr. Philip Boiselle; Beth Israel Deaconess Medical Center, Boston MA
CXR: Sarcoid Stage IV

• Presents with fibrosis as in this radiograph
CXR: Sarcoid Stage IV

- Sometimes can cause cystic spaces on CXR, aka “cystic sarcoid”
Differential for fibrosis

- Collagen vascular disease (RA, scleroderma)
- Sarcoid stage IV
- Silicosis
- Asbestosis
- Hypersensitivity pneumonitis
- Idiopathic fibrosis
- Drug/radiation toxicity
Pulmonary Sarcoid on Chest CT
Sarcoid on CT

1. Hilar and mediastinal adenopathy

- Symmetric hilar adenopathy
- Bulky mediastinal adenopathy (precarinal)
Sarcoid on CT

• 2. Interstitial markings and perilymphatic nodules
Sarcoid on CT

- 3. Fibrosis (end-stage)
- “Honey combing”

HRCT Findings in Sarcoid

- 1. Perilymphatic nodules, 1-10 mm, often subpleural, may calcify
- 2. Patchy, often asymmetric, distribution
- 3. Upper lobe predominance
- 4. Hilar and mediastinal LAN (not always; may calcify)
- 5. Ground-glass opacity (uncommon), indicating the presence of small granulomas
Treatment of Sarcoidosis

- Treat early - Permanent organ derangements are not responsive to glucocorticoids
- Oral prednisone
- Repeat treatment protocol for reactivations
- Other considerations
  - inhaled glucocorticoids not efficacious
  - mild ocular disease: local therapy
  - uveitis: systemic therapy
References

- BIDMC, BIDMC Patient Care Radiology Files. 2000. Beth Israel Deaconess Medical Center, Boston MA. SLIDE#3, 4, 5, 7, 8, 9
- Boiselle, P. 2000. Personal teaching file, Beth Israel Deaconess Medical Center, Boston MA. SLIDE#13, 15, 16, 18, 21, 22
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The End