An Unusual Presentation of Pulmonary Sarcoidosis

Emily Barsky, Harvard Medical School, Year IV
Gillian Lieberman, MD
Our Patient: Ms. WS

- 35F with asthma, recurrent pneumonia, and remote history of intravenous drug use who presents with sudden onset right-sided chest pain and shortness of breath
  - Woke from sleep with sharp, stabbing chest pain
- Seen in emergency department one week prior for chest pain, fevers, productive cough
  - Diagnosed with “multifocal pneumonia” and sent home with course of levofloxacin
Our Patient: History

- **Review of Systems**: Lesions on face and bilateral upper extremities for several months. Review of systems otherwise negative
- **Past Medical History**: Asthma, diabetes, recurrent pneumonia
- **Family History**: No family history of pulmonary disease, cancer, autoimmune disease
- **Social History**: From Puerto Rico, moved to US in 1995, no recent travel. Lives in East Boston, homeless in past. Has positive TB contacts and prison contacts. Positive history of intravenous drug use but none in last 8 years, social alcohol use, 5 pack-year past smoking history. Negative HIV test 1-2 mos ago, no prior PPD.
Our Patient: Physical Exam

- Afebrile, HR 123, BP 114/60, Sp02 97% on 2L O2
- Pulmonary: No breath sounds on right
- Cardiac: No murmur
- Skin: Erythematous, violaceous plaques with scale on forehead, nose and at tattoo sites on upper extremities bilaterally

- CXR was obtained given the patient’s acute dyspnea, chest pain, and hypoxia...
Our Patient: Initial AP CXR

What are the findings?

Image source: MGH, CAS
Our Patient: Right Tension Pneumothorax

- Pneumothorax
- ? Possible lucency
- Collapsed lung tissue
- Mild rib splaying
- Depression of R hemidiaphragm
- Mediastinal Shift

Image source: MGH, CAS
Pneumothorax: Types

SIMPLE
- Small volume lung collapse (generally <30%)
- Patient remains relatively asymptomatic
- Signs on CXR:
  - White visceral line, marking lung/pleural air interface; straight or convex towards chest wall
  - No pulmonary vasculature visible beyond this line
  - No mediastinal shift or occasionally mediastinal shift TOWARDS defect

TENSION
- CLINICAL diagnosis
  - Patient is symptomatic
  - Usually only occurs with large pneumothoraces
- One-way valve: air enters pleural space on inspiration but can’t exit on expiration→ pressure in pleural space builds up→ pleural pressure exceeds atmospheric pressure→ further lung compression→ respiratory failure
- In extreme cases, can lead to cardiovascular collapse (impaired venous return to inferior and superior vena cava)
- Signs on CXR:
  - Visceral pleural line with lack of pulmonary vasculature beyond that point
  - Mediastinal shift AWAY from defect (once pleural pressure sufficiently high)
  - +/- depression/inversion of ipsilateral hemidiaphragm
  - +/- splaying of ipsilateral ribs

Pneumothorax: Classification

- **Primary Spontaneous Pneumothorax**
  - Spontaneous rupture of a pleural bleb
  - Often occurs in thin, tall men
  - Tends to recur
  - Risk factors:
    - Smoking
    - Male gender
    - Family history
    - Marfan’s

- **Secondary Spontaneous Pneumothorax**
  - Usually due to underlying lung disease. Can occur with any lung disease, but most commonly:
    - Chronic Obstructive Pulmonary Disease (70%)
    - Cystic Fibrosis
    - Mycobacterium Tuberculosis
    - Pneumocystis Carinii Pneumonia
    - Interstitial Lung Disease
Pneumothorax: Management

- Small pneumothorax:
  - If <30% hemithorax involvement and if clinically stable --> observe closely
  - Supplemental oxygen (hastens reabsorption)
  - Usually spontaneously resolve, occasionally requires needle aspiration

- Large pneumothorax:
  - If >30% hemithorax involvement or clinically unstable --> requires treatment
  - Needle aspiration, chest tube placement (thoracostomy)

- Tension pneumothorax:
  - Requires emergent chest tube placement
  - If with severe hemodynamic or respiratory compromise, can first decompress with subcutaneous needle, then place chest tube

- What was done for our patient? Given the tension pneumothorax, a chest tube was placed immediately
Our Patient: After Chest Tube Placement

1. Chest tube
2. Small residual R pneumothorax
3. Subcutaneous emphysema

Image source: MGH, CAS
Our Patient: Work-Up

- But why did our patient have a pneumothorax?
- Let’s first look at her CXR from the week prior....
Our Patient: Prior CXR

- Blurring of diaphragms R>L suggesting bibasilar atelectasis vs infiltrates
- Ill-defined areas of opacification throughout both lung fields
- ?Lucency

PA plain film from one week PTA

Image source: MGH, CAS
Our Patient: Prior CXR Cont’d

Focal area of opacity in anterior lung, but difficult to ascertain which side it is on

ED Conclusion one week prior: “Multifocal pneumonia”

Regardless, still no clear cause for tension pneumothorax...

So let’s do a CT

Image source: MGH, CAS
Our Patient: Chest CT without Contrast

Residual pneumothorax

Subcutaneous emphysema

Cavitary lesion in RLL

Lung nodule

Probable Atelectasis
Our Patient: Coronal and Sagital Reconstructions

RLL Cavity    Chest tube
Subcutaneous emphysema

Minor fissure
Major fissure
RLL nodule

Image source: MGH, CAS
Our Patient: Lung Nodules

1.1 cm nodule in RLL

Sub-cm LLL nodule

Image source: MGH, CAS
Our Patient: Bronchial Thickening

- This is an axial cut superior to the location of the cavitary lesion.
- Notice the bronchial and peribronchial thickening on the right, as well as the micronodules and nodular opacities.

Image source: MGH, CAS
Our Patient: Lymphadenopathy

Coronal Reconstruction, Soft Tissue Window

- The soft tissue window allows for visualization of the mediastinum and detection of lymphadenopathy.

- While the absence of contrast renders it suboptimal, there is fullness suggestive of mediastinal, hilar, and subcarinal lymphadenopathy.

Image source: MGH, CAS
Our Patient: Summary of Findings

- RLL cavitory lesion
- Multiple nodules and micronodules
- Bronchial and peribronchial thickening
- Mediastinal, hilar and subcarinal adenopathy

- However, her diagnosis is still unclear
Cavitary Lung Lesions: Differential

- **Infection**
  - Abscess (especially anaerobes, Nocardia, S. aureus, Klebsiella)
  - Mycobacterium (TB, nontuberculous)
  - Fungi

- **Septic emboli** - endocarditis

- **Rheumatologic/Vasculitis**
  - Most commonly Wegener’s
  - Others (rarely): Sarcoidosis, SLE, RA
Cavitary Lung Lesions: Differential Cont’d

- **Neoplasm**
  - Primary
    - Especially squamous cell carcinoma
    - Others: lymphoma, Kaposi’s Sarcoma
  - Metastatic (i.e. osteosarcoma)

- **Misc**
  - Empyema, pulmonary embolism, Langerhan’s cell histiocytosis, bronchiolitis obliterans organizing pneumonia
Our Patient: Work-up Cont’d

- Given significant TB risk factors, induced sputum x 3 --> negative for acid fast bacilli
- Bronchoscopy with bronchoalveolar lavage --> no organisms

- But what about the skin findings?
Companion Patient #1: Skin Lesions in Tattoo

- Derm performed skin biopsy...
- Path: granulomatous dermatitis

Our patient’s diagnosis? Sarcoidosis +/- lung superinfection


Sarcoidosis

- Systemic granulomatous disease of unknown etiology affecting multiple organ systems

- Epidemiology:
  - Usually presents between ages 20-40

- Organ system involvement:
  - Lung > 90%
  - Skin 25% (varied presentations)
  - Occular 20-50% (i.e. uveitis)
  - Liver (hepatitis) 50-80%
  - Other organs systems: musculoskeletal (myositis, polyarthritis), reticuloendothelial (lymphadenopathy, hepatosplenomegaly), cardiac (arrhythmias), renal (focal segmental glomerulosclerosis, membranous nephropathy, crescentic), neurologic
Sarcoidosis Cont’d

○ Signs and symptoms:
  ● Up to 50% asymptomatic, diagnosed on routine CXR
  ● 1/3 have nonspecific symptoms (fever, malaise, fatigue, weight loss, decreased exercise tolerance)
  ● Organ specific symptoms (i.e. cough, chest pain, dyspnea in the case of pulmonary sarcoid)

○ Diagnosis:
  1. Clinical and radiologic features consistent with disease
  2. Noncaseating granulomas on histology (transbronchial lung, skin, or lymph node biopsy most commonly)
  3. Exclusion of other similar diseases
Pulmonary Sarcoidosis: Stages

- Stage 0: Normal CXR
- Stage 1: Bilateral hilar adenopathy
- Stage 2: Bilateral hilar adenopathy and reticular opacities/infiltrates (upper zones>lower zones)
- Stage 3: Reticular opacities/infiltrates, resolved hilar adenopathy
- Stage 4: Fibrosis (irreversible)

Pulmonary Sarcoidosis: Companion Patient #2

Early Stage

- Notice the bilateral hilar adenopathy typical of early stage sarcoidosis
- Also notice the reticulonodular opacities, most prominent in the RUL

PA Plain film
Pulmonary Sarcoidosis: Companion Patient #3

**PA Plain Film**
- Extensive mediastinal and hilar lymph node calcification

**Late Stage**
- Upper lobe volume loss secondary to fibrosis
- Mass-like area of fibrosis
- Traction bronchiectasis

**Coronal CT View**
Pulmonary Sarcoidosis: Other Radiological Findings

- Nodules (multiple, often subpleural)
- Bronchial wall thickening
- Focal ground glass
- Bronchovascular beading
- Apical cysts
- Honeycombing
- Parenchymal masses or consolidation
- Others: traction bronchiectasis, cavitation, pleural disease
Pulmonary Sarcoidosis: Treatment and Prognosis

- **Treatment**: Corticosteroids, cytotoxic agents, immunomodulators
- **Prognosis**: Depends on host characteristics, organ involvement, disease extent
  - Spontaneous resolution in:
    - Stage 1: 90% of patients
    - Stage 2: 70% of patients
    - Stage 3: 20% of patients
  - Permanent functional impairment in 20-25%
Sarcoidosis: Role of Imaging

- **CXR**
  - Initial CXR with staging, prognostic power
  - Suboptimal for evaluating parenchymal abnormalities

- **High Resolution CT**
  - Less prognostic power than CXR
  - Excellent for evaluating parenchymal abnormalities

- **Functional Imaging:**
  - Can identify occult lesions, guide therapy for potential areas of reversible disease
  - PET/CT (FDG)- wave of the future?
    - Excellent sensitivity, but significant radiation
Functional Imaging of Sarcoidosis: FDG PET

Disseminated Sarcoidosis

Functional Imaging: Companion Patient #4

Image courtesy of Dr. Kevin Donohoe
Our Patient: Outcome

- Our patient was thought to have presumed sarcoidosis, with an ATYPICAL presentation (cavitary lung lesion and lung nodules, as well as more typical mediastinal and hilar lymphadenopathy), with probable superinfection.
- Chest tube was removed after several days and lung remained inflated.
- Patient was treated with empiric course of broad spectrum antibiotics for pulmonary superinfection.
- Outpatient follow-up was scheduled after completion of antibiotic course, with repeat CT to look for resolution of cavitary lesion and to better evaluate underlying lung parenchyma.
- The patient will likely start treatment for sarcoidosis at that time.
Summary I

- **Pneumothorax**
  - There are two types of pneumothorax: simple and tension.
  - Tension pneumothorax can lead to respiratory and hemodynamic compromise and is a surgical emergency requiring immediate chest tube placement.
  - When a patient presents with a primary spontaneous pneumothorax, consider further imaging to investigate the etiology.

- **Cavitary lung lesions**
  - There is a broad differential for cavitary lung lesions including infection, septic emboli, rheumatologic disease, and malignancy.
  - Sarcoidosis can present with a cavitary lung lesion in rare instances.
Summary II

- **Sarcoidosis**
  - Sarcoidosis is a systemic granulomatous disease, which most commonly affects the respiratory system.
  - Pulmonary sarcoidosis has four stages of disease.
  - Common pulmonary manifestations include hilar adenopathy and/or reticular opacities in early stages, and fibrosis in end stage disease.
  - However, there are numerous other less common manifestations of pulmonary sarcoidosis (i.e. cavitation).
  - Imaging plays an important role in diagnosing, staging, and prognosticating for pulmonary sarcoidosis.
Acknowledgements

- Mai-Lan Ho, MD
- Marc Camacho, MD
- Kevin Donohoe, MD
- Pamela Mok, MD
- Jennifer Son, MD
- Iva Petkovska, MD
- Gillian Lieberman, MD
- Larry Barbaras
- Emily Hanson
Bibliography


