Case of the Day – Chest

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Disclosures

• Relevant Financial Disclosures
  • None

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  • None
History

• 46 year-old male
• Progressive dyspnea on exertion
• New small amounts of hemoptysis for 2 months
Differential Diagnosis – Unilateral Pulmonary Interstitial Pattern

**ACUTE**
- Infection
- Edema
- Aspiration
- Resolving hemorrhage
- Airspace opacity superimposed on already abnormal lung (i.e. emphysema, fibrosis, etc.)

**CHRONIC**
- Recurrent aspiration/infection
- Radiation fibrosis
- Lymphangitic carcinomatosis
- Pulmonary venous obstruction
- Unilateral lung transplant
- Other rare
Differential Diagnosis

**NEOPLASTIC**
- Bronchogenic carcinoma
- Metastatic disease
- Lymphoma

**NON-NEOPLASTIC**
- Fibrosing mediastinitis
- IgG4-related disease
Subsequent Evaluation

- **18F-FDG PET/CT**
  - Mildly increased metabolic activity in subcarinal and para-aortic lymph nodes ($SUV_{max} = 3.6$)
  - Low level 18F-FDG activity in region of left hilar soft tissue thickening

- **EBUS-guided Biopsy**
  - Negative for malignancy

- **Serology**
  - *Histoplasma capsulatum* antibody positive
Answer

Fibrosing Mediastinitis
Fibrosing Mediastinitis

- Also referred to as mediastinal fibrosis or sclerosing mediastinitis

- Rare benign proliferation of fibrous tissue within the mediastinum
  - Pathologically, a spectrum from loose fibromyxoid tissue to dense collagen with a variable inflammatory cellular infiltrate
Fibrosing Mediastinitis – Etiology

- **Granulomatous infection**
  - *Histoplasma capsulatum*
  - Tuberculosis, actinomycosis, blastomycosis, aspergillosis...

- **Non-infectious granulomatous disease**
  - Sarcoidosis, silicosis, Behcet’s

- **Immunologic/Autoimmune**
  - Collagen vascular disease, IgG4-related disease, multisystem fibrosis

- **Malignancy**

- **Iatrogenic**
  - Methysergide, radiation

- **Trauma**

- **Idiopathic**
Fibrosing Mediastinitis – Presentation

• Variable age at presentation

• $\text{♂} = \text{♀}$

• Signs and symptoms related to compression or obstruction of mediastinal or hilar structures
  • Cough, dyspnea, recurrent infection, chest pain, hemoptysis, superior vena cava syndrome, dysphagia, hoarseness, diaphragmatic paralysis, Horner syndrome, chylothorax,…

• Constitutional symptoms
  • Fever, weight loss, malaise
Fibrosing Mediastinitis – Imaging

- **Chest Radiographs**
  - mediastinal widening
  - distortion of mediastinal lines and stripes
  - hilar enlargement
  - +/- calcification

- **Sequelae:**
  - airway narrowing, post-obstructive atelectasis/consolidation
  - pulmonary venous hypertension, pulmonary edema
  - vascular asymmetry, pulmonary infarcts
Fibrosing Mediastinitis – Imaging

- **CT**
  - infiltrative mediastinal/hilar soft tissue thickening or mass
  - focal (82%) or diffuse (18%)
  - calcification (60-90%)
  - variable enhancement

- **Sequelae**
  - compression/obstruction of central airways, central vessels, esophagus
  - atelectasis, consolidation, edema, infarcts
Fibrosing Mediastinitis – Imaging

• MRI
  • typically intermediate T1 and heterogeneous T2 signal intensity
  • often regions of T2 hypointensity
  • inferior to CT for detection of calcification
  • heterogeneous enhancement post-gadolinium

• Sequelae
  • as demonstrated on CT
Fibrosing Mediastinitis – Imaging

• **Nuclear Medicine**
  - **18-FDG PET**
    - variable metabolic activity within and between lesions

• **Ventilation/Perfusion**
  - perfusion defects reflecting pulmonary arterial or venous obstruction
  - lobar/segmental ventilation defects or air trapping reflecting airway involvement
Summary

• Fibrosing mediastinitis should be considered in the setting of localized mediastinal soft tissue thickening associated with vascular and/or airway compromise.

• Accompanying calcification further suggests the diagnosis.

• In the correct clinical setting, such as positive *Histoplasma capsulatum* serology and no known malignancy, CT findings may obviate the need for biopsy.
Questions?

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Review

At the conclusion of this session, participants should be able to:

1. Provide a differential diagnosis for a unilateral pulmonary interstitial pattern. (CanMEDS Roles: Medial Expert)
2. Describe the typical imaging features and differential diagnosis of fibrosing mediastinitis. (CanMEDS Roles: Medical Expert)
3. List at least four causes of fibrosing mediastinitis. (CanMEDS Roles: Medical Expert)
References

