38-year-old female presents with cough & shortness of breath
Ms. MG is a 38-year-old female with a history of childhood asthma who presents with shortness of breath and a dry cough. Her childhood asthma was mild, and she was hospitalized once for an exacerbation at age 4. She notes improvement in asthma symptoms since adulthood. Additionally, she has a significant allergy history (most recently tested for 75 allergens and positive to all).

She reports development of a non-productive cough with intermittent episodes of labored breathing without clear precipitants. Her cough keeps her up at night now. She also reports generalized fatigue over the past several months.

Never smoker.
Diagnostic work-up

- Chest x-ray
- Non-contrasted high resolution CT scan of chest
CXR – May 2016

- Chest X-ray was reportedly “normal” based on record from outside hospital.
- Imaging modality sequence depends on suspicion for underlying disease process and severity.
Non-contrasted CT scan shows cavitary lesion in left inferior lobe with surrounding fibrosis and scarring.

Patchy nodular infiltrates with dilated airway.
Dilated airways seen in the periphery of the left inferior lobe. Airways are wider than the associated vessels, consistent with bronchiectasis.
Differential is Broad!

- Imaging suggested focal fibro-cavitary left lower lobar bronchiectasis.

- Differential diagnosis broad:
  - Aspiration
  - Primary mycobacterial disease
  - Cystic fibrosis
  - Allergic bronchopulmonary aspergillosis (ABPA)
  - Post-infectious
  - Immunodeficiency
  - A1AT deficiency
  - COPD
  - Asthma
  - Inflammatory bowel disease
  - Ciliary dysfunction
  - Connective tissue disease
  - Idiopathic
Further Diagnostic Work-up

- Test for Alpha-1 anti-trypsin deficiency
  - A1AT level returned <30
  - Determined to have PiZZ genotype (ZZ is most severe form)
- Sputum culture
  - Grew Mycobacterium avium intracellulare (MAC)

Repeat high resolution CT scan of chest after several months of directed MAI treatment and airway clearance…
Significant improvement c/w prior

Inferior lobe predominant bronchiectasis, and multifocal nodular/tree-in-bud opacities, predominately within the left lower lobe and lingula.

No manifestations of emphysema from patient's known alpha-1-antitrypsin deficiency.
Significant improvement c/w prior

Inferior lobe predominant bronchiectasis, and multifocal nodular/tree-in-bud opacities, predominately within the left lower lobe and lingula.

No manifestations of emphysema from patient's known alpha-1-antitrypsin deficiency.
- Treated with **rifampin, ethambutol, and clarithromycin**.
- Given her imaging findings of bronchiectasis with **cavitary disease** in the context of alpha 1 anti-trypsin, she was also treated with **IV amikacin** therapy for 2 months.
- Uses daily hypertonic saline and albuterol for **airway clearance**

Symptomatically, she has done well. Cough has resolved. She is being considered for A1AT replacement therapy.
Discussion: Bronchiectasis

- Localized, irreversible destruction of cartilage-containing airway walls with resultant dilatation
- Characterized by permanent dilation, retention of mucus, and impaired ciliary clearance
- Most common causes: idiopathic (most common), post infectious (pneumonia, TB), primary or secondary immunodeficiencies, CF, ciliary dysfunction, ABPA, and connective tissue disease


Podcast by Dr. Peadar Noone, Bronchiectasis Expert
Imaging: Bronchiectasis

- **Gold standard = High res CT**
- Bronchial dilation is most important CT finding (usually defined as internal airway diameter > adjacent pulmonary artery = “signet ring sign”)
- Lack of airway tapering >2 cm distal to point of bifurcation
- Airway visibility within 1 cm of the costal pleura of fissures
References


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