Pulmonology Case Presentation

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UNC SCHOOL OF MEDICINE
Radiology

38-year-old female presents with cough & shortness of breath

Patient history

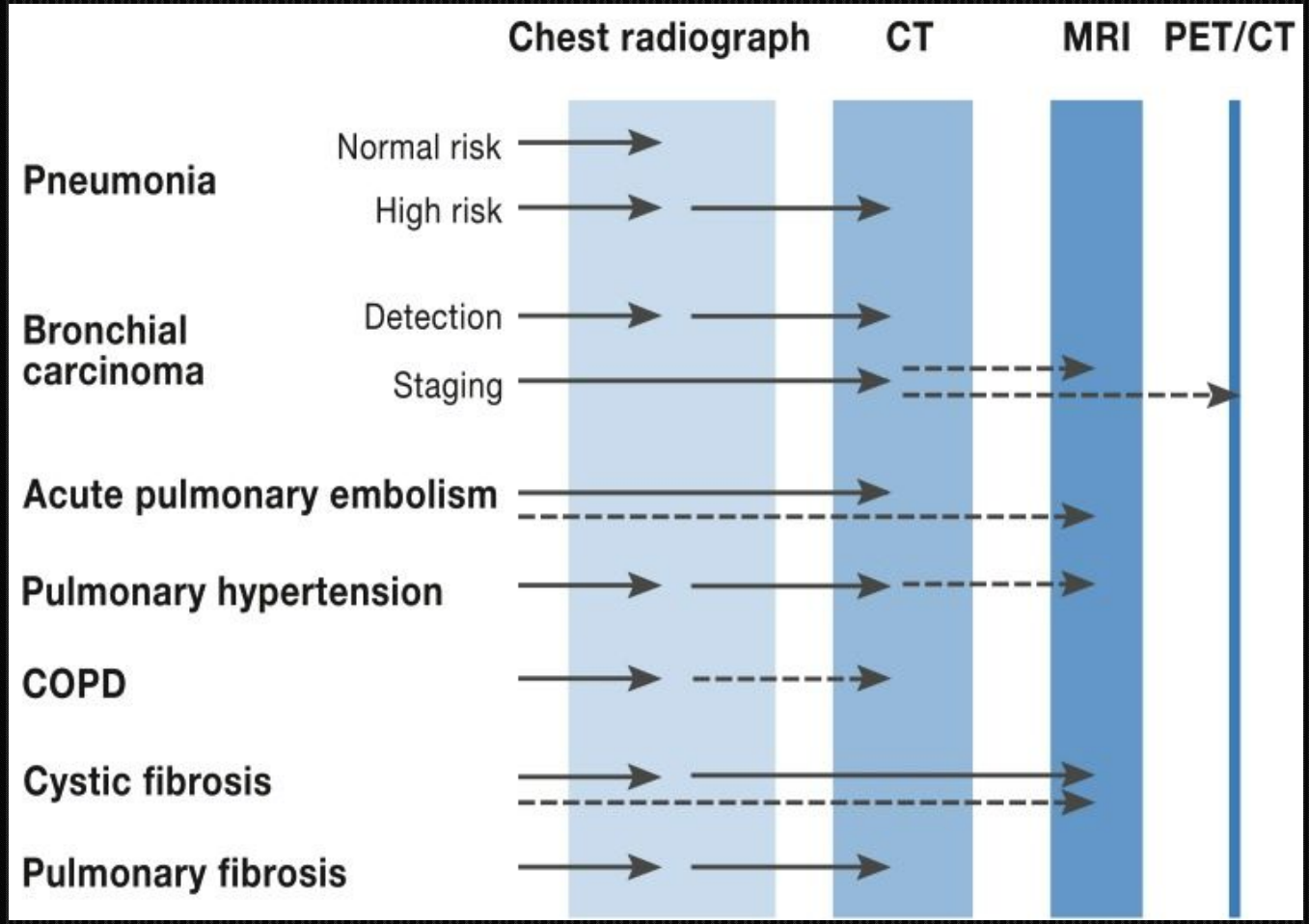
- Ms. MG is a 38-year-old female with a history of childhood asthma who presents with shortness of breath and a dry cough. Her childhood asthma was mild, and she was hospitalized once for an exacerbation at age 4. She notes improvement in asthma symptoms since adulthood. Additionally, she has a significant 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

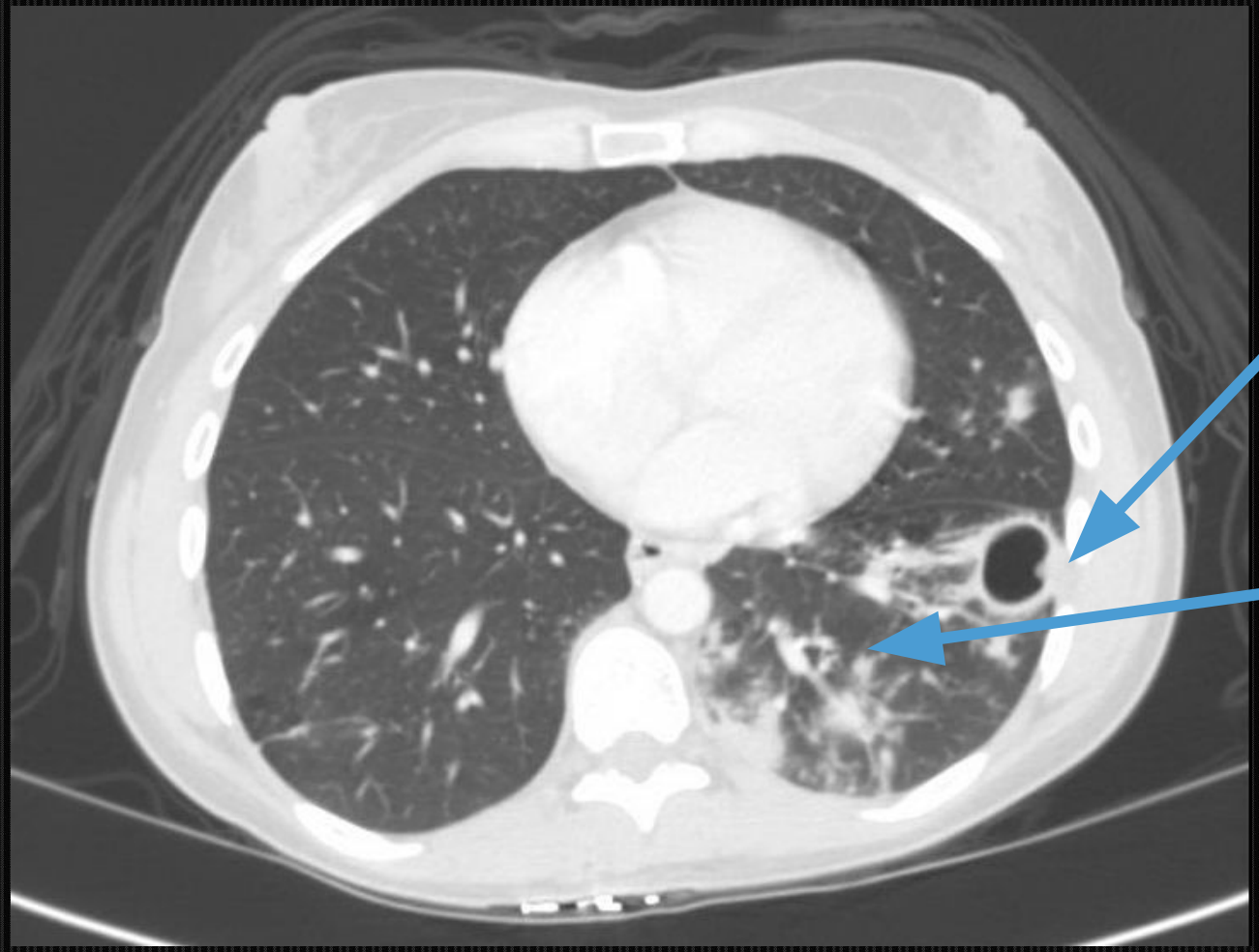
- Chest x-ray
- Non-contrasted high resolution CT scan of chest

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



Chest CT - June 2016



Non-contrasted CT scan shows cavitary lesion in left inferior lobe with surrounding fibrosis and scarring.

Patchy nodular infiltrates with dilated airway.

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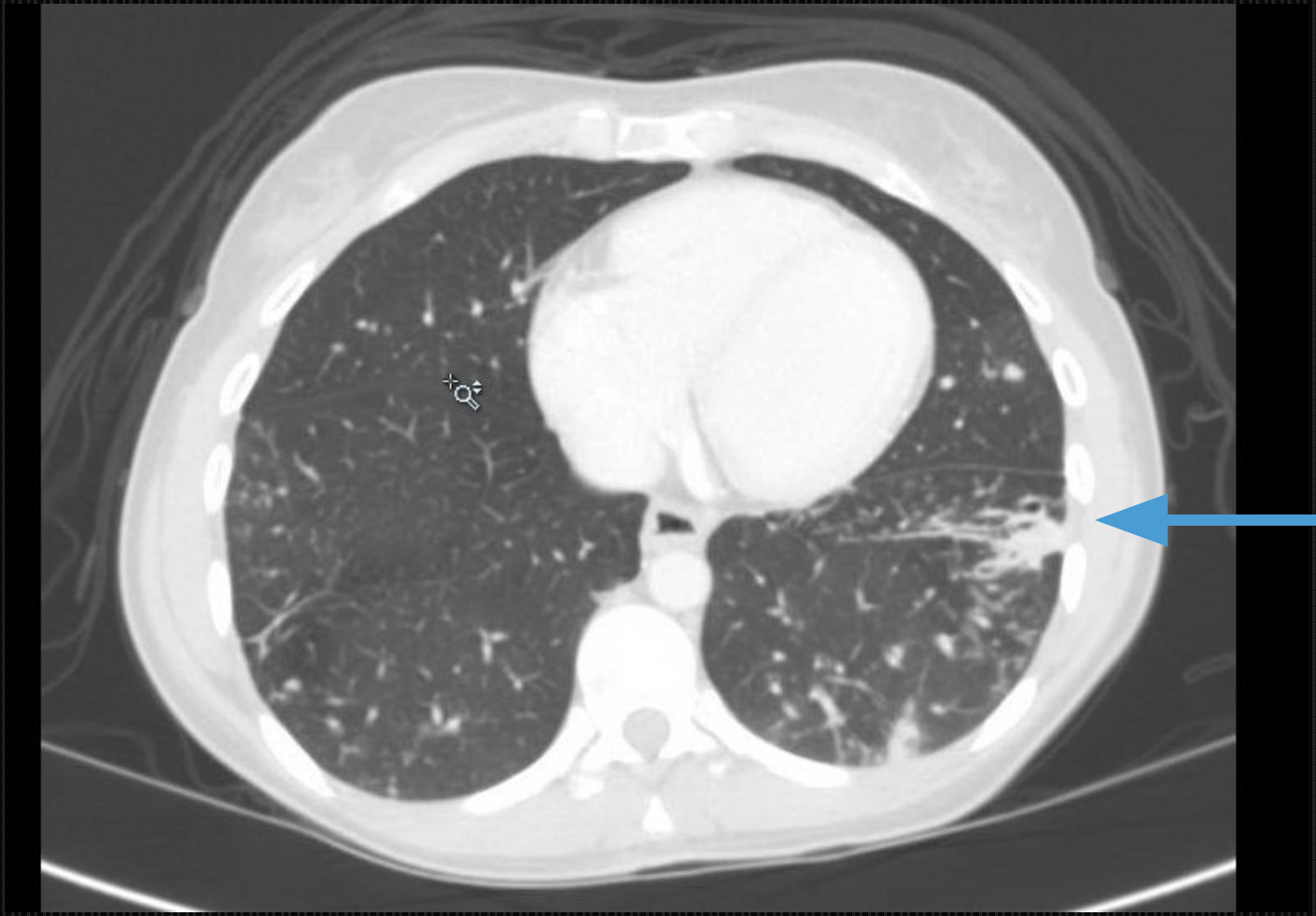
Chest CT - June 2016



Dilated airways seen in the periphery of the left inferior lobe. Airways are wider than the associated vessels, consistent with bronchiectasis.

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Differential is Broad!

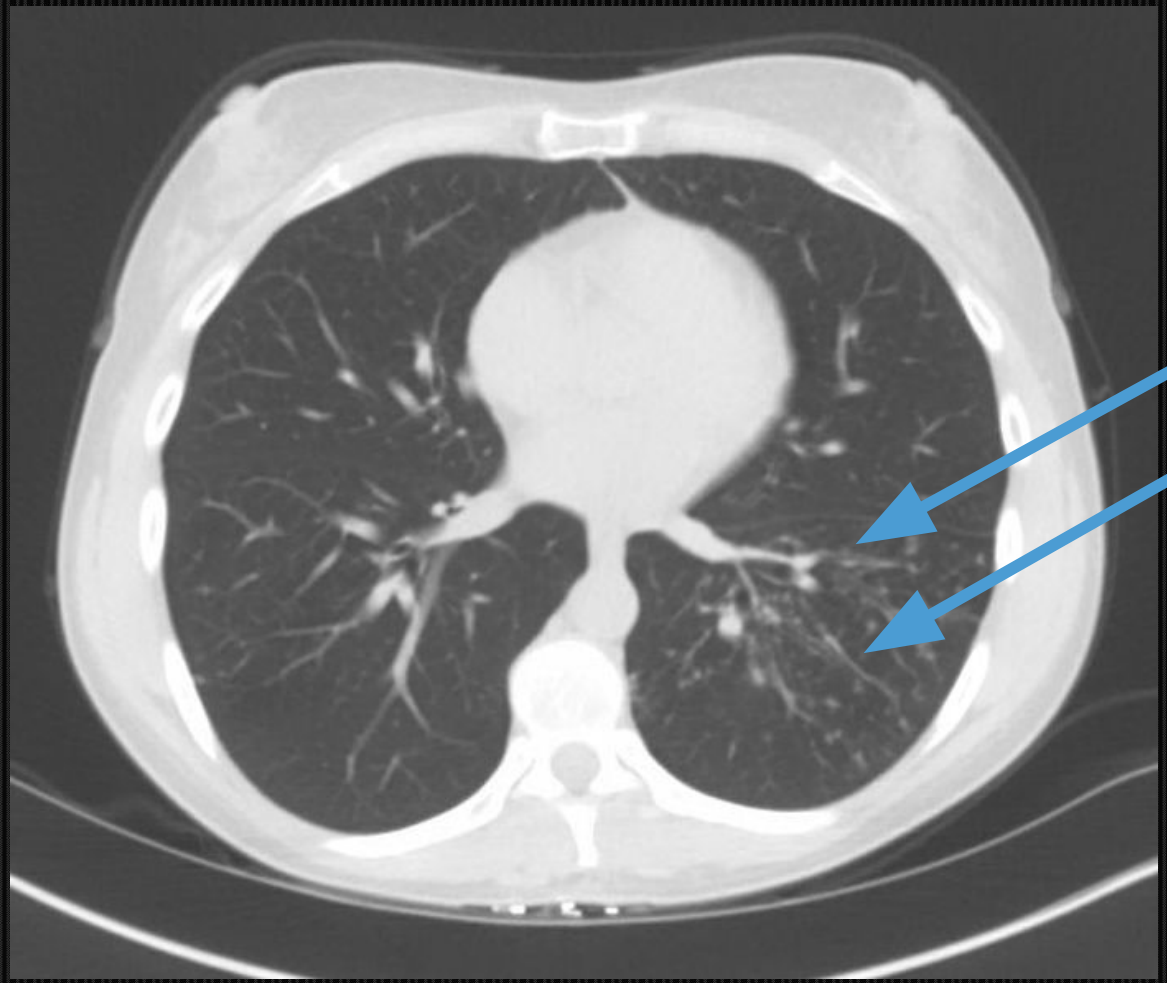
- Imaging suggested focal fibrocavitary left lower lobar **bronchiectasis**.
- Differential diagnosis broad:
  - Aspiration
  - Primary mycobacterial disease
  - Cystic fibrosis
  - Allergic bronchopulmonary aspergillosis (ABPA)
  - Post-infectious
  - Immunodeficiency
  - A1AT deficiency
  - COPD
  - 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 …

CT - October 2016

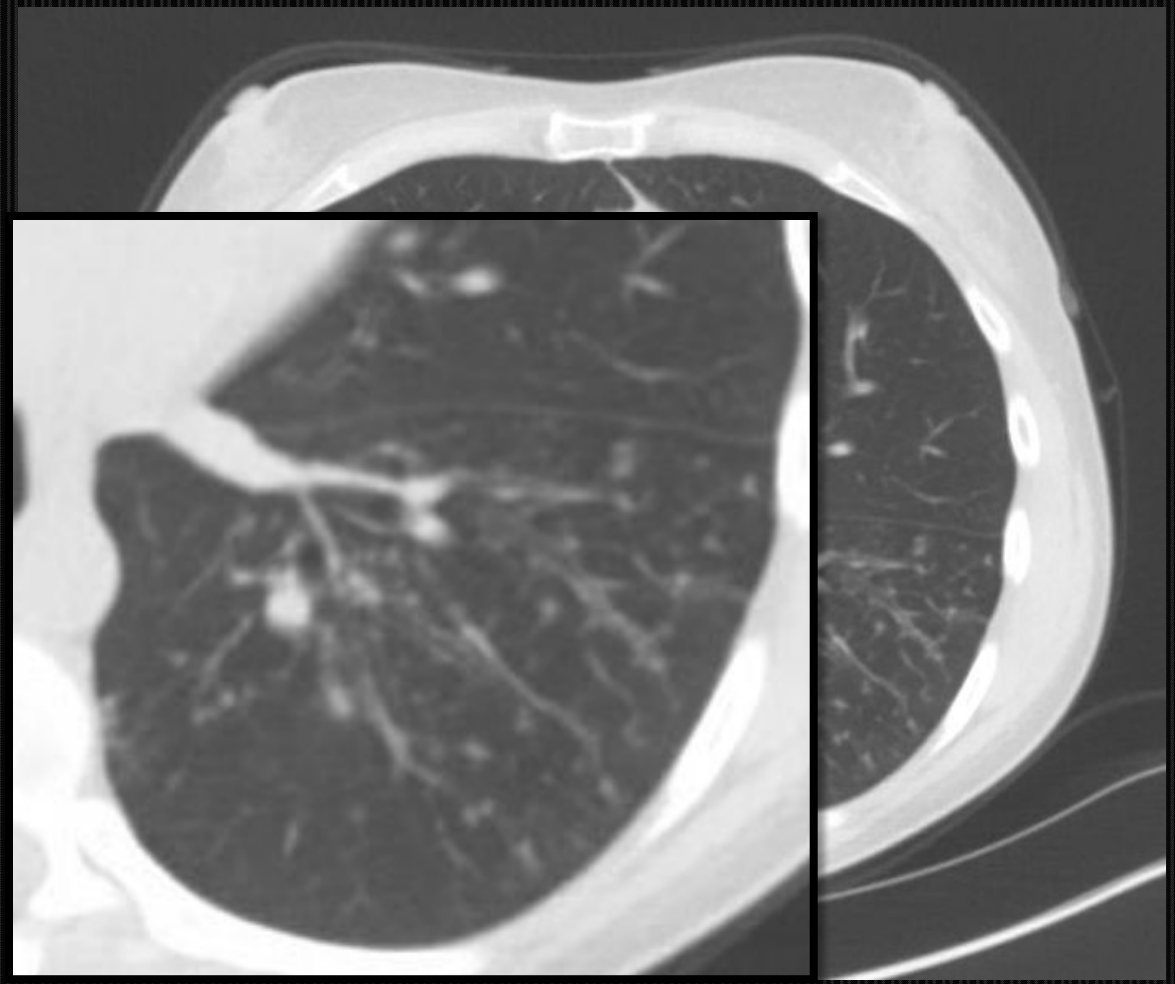


Significant improvement c/w prior

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.

CT - October 2016



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Patient treatment or outcome

- 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

https://www.med.unc.edu/medicine/news/chairs-corner/unc-bronchiectasis.mp3

Podcast by Dr. Peadar Noone, Bronchiectasis Expert

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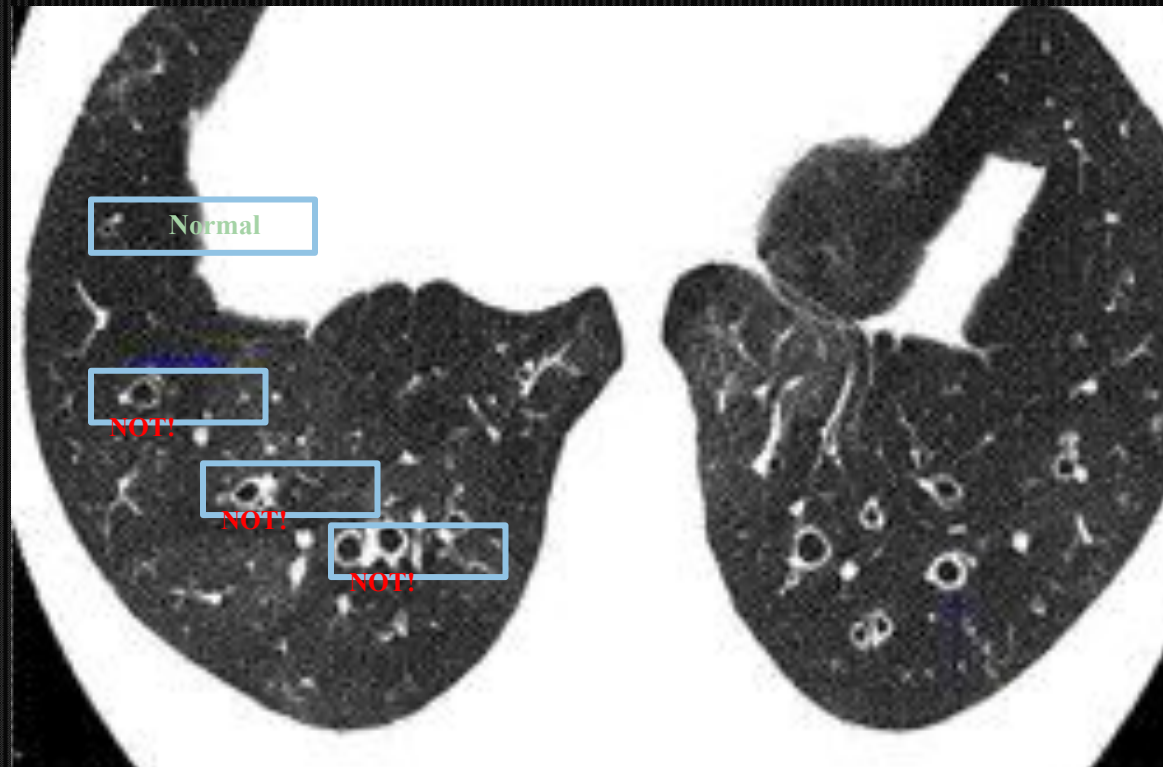
Imaging: Bronchiectasis

- **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



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