Bronchiectasis Case Presentation

California Thoracic Society

January, 2018

Bronchiectasis Case Presentation

- **Presents in September, 2017 with progressive dyspnea on exertion**
  Currently walks 2 blocks. Could walk 1 mile a year earlier

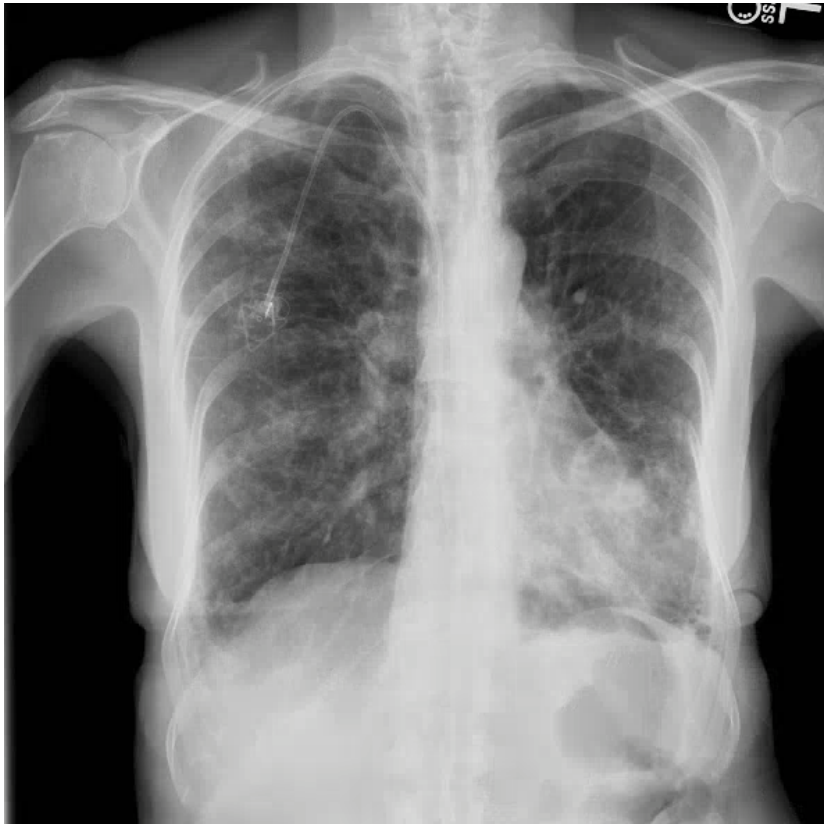
- **Cough, chronic sputum production.**

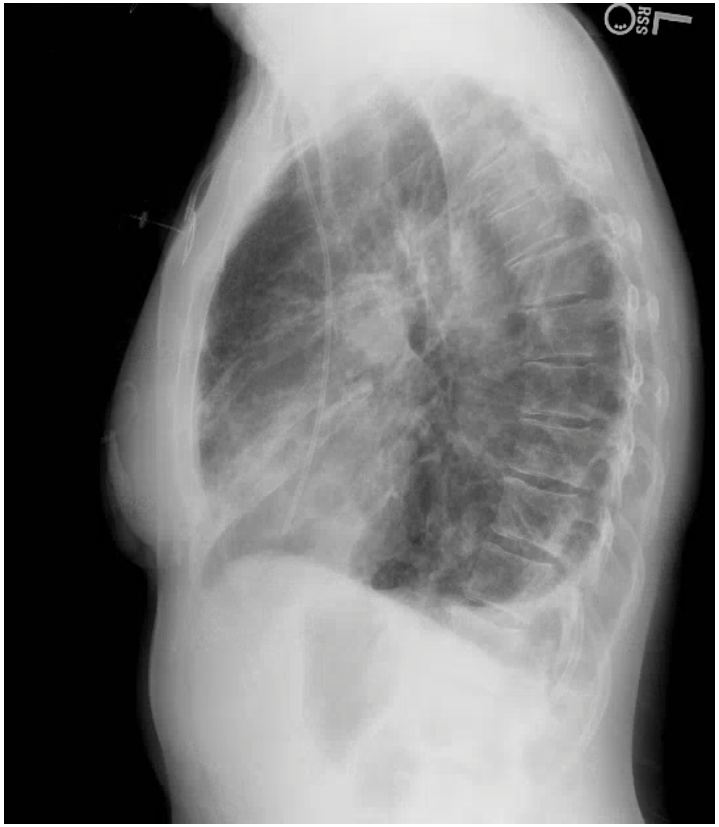
- **PMH:GERD, Hiatal hernia, Gastroparesis, LV diastolic dysfunction**

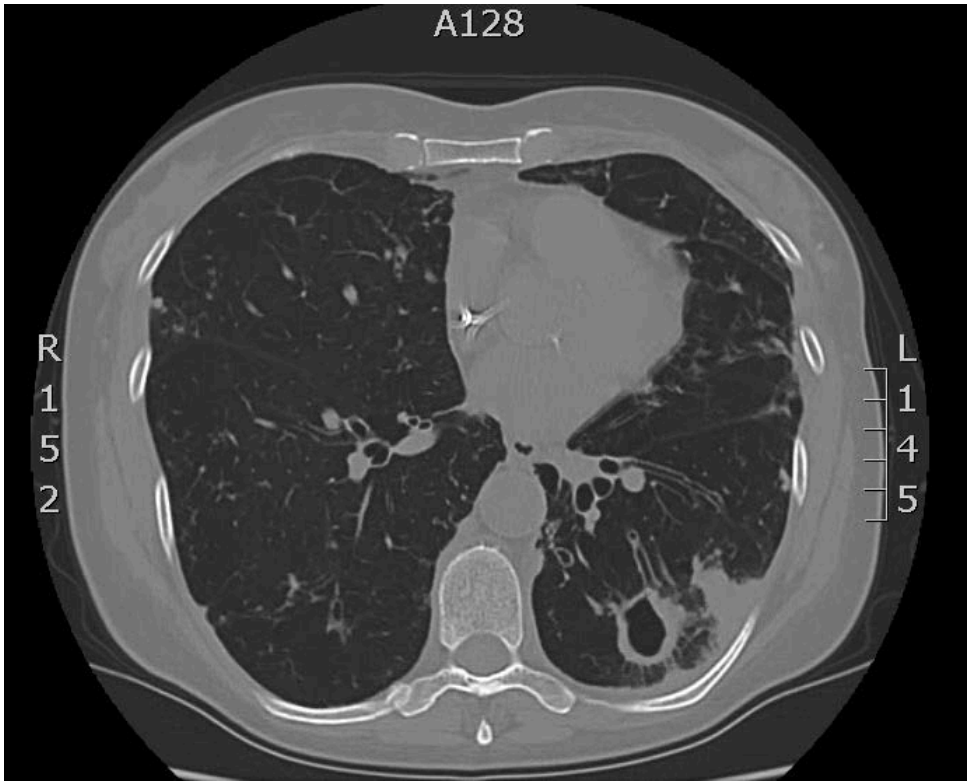
- **PMH:MAC Rx a) 1996 x18 months**
  b) 2002 x 13 weeks
  c) 2004 x 6 months
  d) 2015 x 8 months
  e) 2017 6 months to current (fevers, weight loss, cough and dyspnea)

- **Pancreatic Insufficient. Failure to thrive as an infant.**

- **Chronic Sinusitis. Recurrent nasal polypectomy**



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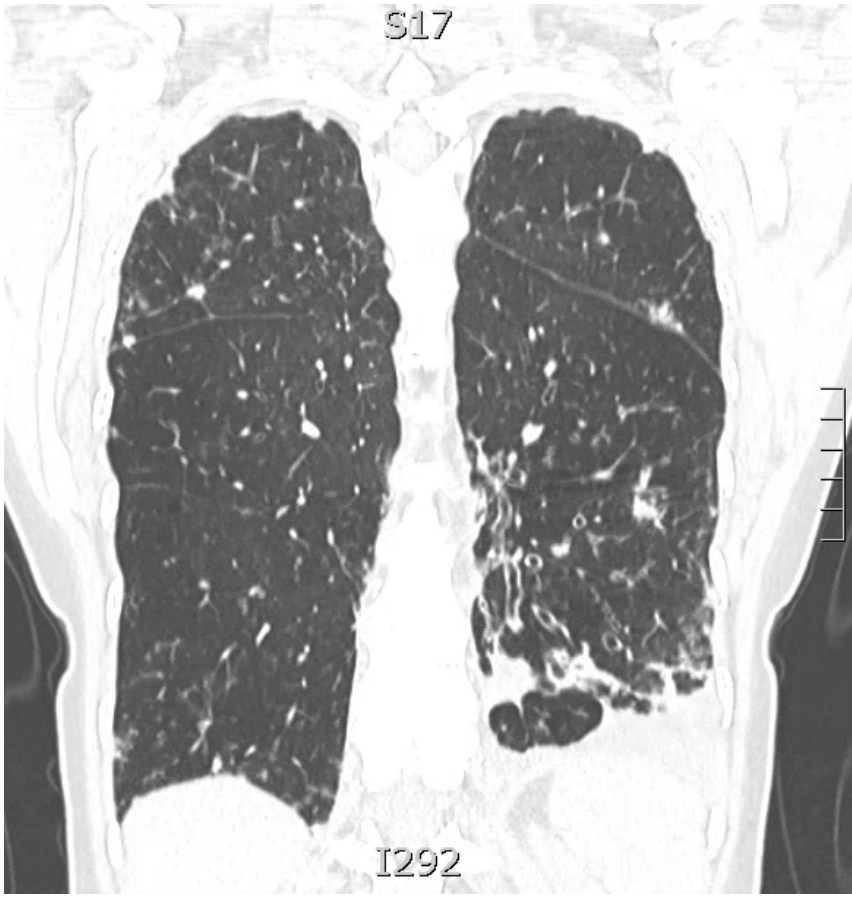
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- **Physical Exam**
  - **-Thin. BMI =18.5**
  - **-Mild maxillary Tenderness**
  - **-Chest: Decreased breath sounds with scattered rhonchi. Rare exp wheezing**
  - **- No clubbing**

- **Laboratory Data**
  - **- CBC: Normal**
  - **-ESR : 110**
  - **-CRP: 5.2**
  - **-Chem20: Normal**
  - **-Ig and IgG subclasses: Normal**
  - **-IgE: 4**

- **Microbiology Data**
  - **AFB: MAC/MAI**
  - **Fungal: Aspergillus intermittently**
  - **Routine: *Pseudomonas aeruginosa***
  - ***Stenotrophomonas maltophilia***

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- **Relevant CF data**
  - **-Sweat Chloride: 25meq/ml**
  - **-Borderline nasal potential difference**
  - **-CFTR genetics**
    - **a) M470V/M470V**
    - **b) IVS 5T allele**
  - **-Fecal pancreatic elastase was 52 ug/g (nl > 200ug/g)**
  - **-Vitamins E and A were normal**
  - **-Vitamin D was low**

- **Relevant PCD data**
  - **-EM cilia showed normal outer and inner dynein arms and questionable central apparatus.**
  - **-Single DNAH5 variation**
    - **p.F4392C: uncertain pathogenicity**
  - **-Single DNAH11 variation**
    - **p.D892V: uncertain pathogencity**

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Leading considerations for etiology of bronchiectasis

- **Atypical Cystic Fibrosis**
  Strong clinical phenotype but lacks convincing laboratory confirmation
- **Chronic NTM (MAC/MAI) airway infection**
- **Chronic aspiration**
- **Atypical PCD**
  PCD is recessive.
  Unclear if the carrier state is clearly normal

- **“Multiple hit” concept**