Bronchiectasis

Sarah Tapyrik, MD, FCCP
Assistant Professor - Clinical
Medical Director of Respiratory Therapy
Department of Internal Medicine
Division of Pulmonary, Critical Care and Sleep Medicine
The Ohio State University Wexner Medical Center

Bronchiectasis - Outline

- Definition & Radiology Review
- Epidemiology & Pathophysiology
- Etiologies
- Diagnosis & Work-up
- Treatment
- Case review
Case

- 64 yo man with 5 years of chronic cough
- Dry cough with minimal mucous production
- Antibiotics at least 4 times/year
- Sinus disease with improvement s/p surgery

- Never smoker
- Grew up on a farm – livestock and chickens
- Works as an engineer
- PCP sent him for a CT scan

And the CT Scan is Read As:

- BRONCHIECTASIS

- What IS this anyway?
Radiology

- What exactly do we see on imaging?
  - Bronchial diameter exceeding that of the adjacent pulmonary artery
  - Lack of normal tapering of terminal bronchioles in the lung periphery

Bronchiectasis

- Imaging: abnormal and permanent dilatation of the bronchi
Bronchiectasis

Normal

Image: Yale Rosen CC BY-SA 2.0
https://www.flickr.com/photos/pulmonary_pathology/3677946871
Bronchiectasis

- Clinical syndrome: cough, sputum production and recurrent bronchial infection
  - +
- Radiological findings of dilated airways
- Occurs in multiple pathologic processes

Eur Respir J 2017; 50: 1700629
UpToDate Feb 2021. Clinical manifestations and diagnosis of bronchiectasis in adults
Epidemiology

- Up to 500,000 US adults have bronchiectasis
- Prevalence increases with age, highest rates in adults >60 years old
- More common in women
- Extensive use of healthcare resources

Pathophysiology

- Two major factors
  1. Infectious insult
  2. Impaired drainage, airway obstruction or defect in host defense

- Airway neutrophils, cytokines and other immune responses cause abnormal dilatation and destruction of airways (bronchi and bronchiole walls)
# Etiologies

**Acquired bronchial obstruction**
- Foreign body aspiration
- Tumors
- Hilar adenopathy
- COPD
- Mucoid impaction
- Other

**Congenital anatomic defects**
- Tracheobronchial
- Vascular
- Lymphatic

**Immunodeficiency states**
- IgG deficiency
- IgA deficiency
- Leukocyte dysfunction
- Other rare humoral immunity immunodeficiencies

**Abnormal secretion clearance**
- Ciliary defects
- Cystic fibrosis
- Young’s syndrome

---

# Etiologies (continued)

**Infections**
- Childhood infections
- Bacterial infections
- Viral infections
- Other infections

**Miscellaneous disorders**
- Alpha-1-antitrypsin deficiency
- Recurrent aspiration pneumonia
- Rheumatic disease
- Inflammatory bowel disease
- Toxic fume & dust inhalation
- Chronic rejection after solid organ transplantation
Etiologies

- Multiple etiologies can cause or contribute to pathophysiologic process
- Cystic Fibrosis (CF) Bronchiectasis
  - Recurrent and chronic airway infections
  - Most recognized cause
- Non-CF Bronchiectasis
  - All etiologies other than CF

Non CF Bronchiectasis

- Airway Obstruction
- Defective host defense - common variable immunodeficiency
- Rheumatic disease
- Primary Ciliary Dyskinesia
- Infections
- Allergic Bronchopulmonary Aspergillosis (ABPA)
- Fibrosing Lung Diseases
- Aspiration
- Congenital

UpToDate Feb 2021. Clinical manifestations and diagnosis of bronchiectasis in adult
Airway Obstruction

- Foreign body aspiration
- Intraluminal obstruction lesion (carcinoid)
- Extra luminal compression (mass or lymph nodes)
- Focal bronchiectasis at sight of obstruction

Defective host defense

- Ciliary defects, prolonged immunosuppression, hypogammaglobulinemia (IPH), CVID
- Bronchial wall injury from repeated infections
- Recurrent sinus and respiratory infections
- ? IgG subclass deficiency
Rheumatic & systemic disease

- RA and Sjogrens syndrome can be complicated by bronchiectasis
- Inflammatory bowl disease (UC > Crohn’s)
- Bronchiectasis can occur prior to rheumatic symptoms/diagnosis
- Mechanism not known
- RA + bronchiectasis (and COPD) has higher mortality than other bronchiectasis associations

UpToDate Feb 2021. Clinical manifestations and diagnosis of bronchiectasis in adult

Primary Ciliary Dyskinesia

- Immotile-cilia syndrome with defect in airway cilia
- Autosomal recessive with 30+ variants
- Recurrent infections – upper and lower respiratory tracts
- Bronchiectasis middle lobe and lingula
- Nasal nitric oxide analysis (low level is consistent with PCD)
- Extended genetic testing

UpToDate Feb 2021. Overview of Primary Ciliary Dyskinesia
UpToDate Feb 2021. Clinical manifestations and diagnosis of bronchiectasis in adult
Infections

- Multiple infections associated with bronchiectasis
  - Bacterial
  - Atypical bacteria (mycoplasma, chlamydia and legionella species)
  - Viral
  - Mycobacterial (TB and NTM)
  - Nocardia

- Childhood and recurrent infections

Mycobacterial Infections

- Sequela of virulent infections
  - Direct tissue injury
  - Obstruction from enlarged lymph nodes

- Bronchiectasis is both a risk and consequence
Infections – NTM: chicken or egg?

- Underlying bronchiectasis may have NTM infection or colonization
- NTM infection (MAC and M abscessus) can cause bronchiectasis
- Fibronodular bronchiectasis caused by MAC usually in women >60 in RML and lingula.
  - Peribronchial inflammation and thickening that leads to bronchiectasis

Allergic Bronchopulmonary Aspergillosis (ABPA)

- Complex hypersensitivity reaction in response to colonization of the airways with *Aspergillus fumigatus*
- Occurs in patients asthma or cystic fibrosis (CF)
- CT with peripheral and central airway bronchiectasis

- Blood eosinophilia
- Elevated plasma IgE
- Precipitating specific antibodies Aspergillus
Fibrosing Lung Diseases

- Sarcoidosis – upper/central airways
- Idiopathic Pulmonary Fibrosis (IPF) – lower airways
- Sequela of acute respiratory distress syndrome (ARDS) – middle lobe and lingula

Other contributors

- Vitamin D Deficiency
  - Observational study of 402 patients with bronchiectasis
  - 50% deficient and 43% insufficient
  - Deficient patients with more pseudomonas colonization, more exacerbations and worse symptoms

- Cigarette smoking → COPD
  - Causal role not clear
  - Repeated infections/exacerbations can accelerate disease
Other contributors

- Chronic aspiration
  - Airway destruction from acidic GI contents
  - Lower lobe airways

- Alpha-1 Antitrypsin (A1AT) deficiency – most associated with premature panlobular emphysema
  - Abnormal elastase
  - Lower lobe predominance

Congenital syndromes

- Williams-Campbell Syndrome: rare cartilage deficiency of the mid-order bronchi.
- Swyer-James Syndrome: post infectious bronchiolitis obliterans
  - smaller lucent lung usually accompanied by diffuse bronchiectasis
Work-up

- Imaging → CT Chest (preferably thin sections)
  - CXR insensitive
- Bronchoscopy
- CBC with differential, Immunoglobulins, autoimmune, sweat chloride, CFTR gene mutation, nasal nitric oxide analysis, PCD gene testing, A1AT, RAST (aspergillus testing)
- Cultures – AFB, fungal
- PFTs (often obstructive impairment)

UpToDate Oct 2020. Bronchiectasis in adults: Treatment of acute exacerbations and advanced disease

Bronchiectasis - Exacerbations

- Deterioration of 3 or more symptoms for ≥48 hour
  - Cough
  - Sputum volume and/or consistency
  - Sputum purulence
  - Breathlessness and/or exercise intolerance
  - Fatigue and/or malaise
  - Hemoptyisis

- Mucous tends to more tenacious and concentrated as compared to healthy controls and other conditions

UpToDate Oct 2020. Bronchiectasis in adults: Treatment of acute exacerbations and advanced disease
Guidelines - 2017

European Respiratory Society guidelines for the management of adult bronchiectasis

Eva Polverino, Pieter C. Goeminne, Melissa J. McDonnell, Stefano Aliberti, Sara E. Marshall, Michael R. Loebinger, Marlene Murris, Rafael Cantón, Antoni Torres, Katerina Dimakou, Anthony De Soyza, Adam T. Hill, Charles S. Haworth, Montserrat Vendrell, Felix C. Ringhausen, Dragan Subotic, Robert Wilson, Jordi Vilaró, Bjorn Stallberg, Tobias Wette, Gernot Rohde, Francesco Blasi, Stuart Elborn, Marta Almagro, Alan Timothy, Thomas Ruddy, Thomy Tonia, David Rigau, and James D. Chalmers

Treatment - Exacerbations

- Antibiotics – choice of agent based on cultures
- 14 days of treatment
- Long term antibiotics (> 3 months) in adults with 3 or exacerbations/year
  - Inhaled antibiotics with chronic *P. aeruginosa* infection
  - Macrolide therapy
- Eradication therapy with new isolation of *P. aeruginosa*
  - Combination of oral, IV and/or inhaled therapies

Eur Respir J 2017; 50: 1700629
Treatments - inhalers

- Inhaled corticosteroids and long-acting bronchodilators – not recommended for routine use
- Continued in patients with co-morbidities of asthma and/or COPD
- Trial of short or long acting bronchodilators in certain patients (significant breathlessness)

Eur Respir J 2017; 50: 1700629

Treatment – airway clearance

- Mucous clearance therapies
  - Directed cough
  - Exercise
  - Forced expiration
  - Chest physical therapy – postural drainage, hand or mechanical chest clapping
  - Oscillation vest
  - Vibratory Positive Expiratory Pressure (PEP)

UpToDate Oct 2020. Bronchiectasis in adults: Treatment of acute exacerbations and advanced disease
In practice....

- Steroids – most often inhaled
- Inhaled and oral antibiotics
- Clearance aides (flutter/acapella and chest vest)
- Nebulized hypertonic saline

Case 1

- Recurrent cough and sinus drainage
- Improves with oral steroids
- IgG, IgM and IgA low end of normal
Case 2

- 37 yo old with 4 years of chronic cough
- BAL with MAC
- Chronic sinus disease

Case 3

- 87 yo old with mild, intermittent, chronic cough
- Minimal antibiotic, bronchodilator or steroid use
Summary – Bronchiectasis

- Irreversible regional or diffuse bronchial dilatation
- Repeated pattern of airway infection, inflammation, and injury
- Multiple causes, including congenital diseases, infection, obstructing lesions, immunodeficiency, aspiration
- History + lab work + imaging findings can help with diagnosis
- Treatment consists of antibiotics, steroids (inhaled, oral), bronchodilators, mucous clearance assistance as well as treatment of underlying conditions

Eur Respir J 2017; 50: 1700629

References

- UpToDate
  - Clinical manifestations and diagnosis of bronchiectasis in adults
  - Clinical manifestations and diagnosis of allergic bronchopulmonary aspergillosis
  - Primary Ciliary Dyskinesia (immotile-cilia syndrome)
  - Overview of nontuberculous mycobacterial infections
