Bronchiectasis

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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?

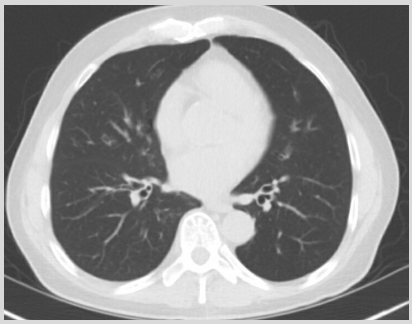
Radiology

- 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

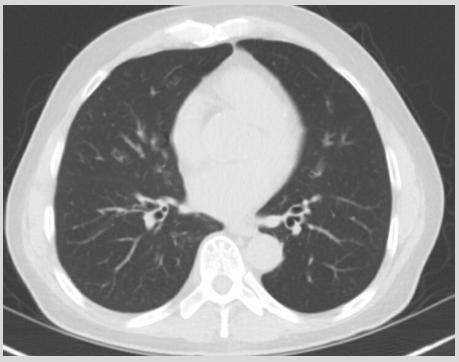
RadioGraphics 2015; 35:1011–1030

Bronchiectasis

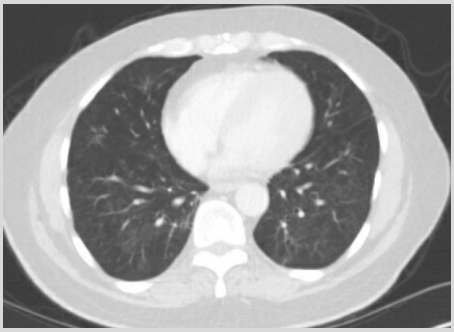
- Imaging: abnormal and permanent dilatation of the bronchi



Bronchiectasis



Normal



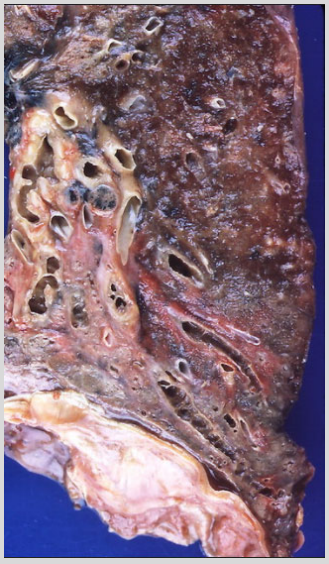
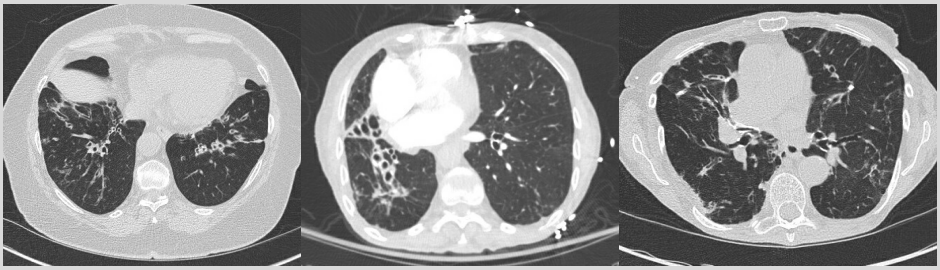


Image: Yale Rosen CC BY-SA 2.0
https://www.flickr.com/photos/pulmonary_pathology/3677946871



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

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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)

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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

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Airway Obstruction

- Foreign body aspiration
- Intraluminal obstruction lesion (carcinoid)
- Extra luminal compression (mass or lymph nodes)

- Focal bronchiectasis at sight of obstruction

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Defective host defense

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

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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

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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

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Infections

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

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Mycobacterial Infections

- Sequela of virulent infections
  - Direct tissue injury
  - Obstruction from enlarged lymph nodes
- Bronchiectasis is both a risk and consequence

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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

UpToDate Oct 2020. Overview of NTM infections

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

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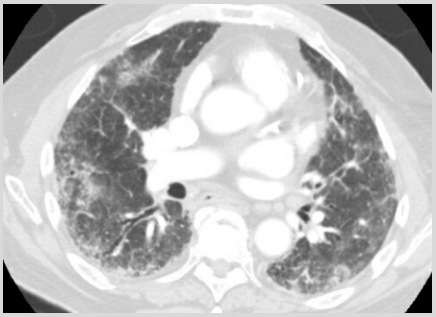
Fibrosing Lung Diseases

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



RadioGraphics 2015; 35:1011–1030

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

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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

RadioGraphics 2015; 35:1011–1030

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

RadioGraphics 2015; 35:1011–1030

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)

RadioGraphics 2015; 35:1011–1030

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
  - Hemoptysis
- 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[1], Pieter C. Goeminne[2,3], Melissa J. McDonnell[4,5,6], Stefano Aliberti[7], Sara E. Marshall[8], Michael R. Loebinger[9], Marlene Murris[10], Rafael Cantón[11], Antoni Torres[12], Katerina Dimakou[13], Anthony De Soyza[14,15], Adam T. Hill[16], Charles S. Haworth[17], Montserrat Vendrell[18], Felix C. Ringshausen[19], Dragan Subotic[20], Robert Wilson[9], Jordi Vilaró[21], Bjorn Stallberg[22], Tobias Welte[19], Gernot Rohde[23], Francesco Blasi[7], Stuart Elborn[9,24], Marta Almagro[25], Alan Timothy[25], Thomas Ruddy[25], Thomy Tonia[26], David Rigau[27] and James D. Chalmers[28]

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)

VibraPEP by Curaplex
Medicalequipment4sale.com

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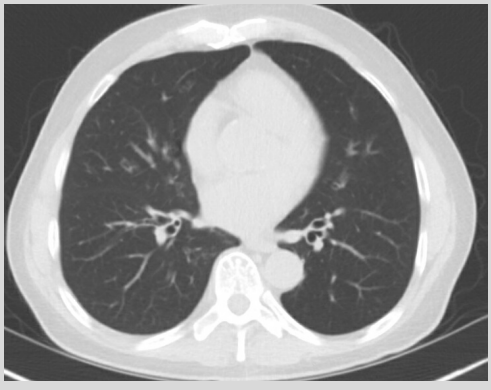
Acapella Vibratory PEP.
DMESupplyUSA

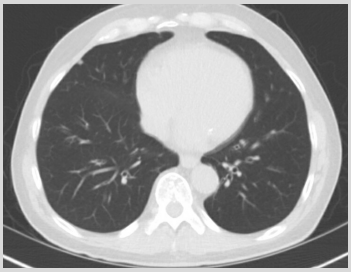
In practice….

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

Case 1

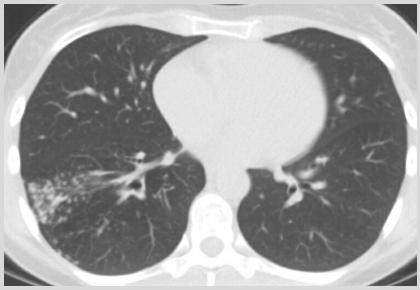
- Recurrent cough and sinus drainage
- Improves with oral steroids
- IgG, IgM and IgA low end of normal





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
- Multiples 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

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