Bronchiectasis – Clinical Quick Talk

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Definition:

- Bronchiectasis – chronic lung disease characterized by either focal or diffuse dilation of the airways (12)

Pathophysiology:

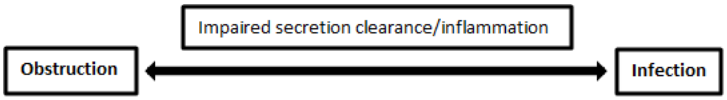

Figure 1: Two interrelated processes involving infection and obstruction lead to inflammation and damage of bronchial walls. This further leads to impaired secretion and infection clearance, which perpetuates the cycle. (13)

Etiology:

Etiologies	Examples	Important notes
Obstruction	• Tumor • Lymph node • Foreign body	Focal pattern
Infection	• Bacterial (staph, pseudomonas, etc.) • Mycobacterial (tuberculous and nontuberculous) • Viral pneumonias (flu, covid) • Allergic bronchopulmonary aspergillosis (ABPA)	ABPA seen in asthma – peripheral and central airway bronchiectasis
Immunodeficiency	• Common Variable Immunodeficiency (CVID) • HIV	
Autoimmune	• Rheumatoid arthritis • Sjogren's syndrome • IBD	Diffuse pattern
Genetic	• Cystic fibrosis (CF) • Primary ciliary dyskinesia • Alpha-1-antitrypsin deficiency	CF – upper lung involvement Kartagener syndrome (chronic sinusitis, situs inversus, bronchiectasis)
Congenital	• Tracheomalacia • Bronchomalacia • Cartilage deficiency (Williams-Campbell syndrome) • Tracheobronchomegaly (Mounier-Kuhn syndrome)	Expiratory chest CT shows airway collapse
Miscellaneous	• Post-radiation fibrosis • Idiopathic pulmonary fibrosis • Young syndrome (bronchiectasis, sinusitis, azoospermia) • Idiopathic	Fibrosis will cause "traction bronchiectasis"

Table 1 (9, 12, 13)

Presentation: Patients have a history of frequent respiratory infections, chronic cough, sputum production, dyspnea; less commonly hemoptysis and pleurisy (1).

Physical Exam: Crackles (diffuse or focal), wheezes, digital clubbing (1).

Diagnosis:

- Detailed medical and family history
- Chest radiographs – not sensitive, but may show linear atelectasis and "tram tracks" suggestive of airway dilation (12)
- CT – confirms diagnosis and can specify subtypes (cylindrical, varicose, or cystic)
    - Airway to arterial ratio ≥ 1.5 (cross-sectional diameter of airway is 1.5x that of accompanying vessel) (12)
    - Lack of distal tapering of bronchi - "tram tracking" (12)
    - Presence of visible airways ≤1cm from pleural surface (12)
- Investigation should also seek underlying cause – CBC w/ differential, immunoglobulin quantitation, CF testing, aspergillus titers, HIV, alpha-1 antitrypsin deficiency, rheumatoid arthritis, Sjogren's Syndrome, sputum cultures

Treatment:

- Exacerbations - worsening of symptoms (cough, sputum, dyspnea, fatigue, hemoptysis, reduced FEV1/FVC ratio) in 48 hours (2)
    - Obtain sputum culture
    - Empiric treatment based on prior cultures
        - Clinically stable – PO antibiotics
            - No prior cultures – fluoroquinolones
            - Prior cultures without pseudomonas – amoxicillin or macrolide
            - Prior cultures with pseudomonas – ciprofloxacin; if suspect quinolone resistance, use IV antibiotics (2,10)
        - Clinically unstable (sepsis, hemoptysis, failed OP therapy) – IV antibiotics
            - Pseudomonal and MRSA coverage pending culture results; if suspect resistance, consider dual antipseudomonal coverage (3,10)
- Prevention/Maintenance
    - Maximize airway clearance
        - Mucokinetics (albuterol), mucolytics (nebulized hypertonic saline), chest physiotherapy, pulmonary rehab, exercise, hydration (11)
        - Dornase alpha (breaks down DNA) - beneficial in CF; not beneficial in non-CF (4)
    - Minimize infection
        - Recurrent exacerbations without pseudomonas or nontuberculous mycobacterium in sputum – chronic macrolide therapy (azithromycin) (6)
        - Recurrent exacerbations with pseudomonas in sputum – inhaled anti-pseudomonal therapy (tobramycin) (5,6)
        - Vaccines for patients with chronic respiratory conditions
        - Treat underlying causes

Clinical Pearls:

- The presence of pseudomonas in sputum is associated with increased death, exacerbations, and hospital admissions. (7)
- Patients with three or more exacerbations per year have twice the mortality rate of those who do not. (8)

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