Bronchiectasis

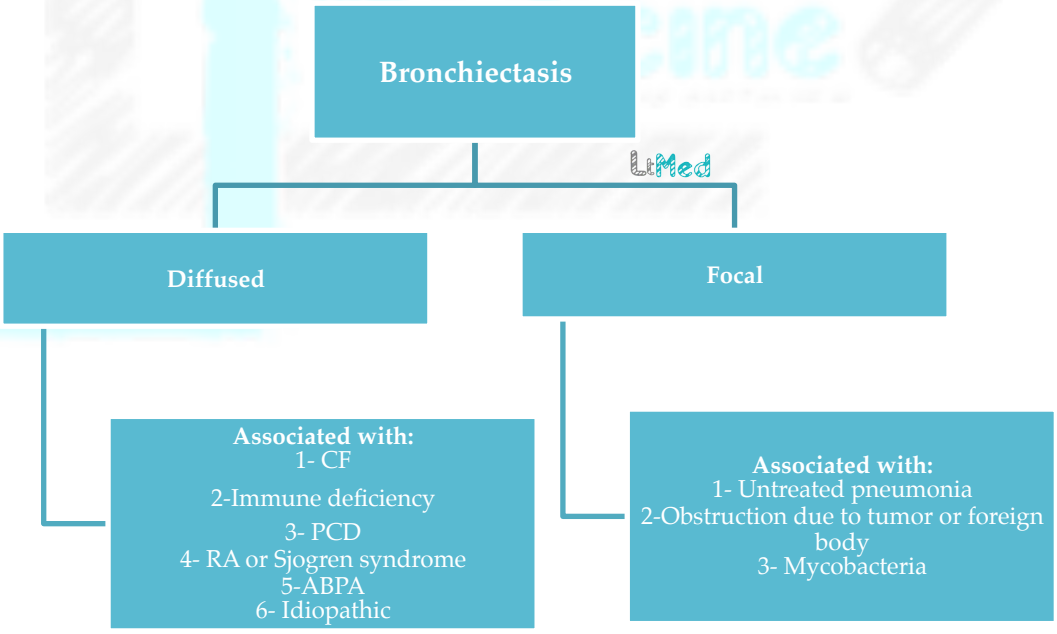
- **Definition:** Abnormal permanent anatomical dilatation of bronchioles that cannot be cured or reversed.
  - The large bronchi become *inflamed, and walls become thickened, accompanied with mucociliary dysfunction.*

Figure 1: Bronchiectasis gross appearance.

- **Epidemiology:**
  - A rare disease nowadays, due to effective treatment of pulmonary infections.
  - The prevalence of bronchiectasis *increases with age.*
  - Onset of the process is usually at *childhood.*

- **Causes:**
  - **Cystic fibrosis (CF):**
    - *The most common cause, accounting for more than 50% of cases.*
  - **Other:**
    - Recurrent infections: tuberculosis, pneumonia, and lung abscess.
    - Pan-hypogammaglobulnemia or immune deficiency.
    - Allergic bronchopulmonary asperigellosis (ABPA).
    - Collagen-vascular disease (rheumatoid arthritis RA, or Sjögren syndrome).
    - Lung tumors or foreign body.
    - Primary ciliary dyskinesia (PCD).
    - Idiopathic.

- **Types:**



- **Pathophysiology:**
  - Recurrent infections will trigger the neutrophils & cause inflammation.
  - Inflammatory mediators destroy elastin, muscles, and cartilage in the large & medium airways➔ irreversible bronchodilation.
  - Lymphocytes and macrophages will infiltrate the mucosal walls and cause thickening, which is responsible for the airway obstruction.
  - With time, the disease will progress and spread to the lung parenchyma causing fibrosis.
  - Impaired airway clearance mechanism will also contribute to the airway obstruction.
  - Various organisms will frequently colonize chronically dilated bronchi.

- **Clinical Presentation: (can be difficult to differentiate from chronic bronchitis)**
  - *Recurrent mucopurulent (khaki colored) foul smelling productive cough, in large amounts.*
  - *Hemoptysis*: due to rupture of the bronchial arteries.
  - Dyspnea and wheezes.
  - Weight loss.
  - Coarse crackles.
  - Digital clubbing (rare).
  - Signs of anemia (anemia of chronic diseases).

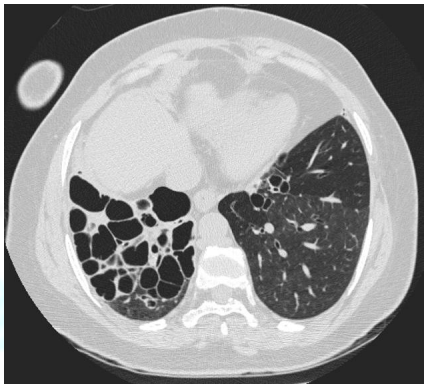


Figure 2: Bronchiectasis.

- **Diagnosis:**
  - **Chest X-ray (CXR):**
    - The best initial test.
    - Dilated thickened bronchi (tram-track appearance).
  - **High-resolution CT: gold stabdard**
    - The most accurate diagnostic test (nearly 100% sensitive & specific).
    - Airways are larger than their associated vessels.
    - Dilated thickened bronchi (signet ring appearance).
  - **Sputum culture & gram staining:**
    - Important for effective treatment.
    - Major pathogens are:
      - *H.influenzae (35%)*
      - *P.aeruginosa (31%)*
      - *M.catarrhalis (20%)*
      - *S.aureus (14%, especially in CF patients)*
      - *Anaerobes*
      - Others*: S.pneumoniae, K.pneumoniae, aspergillus fumigatus, M.avium.*
  - **Sweat chloride test:**
    - If cystic fibrosis is suspected.

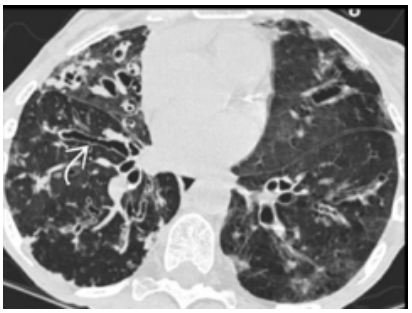


Figure 3: Tram-track appearance.

- **Treatment:**
  - Chest physiotherapy and postural drainage:
    - Cupping and clapping.
    - At least 3 times daily for 10-20 minutes.
  - Antibiotics:
    - Oral, IV, or inhaled.
    - Rotate antibiotics, 1 drug weekly each month.
    - Choices:
      - *Macrolides (azithromycin, or clarithromycin).*
      - Cephalosporins (Cefuroxime, cefaclor, or cefixime).
      - Quinolones (levofloxacin, or moxifloxacin).
  - Bronchodilators: in patients with airflow limitation.
  - Mucolytics
  - Oxygen
  - Inhaled or oral steroids: to delay disease progression and for ABPA.
  - Surgical resection of localized lesions (rarely used).

- **Complications:**
  - Pneumonia.
  - Empyema.
  - Pneumothorax.
  - Metastatic cerebral abscess.
  - Massive hemoptysis:
    - Mortality is 25%.
    - Due to rupture of high-pressure systemic bronchial arteries.
    - Usually self-limited, if did not stop bronchial artery embolization is the treatment of choice.
- **Prognosis:**
  - Patients with cystic fibrosis have the poorest outcome.
  - Most bronchiectasis patients will eventually develop respiratory failure or cor pulmonale.

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Written by: Roaa Amer
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Reviewed by: Haifa Al Issa
Areej Madani