**Allergic bronchopulmonary aspergillosis**

Lim and Weller, “Allergic bronchopulmonary aspergillosis.” *UpToDate 11.3*


Types of aspergillus-related disease:
- Aspergilloma
- Chronic necrotizing pneumonia
- Invasive pulmonary aspergillus
- ABPA

Prevalence:
- Asthma: 1-2%, 7-14% if steroid-dependent
- CF: 2-16%

Pathophysiology: limited understanding
- Host response: IgE, IgG response; T cells
- Exposure to airborne Aspergillus spores (2-3 ug) – not dose related
- Fungal enzymes / mycotoxins
- Result: mucoid impaction, eosionophilic pneumonia, bronchocentric granulomatosis, reactive airways

Stages
- I: Acute flare – upper / middle lobe, IgE sharply elevated
- II: Remission – no infiltrates off prednisone 6 months, IgE normal / elevated
- III: Recurrent exacerbations / flares – upper / middle, IgE sharply elevated
- IV: corticosteroid-dependent asthma - variable infiltrates and IgE
- V: fibrotic lung disease – fibrotic, bullous, cavitary lesions

Clinical features:
- Bronchial obstruction: mucus plugs, wheeze
- Hemoptysis
- Fever
- Peripheral blood eosinophilia
- Reduced FEV1 and increased RV, reduced FVC if mucus plugging, occasional reversibility with bronchodilator

Radiographic features:
- Parenchymal infiltrates
- Atalectasis
- Bronchiectasis
  - CXR:
    - Tram line (thickened)
    - Parallel line (ectatic bronchi)
    - Ring / toothpaste / gloved finger shadows (filled bronchi)
    - Perihilar infiltrates
  - CT
    - Upper lobe predominance, proximal cylindrical bronchiectasis (not sensitive or specific for ABPA)

Diagnosis:
- Major diagnostic features:
  - History of asthma
  - Central bronchiectasis
  - Immediate skin test reactivity to Aspergillus antigens
    - Positive in asthma along in 20-30%
  - Elevated serum IgE and/or IgG to A. fumigatus
Serum total IgE > 1000ng/mL
- Supportive
  - Precipitating serum antibodies to A. fumigatus
    - Positive in asthma and COPD in 10%
  - Lung infiltrates
  - Peripheral blood eosinophilia >500/mm
    - DDx: Acute or chronic eosinophilic pneumonia, drug-induced EP, Churg-Strauss, Loeffler’s, autoimmune diseases, crack cocaine, hypereosinophilic syndrome
- Sputum / mucus plug culture of variable yield
- Biopsy rarely performed
- ABPA-seropositive: asthma, skin reaction, IgE, IgE-IgG to A. fumigatus without bronchiectasis

Treatment
- Corticosteroids: treats bronchospasm, infiltrates, IgE;
  - 0.5 mg/kg/day for 1-2 weeks, tapering over 2-6 months
  - Monitored with serial (monthly / bimonthly) serum total IgE (35% reduction)
  - Resolution of radiography (within 2 months)
- Inhaled steroids: for symptoms, but not preventing flares
- Itraconazole: RCT study of 55 patients on steroids → 16 weeks itraconazole, 46% vs. 19% likelihood of response (reduced steroid dose, IgE, PFT or exercise tolerance or radiography)
- No data on voriconazole
- Environmental modification?

Bronchiectasis


Causes
- Postinfectious conditions: bacteria, TB/MAI, aspergillus, viral (adenovirus, flu, HIV)
- Congenital: CF, primary ciliary dyskinesia, alphal-antitrypsin deficiency, Marfan’s, pulmonary sequestration
- Immunodeficiency
  - Primary Hypogammaglobulinemia
  - Secondary: cancer, chemotherapy, immune modulation after transplant
- Inhalation / aspiration (chlorine, heroin OD, foreign body)
- Rheumatic conditions: RA, SLE< Sjogren’s, relapsing polycondritis
- Other: IBD, etc.

Diagnostic testing for bronchiectasis:
- CBC, IgG, IgA, IgM
- HRCT
- Spirometry
- Consider: RF, IgE, ABPA, IgG subclasses, alphal-AT, sinus CT, sputum, bronchoscopy, sweat chloride test analysis

Exacerbations:
- Clinical: sputum, dyspnea, cough, fever, wheeze, decreased exercise tolerance, reduced PFT, CXR changes
- Treatment: antibiotics (FQ) X 7-10 days
- Prevention: variable results in variable studies on antibiotic prophylaxis (Cochrane review)
- Pulmonary toilet (PT/drainage not validated in Cochrane review)
- IR/surgery for bleeding