Pulmonary Fibrosis

Definition of pulmonary fibrosis

- The end-stage of many different pulmonary diseases. The lungs become fibrosed (thickened, stiff and scarred) and lack their usual elasticity and compliance. It can be localised (e.g. one lobe) or affect the whole lung.

Epidemiology of pulmonary fibrosis

- Idiopathic disease approximately 10 per 100,000 per year
- Increases with age: peak incidence 50-70 year-olds

Causes of pulmonary fibrosis (see fibrosis question section for mnemonics)

- Usual interstitial pneumonia (UIP)
  - Also called Idiopathic Pulmonary Fibrosis (IPF) and Cryptogenic Fibrosing Alveolitis (CFA)
- Rheumatological
  - Rheumatoid arthritis; SLE; dermatomyositis; polymyositis; mixed connective tissue disease; Sjogrens; anklylosing spondylitis; systemic sclerosis (CREST)
- Occupational
  - Asbestosis
  - Silicosis
  - Beryliosis
  - Pneumoconiosis
  - Extrinsic Allergic Alveolitis (EAA)
- Infection
  - Aspergillosis (ABPA)
  - TB
- Drugs
  - Amiodarone
  - Methotrexate
  - Bleomycin
  - Cyclophosphamide
  - Nitrofurantoin
- Vasculitis
  - Wegeners; Churg-strauss; Goodpastures
- Radiation fibrosis
- Aspiration
- Sarcoid
- Histiocytosis

Presentations of pulmonary fibrosis

- Shortness of breath
- Cough: usually non-productive
- Haemoptysis
- Chest pain
- Wheeze
- On examination:
  - Clubbing
  - Fine bilateral basal ‘velcro’ crackles

- Signs of cor pulmonare (RV heave, peripheral oedema, raised JVP)
- Signs of underlying cause (e.g. associated auto-immune disease)

Differential diagnosis of pulmonary fibrosis

- COPD (predominantly bronchitis)
- Asthma
- Heart failure

Investigation of pulmonary fibrosis

- Bloods
  - FBC, U&E, LFT, CRP
  - ESR
  - Autoimmune profile
    - RF; ANA; ANCA; anti-GBM
  - CK (raised in dermatomyositis)
  - ACE (may be raised in sarcoid)
- ABG (Type 1 respiratory failure)
- CXR
  - Reduced volume and reticulonodular shadowing
  - Honeycombing
- Lung function tests
  - Restrictive pattern (FEV1/FVC>70%)
  - Low TLC
  - Reduced K_{CO}
- HRCT
  - Distribution of fibrosis to sub-pleural lung is typical of IPF/ UIP
  - Reticular opacities
  - Sub-pleural honeycombing
  - Traction bronchiectasis
- Broncho-alveolar lavage (BAL) and biopsy
  - Important as lymphocytes > neutrophils indicate a better response to steroids and a better prognosis

Diagnostic criteria for Idiopathic pulmonary fibrosis:

- Major Criteria
  - Exclusion of other causes e.g. drug toxicities, connective tissue disease
  - Abnormal pulmonary function studies (evidence of restriction)
  - Bibasilar reticular abnormalities with minimal ground glass opacities on HRCT
  - No evidence of alternative diagnosis on transbronchial biopsy or BAL
- Minor Criteria
  - Age > 50
  - Insidious onset of otherwise unexplained dyspnoea on exertion
  - Duration > 3 months
  - Bibasilar inspiratory crackles
- Usual interstitial pneumonia (UIP) is defined by a heterogenous appearance with alternating areas of normal lung, interstitial inflammation, fibrosis and honeycomb change affecting the peripheral supleural parenchyma most severely.

Management of pulmonary fibrosis

- Medical
  - Prednisolone
  - Azathioprine and cyclophosphamide
  - Pirfenidone
    - Licensed for use in patient with UIP with FEV 50-80% predicted (NICE guidelines)
- Home oxygen
- Pulmonary rehabilitation
- Lung transplant
  - Indications
    - Age under 60 with expectancy under 18 months
    - Match for ABO, not Rh
    - Double transplant preferred
    - Donor
      - Good cardiac and lung function, age under 40
      - Chest diameter slightly less than recipient

Complications of pulmonary fibrosis

- Respiratory failure
- Pulmonary hypertension
- Pneumothorax
- Increased risk of bronchogenic cancer

Prognosis of pulmonary fibrosis

- Variable and depends on type
- Highly cellular with ground glass infiltrate: good response to immunosuppression
  - 80% 5 year survival
- Honey-combing on CT and no response to immunosuppression
  - 20% 5 year survival

Common questions concerning pulmonary fibrosis

- What are the respiratory causes of clubbing?
  - Fibrosis (and TB)
  - Chronic suppurative lung disease (bronchiectasis, empyema, abscess)
  - Lung cancel (not small cell – as progresses too quickly)
- What causes upper vs lower lobe fibrosis?
  - Upper lobe (A TEA SHOP)
    - ABPA
    - TB
    - Exrinsic allergic alveolitis
    - Ankylosing spondylosis
    - Sarcoid
    - Histiocytosis
    - Occupational
      - Berylliosis, silicosis
    - Pneumoconiosis
  - Lower (BRAD)
    - Bronchiectasis

        - RA (and other connective tissue disease)
        - Asbestos
        - Drugs
- What is histiocytosis?
    - Aulti-system disease with abnormal proliferation of histiocytes. Commonly affects young people.
        - Respiratory
            - Mixed obstructive and restrictive pattern
            - Low KCO, low FEV1/FVC ratio
        - Renal damage
        - Skin
            - 1-5mm papules
        - Lymphadenopathy
        - Bones
            - Radiolucent skull lesions
            - Marrow invasion
- What is an acute exacerbation of pulmonary fibrosis?
    - An unexplained worsening or development of dyspnea within 30 days with new radiological infiltrates on HRCT
    - Poor prognosis
    - Treatment is iv steroid as 1st line and other immunosuppressant (e.g. cyclophosphamide) as 2nd line