Sarcoidosis

Multisystem chronic inflammatory idiopathic condition characterised by non-caseating epithelioid granulomata at various sites particularly the lungs and thoracic cavity.

Epidemiology/Risk Factors

- F>M. Commoner in Scandinavian & Caribbean-origin people. Rare in Aborigine or SE Asians
- Occupational exposure to Be, Al and zirconium.
- Family history

Presentations

Asymptomatic: up to 50% & diagnosed on routine CXR.
Non-specific symptoms: 30% present with fever, fatigue, cachexia and lassitude esp if Black.
Acute presentation: Erythema nodosum & polyarthritis. Commoner in white F, often remits<2yrs.
Chronic, progressive pattern: ~10-30% have onset over 2yrs.

Symptoms

Constitutional upset: Fever and night sweats, malaise, fatigue, weight loss.
Lung: >90%. Usually interstitial disease. Dry cough, fever and SOB, chest discomfort.
Lymphadenopathy 75%. All common groups may be involved. Often asymptomatic.
Skin: 30%. Erythema nodosum, cutaneous granulomas, lupus pernio (pink nose/cheek plaques). Scars may have granulomatous infiltration.
Eye: 20%. Granulomatous uveitis, conjunctivitis. Dry eyes.
Joints: 15%. Acute polyarthrits of lower leg or hands.
Hypercalcaemia and hypercalciuria: 10-15%.
Neurosarcoidosis: CN palsies (VII, II, VIII, IX, X), seizures, CVA, peripheral neuropathies.
Heart disease: arrhythmias, HF, cardiomyopathy.
Liver: HSM, asymptomatic LFT derangement.
Heerfordt's syndrome: parotiditis with uveitis and facial nerve palsy.
Other: Bone marrow suppression, nosebleeds, rhinitis, tonsillar involvement.

Investigations

Bedside: Urine (?hypercalciuria), ECG (arrhythmias)
Bloods: FBC, ESR↑, UEC, CMP (↑Ca), LFT, ACE (↑ in 60%)
Imaging: CXR/CT show bilateral hilar or paratracheal lymphadenopathy + interstitial disease.
Kveim test: intra-dermal inj sarcoid splenic material & biopsy of any nodule formed.
Other: Gallium scan (detect extra-pulmonary disease), Lung function tests (restrictive pattern), liver or LN biopsy (non-caseating granulomata), bronchoalveolar lavage (↑CD4:CD8 ratio)

CXR Staging

Stage 0 - Normal findings
Stage I - Bilateral hilar (± paratracheal) LN
Stage II - Bilateral hilar LN + pulm. infiltrates
Stage III - Parenchymal infiltrates w/o LN
Stage IV - Parenchymal involvement + vol loss (pulmonary fibrosis). May be other features (cavitations, calcifications, hilar retraction, bullae, cysts, and emphysema)

Management

Pulmonary disease: Stage 0-I: symptomatic Rx only. Stage II+: 1-2y steroids + bisphosphonates (to ↓osteoporosis). MTX, azathioprine. infliximab, leflunomide & chloroquine may be used.
Extra-pulmonary disease: Treat with std Rx and often with (high-dose) steroids.

Prognosis

- ~66% resolve in the long term. 10% have progressive disease. Mortality is ~1-5%.