Giant cell arteritis (GCA) and polymyalgia rheumatica (PMR)

Definitions of giant cell arteritis (GCA) and polymyalgia rheumatica (PMR)

- Giant cell arteritis is an inflammatory vasculitis affecting large vessels.
  - Commonest vasculitis & among the commonest causes of acute vision loss
  - Unknown aetiology, but is associated with polymyalgia rheumatica,
- Polymyalgia rheumatica (PMR) an inflammatory condition affecting the muscles and joints of the pelvic and shoulder girdles

Epidemiology of GCA and PMR

- 29 per 100,000 (in Europe)
  - 50% of GCA have PMR
  - 15% of PMR have CGA
- Increases with age
  - Very rare in under-50s, with mean age of onset at 70 years
- Women > men 3:1
- Caucasian > Afro-Caribbean

Aetiology of GCA and PMR

- Aetiology unknown, except that it is an inflammatory vasculitis of large arteries.
- May be a genetic component as familial clusters sometimes occur

Pathology of giant cell arteritis (GCA)

- Abnormal, sometimes pulseless temporal arteries
- Histology on biopsy
  - Inflammatory infiltrates
  - Fragmentation & distortion of internal elastic lamina
  - Multinucleated giant cells in <50%

Presentation of giant cell arteritis (GCA)

- Headache (usually temporal)
- Jaw claudication
- Scalp tenderness
- Tenderness and/or thickening of one of the temporal arteries
- Visual impairment (involvement of ophthalmic artery)
- Systemic features
  - Anorexia, weight loss, fever, sweats, malaise
- Other systems
  - Aortic root involvement in 15%, which can cause aneurysms years later.
    - Can cause asymmetric pulses and BP, and bruits
  - Strokes can occur due to ischaemia and narrowing of aortic or intracerebral vessels

Differential diagnosis of giant cell arteritis (GCA)

- Headaches
  - Migraine, tension headache, trigeminal neuralgia, cluster headache
- Intracranial pathology (tumour, vascular malformation)

- TIA (vision loss)
- Cervical spine disease
- Sinus disease
- TMJ pain
- Systemic vasculitides/connective tissue disorders (all rare) e.g.
    - Takayasus (young women, absent arm pulses, visual loss, carotid bruits)
    - Polyarteritis nodosa (muscle pains, "abdominal angina", livedo reticularis, anaemia, raised WCC, raised ESR, proteinuria)
    - Polymyositis (proximal muscle weakness, raised CK)

Diagnosis of giant cell arteritis (GCA)

- The American College of Rheumatology's criteria for diagnosing giant cell arteritis is:
    - Age >50 years
    - New onset headache
    - Abnormalities of the temporal arteries on palpation
    - ESR >50 mm/hour
    - Abnormal temporal artery biopsy.
- The presence of three or more criteria has a sensitivity of 97% and a specificity of 79% for a diagnosis of giant cell arteritis.

Presentation of polymyalgia rheumatica (PMR)

- Age >50
- Sudden onset, severe pain of shoulders, neck, hips and lumbar spine
    - For >2/52
    - Difficulty combing hair or reaching to shelves.
- Symptoms are worse in the morning, and can last for several hours (at least >45min)
- Systemic features
    - Anorexia, weight loss, fever, sweats, malaise
- Even PMR predisposes to vascular events so should be treated promptly

Investigations in giant cell arteritis (GCA) and polymyalgia rheumatica (PMR)

- Bloods
    - Anaemia of chronic disease
        - Can have deranged LFTs
    - ESR and CRP usually very raised
        - 95% of GCA will have an ESR over 50
        - CK normal
    - Protein electrophoresis to exclude myeloma
- CXR to look for aneurysm formation
- Urinalysis (blood and protein for systemic vascular involvement)

- **PMR-specific investigations**
    - Search for malignancy with a good history and full examination including skin breast exam and digital rectal exam (DRE).

- **GCA-specific investigations**
    - Temporal artery biopsy
        - Can be done within 14 days of starting steroid treatment but loses sensitivity over time

        - May be negative due to skip lesions
        - Shouldn't delay steroids for biopsy
    - PET scan if suspicion of large vessel involvement
        - E.g. Marked systemic features, high inflammatory markers despite steroid treatment
        - NB may be non-diagnostic if on >10mg/day prednisolone
    - Colour duplex of temporal arteries
        - Sensitivity (and negative predictive value) 95%

Treatment of giant cell arteritis (GCA) and polymyalgia rheumatica (PMR)

- Treatment of both GCA and PMR is steroids.
    - The reducing regime must be tailored to the patient's symptoms and inflammatory response. The dose depends on whether GCA is present
    - Both diseases tend to require 18-24 months of treatment

- **Giant cell arteritis (GCA)**
    - If no visual involvement: 40-60mg prednisolone OD
        - Symptoms should improve in 7-14 days
        - Taper dose after 1-2 months gradually
            - By 10mg every 2/52 to 20mg, then by 2.5mg every 2-4/52 to 10mg, then by 1mg every 1-2 months
    - Vision loss/amaurosis fugax
        - IV methylprednisolone 500mg-1g od for 3 days
        - Consider 60mg prednisolone PO if established visual loss
    - 30-50% can stop taking steroids after 2 years. Some patients will be unable to come off of steroids completely and may be on a low dose long-term; in these people a steroid-sparing immunosuppressant (usually methotrexate) may be considered.

- **Polymyalgia rheumatica (PMR)**
    - Prednisolone PO 15mg for 3-4/52
        - Reduce by 2.5mg every month until 10mg, then by 1mg/month
    - Symptoms often improve quicker than GCA

- Steroid protection (50% on long-term steroids will get a fracture)
    - Calcium and vitamin D supplements
    - Bisphosphonate
    - PPI
    - 75mg aspirin (reduces visual loss & CVAs) unless contraindications
    - Consider DEXA in under 70s

Prognosis of GCA and PMR

- Visual symptoms are often irreversible
    - If one eye affected, 20-50% chance of bilateral vision loss without treatment
- Most patients respond to steroid treatment, but on average need two years of treatment
    - There is therefore treatment-related morbidity: osteoporosis, mood changes, obesity, hypertension, hyperlipidaemia, myopathy, diabetes, cataracts, skin changes, bruising, fluid retention.
    - If poor or inadequate response to steroids, steroid sparing agents such as methotrexate can be used under rheumatological guidance