Temporal Arteritis and Polymyalgia Rheumatica

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Comparison

• Both vasculitides
• Similar age and sex distributions
• Both associated with HLA-DR4
• 50% GCA patients have PMR
• 15% of pts with PMR go on to develop GCA
• 10-30% PMR patients have positive temporal artery biopsy
Temporal Arteritis

- Giant Cell Arteritis
- First described clinically by Hutchinson in 1890
- First histological description Horton et al 1932
- Systemic, chronic granulomatous vasculitis
- Cranial branches originating from aortic arch
Epidemiology

- Commonest vasculitis
- Incidence highest in Scandinavia
- 15-35/100,000 >50 yrs
- Prevalence ≤1/500
- Mean age 72
- Female predominance ~5:1
- Pathogenesis unknown
  - ? Infectious aetiology
Presentation

- Headache (68%)
- Scalp tenderness
- Visual disturbance (39%)
- Jaw claudication (45%)
- Facial Pain
- Earache/ toothache/ pain in palate
- Non-specific picture
  - PUO
- Cerebrovascular disease (1-25%)
- Myopathy/ neuropathy
- Seizure
- May have a prodrome of general malaise, fever, night sweats and weight loss lasting days to weeks (40%)
Examination

- Visible +/- tender temporal artery
- Pulselessness
- Scalp tenderness
- Bruits
- Eye signs
  - Diplopia, ptosis, INO, abnormal pupils, fundoscopy
Labs

• Raised ESR + CRP

• Normocytic normochromic anaemia

• Thrombocytosis

• Raised alkaline phosphatase and _GT
1990 Criteria for the Classification of Giant Cell (Temporal) Arteritis

1. Age at disease onset >=50 years
2. New headache
3. Temporal artery abnormality
4. Elevated erythrocyte sedimentation rate
5. Abnormal artery biopsy

* For purposes of classification, a patient shall be said to have giant cell (temporal) arteritis if at least 3 of these 5 criteria are present. The presence of any 3 or more criteria yields a sensitivity of 93.5% and a specificity of 91.2%

Temporal Artery Biopsy

- Within one week of commencing treatment
- Positive in 85% GCA
- Approximately 35% positive overall
- Usually unilateral
- Length $\geq 1\text{cm}$ necessary, ideally $>3\text{cm}$
  - Skip lesions
- Complications
  - Scalp necrosis, facial nerve damage, stroke

Histology

- Normal Biopsy
- Giant Cell
- Narrowed Lumen
- Thickened intima
- Positive Biopsy
Ultrasonography in GCA

- Non-invasive
- “Halo sign”
- 69% sensitive, 82% specific compared to biopsy
- Presence of stenosis/occlusion as sensitive as biopsy
- User dependent

Complications of GCA

- Anterior ischaemic optic neuropathy
  - Most common cause of blindness in GCA
- Ophthalmic artery or branch occlusion

15-20%
Steroids

• Initially 40-60mg prednisolone daily x 8/52
• Over 4 weeks, half dose
• Reduce by 5mg/ 3-4 weeks until 10mg/day
• Then by 1mg every 6-8 weeks
• IV Methylprednisolone used in some centres if visual loss present
• Average length of Rx 1.5-2 years. Many pts on long term low dose prednisolone
Flares

• Important to diagnose accurately due to risk of complications vs side effects of steroids
• Symptoms, ESR and CRP
• Anticardiolipin antibodies high in temporal arteritis but not other inflammatory dx
• Return to dose of prednisolone that last controlled the TA

Other treatments

• Bisphosphonates

• Aspirin 75-100mg/day thought to reduce risk of ischaemic events once diagnosed

• Evidence does not support addition of steroid sparing agents
Polymyalgia Rheumatica

- Autoimmune inflammatory disease
- First description probably made by Bruce in 1888
- Barber suggested the present name in 1957
- Aching and morning stiffness
- Subacute onset
- Often diagnosis of exclusion
- Response to steroids excellent
Epidemiology

• Almost exclusively in >50 yrs
• Highest incidence in Northern Europe
  – 113/ 100,000 Norway
• Prevalence up to 1%
• Female: male = 2:1
• Mortality similar to general population
• Pathogenesis unknown
Presentation

- Ache + chronic stiffness
  - Shoulders, hips, neck, torso
  - Gel phenomenon
  - Pain worse at night
- Usually symmetric
- Difficulty performing daily tasks like dressing
- Systemic symptoms
Physical Examination

- Normal muscle strength
- May have decreased active ROM
  - Disuse atrophy
  - Pain
- Tenderness due to synovitis
- May have signs GCA
- Uncommonly have pitting oedema of extremities
Labs

• Raised ESR
• Raised CRP
• Normochromic, normocytic anaemia (50%)
• Serology usually negative
• Normal CK
Differential

- Joint disease
- Bone disease
- Muscle disease
- Infections
- Hypothyroidism
- Functional
- Myeloma
Subacromial Bursitis
Diagnostic Criteria: Bird/Wood

- Bilateral shoulder pain and/or stiffness
- Less than two weeks from onset of symptoms to maximal symptoms
- ESR greater than 40 mm per hour
- Morning stiffness lasting longer than one hour
- Patient older than 65 years
- Depression and/or weight loss
- Bilateral upper arm tenderness

- Dx requires 3 of 7 listed features
- Presence of 3 confirms a sensitivity of 92% & specificity of 80%

- BHPR exclusion criteria
  - Active infection
  - Active cancer
Treatment

- Daily prednisolone 15 mg for 3 weeks
- Then 12.5 mg for 3 weeks
- Then 10 mg for 4-6 weeks
- Followed by reduction by 1mg every 4-8 weeks or alternate day reductions (e.g. 10/ 7.5 mg alternate days etc)
- Usually need steroids for 1-3 years
- Bone protection
- Recommend adding DMARDs e.g. MTX after two relapses
- Treat relapse with previously successful steroid dose

BSR & BHPR Guidelines for the Management of Polymyalgia Rheumatica (PMR)
Diagnosis

- Essentially clinical
- Guided by response to treatment
- Improvement quickly with steroids
- Reassess diagnosis if not improving
- Reconsider diagnosis if atypical presentation
Referral

- Primary care diagnosis
- Referral to specialist warranted if
  - Young patient
  - Red flags
  - Suboptimal response to steroid
  - Difficulty weaning Rx
Follow up

• 3 monthly GP follow up for 1st yr
• Assess complications
• Any features suggesting alternate diagnosis?
• Inflammatory markers
Summary

• PMR and TA considered as two separate diseases of older patients
• May coexist
• Both responsive to steroids
• Both prevalent and can lead to both disease and treatment related complications
• TA should be treated as an emergency
Thank You!

Oh, sure I'm fine... considering the shape I'm in!