Polymyalgia, Temporal Arteritis and pineapples

Rod Hughes Consultant Rheumatologist Ashford St Peter’s Hospital Trust Chertsey Wed 11th May 2011

Meeting aims

• Pineapples – their significance in disease
• Defining both diseases
• Give clinical examples
• Outline Investigation and treatment
• Give adverse effects of treatment
• Make recognition easy

Pineapples - Ananus comosus Bromeliaceae
A sign of welcome

Pineapples

- Both the fruit and root can be applied topically or eaten as an anti-inflammatory and proteolytic agent
- Bromelain from pineapples can be used in the diet to treat colitis in mice
- Pinapples are rich in manganese and can be used to induce menstruation or abortion
Mrs SE aged 68

- Normally fit and well – playing golf and walking regularly
- Developed shoulder pain – gradual onset left, then right, over two weeks
- Diagnosed as ‘frozen shoulders’ by GP and given anti-inflammatories
- Slightly better

- Neck ache
- Fatigue and waking at night due to stiffness
- Gradual onset of thigh aching – attributed to bursitis by GP
- Some response to diclofenac – taking regularly
- Starting to feel very stiff
- GP – ESR 15 – stiffness attributed to OA

- Carried on for four months
- Unable now to use the bath
- Cannot easily get restful sleep at night
- Sent to physiotherapy by GP
- 7 sessions but no benefit – reliant on anti-inflammatories every day
- Eventually referred to rheumatology
• Very stiff especially in proximal muscles
• Unable to rise from chair without using arms
• Unable to actively move arms above shoulders – passive movement full
• Tender and uncomfortable on resisted movement of thighs and shoulders

• ESR 24  CRP 12
• Diagnosis – likely polymyalgia
• No headaches, no visual symptoms
• 15mg steroids given for two weeks
• Within twelve hours 95% symptoms gone
• Fatigue responded
• Stiffness resolved

• Diagnosis - PMR

Polymyalgia (PMR)
• Clinical syndrome of the middle aged and elderly
• Pain and stiffness in the neck, shoulder and pelvic regions
• Gradual or sudden onset
• Dramatic response to steroid therapy
Prominent temporal arteries
Difficulty attending morning service
Rheumatic pains
Generalised ill health

Brought to Europe by Columbus from South America in 1493
Temporal Arteritis

- A vasculitis often accompanying PMR
- Known as Giant Cell arteritis (GCA), Temporal arteritis (TA) or granulomatous arteritis
- Early recognition essential to avoid irreversible blindness
- Late complications if not treated early and aggressively – higher dose steroids than PMR

Epidemiology

PMR
- White
- Over 50’s
- 3300/100000
- HLA link
- women x2
- Average age 70

TA
- White
- Over 50’s
- 223/100,000
- HLA link
- women x2

PMR presentation

- Pain + stiffness shoulder/neck/pelvis/back
- Bilateral/symmetrical
- Synovitis - low grade knees/wrists/ sternoclavicular joints
- Muscle strength unimpaired but movement inhibited
- Fever/ weight loss/malaise
Diagnostic Criteria for PMR - Summary
1st 3 +ve and any number of others

- Pain/Stiffness in proximal girdle
- Steroid responsive. Greater than 70% improvement with 15mg oral pred over 2 weeks
- Older than 50

- ESR greater than 40
- Normal TFT and myeloma screen
- Weight loss, fever or joint pains

Complications of PMR

- Can sometimes develop into more classical Rheumatoid arthritis – joints become inflamed
- PMR-onset RA
- PMR and TA can sometimes develop alongside cancers either before diagnosis or when known about

TA presentation

- Headache - severe and localized to temples
- Fever
- Scalp tenderness
- Jaw pains after eating (claudication)
- 25-50% have transient visual field loss/mistiness
- Blindness 6-10% when unrecognised
Rare manifestations of TA

- Stroke
- Peripheral neuropathy
- Deafness
- Depression
- Systemic affects of vasculitis - heart/liver/thyroid/aortic dissection
TA biopsy

- Often under local anaesthetic
- Minor operation
- Results in a small piece of artery sent to the laboratory
- Sometimes misses the artery
- Should always be considered in TA
Aims of new imaging

- Ultrasound and PET scanning
- Avoid the need for temporal artery biopsy
- Diagnose TA
- Reveal extent of vasculitic involvement in PMR and TA
- Assess treatment effects
Sites

- **Temporal arteries** – showing stenosis or vessel wall swelling
- **LV-GCA** – imaging subclavian, axillary and proximal brachial arteries and aortitis

Ultrasound

- **Conventional** US gives two-dimensional, black and white images
- **Doppler** US bounces short bursts of sound waves off moving RBC producing 2-D colour images
- **Duplex** combines both modes
PET scan

• Like a CT scan
• Uses activation and release of labelled glucose to image
• Available in Guildford at St Luke’s

Blood vessel effects of TA

PET Scan

Temporal arteritis can affect large and medium vessels throughout the body
Relationship between PMR and TA

- Clinical overlap
- Biopsy proven TA in 10-15% PMR
- Headaches almost always occur with TA – rare in PMR without TA
- Always enquire

Long term consequences of PMR - TA

- Steroid adverse effects
- Long term vascular effects of sustained inflammation – aortic aneurysm
Thoracic aortic aneurysm

Aortic aneurysm

- 1995 study from Minnesota
- TA patients 17x more likely to develop thoracic aortic aneurysms 5-10 years after TA
- TA patients 2.5 x more likely to develop isolated abdominal aortic aneurysms
- **Annual chest X rays** for patients with TA after treatment

**Investigations and diagnosis**

- ESR - normal in some cases
- Indicator of diagnosis
- Very non-specific
- Not a good way to monitor response
- Hb low
Can PMR/TA be mistaken for any other diagnosis

• Under active thyroid
• Muscle inflammation – CPK blood test
• Myeloma

Management

• NSAI may help and result in a delay in diagnosis

Steroid trial
• 15mg for two weeks for PMR
• 40mg for two weeks for TA

Reducing dose of steroids over 12 - 24 months

Bone prophylaxis to prevent OP

Steroid reduction

• Not too fast
• Beware flares
• Beware reducing according to the ESR
• A clinical disease with a clinical course
• Steroid adverse effects - may use steroid spacers (Aza/MTX/leflunomide)
Steroid reduction - PMR

- 15mg 1 month
- 12.5mg 1 month
- 10mg 1 month
- 9mg 1 month
- 8mg 1 month
- 7mg 1 month etc
Total minimum treatment time circa 1 year

Resolution of PMR/TA

- Usually self-limiting
- Duration 18 – 36 months
- Only 5% need steroid for longer that 36 months
- Unlikely to recur (but possible)
- Occasionally TA develops when PMR being treated

Steroid adverse effects

Not ‘important’ - reversible

- Weight gain
- Easy bruising
- Sweating/flushing
- Dreams ++
- Fluid retention
Steroid adverse effects

**Important**

- Osteoporosis
- Blood pressure elevation
- Skin thinning/ leg ulcers
- Cataracts
- Diabetes mellitus

- Upper body obesity with the arms and legs
- Buffalo hump
- Red, Re and Fad
- High Blood Sugar
- High Blood Pressure
- Varicose
- Acne
- Female Balding
- Water Retention
- Hyperthyroidism
- Thin Skin and Hyperpigmentation
- Purple Streak
- Poor Wound Healing
- Hyperpigmentation
- Severe Depression
- Cognitive Difficulties
- Enlarged Kidney
- Sleep Disorders
- Fatigue

*Minnie G*, Dr. Couling's first patient
Scanning electron microscopy of normal and osteoporotic bone

Burden of risk and fracture occurrence

• Steroids can cause bones to become weak

• More bone loss soon after steroids started than later

Steroid induced OP – always start bone protection early

• **Treatment with an anti-resorptive** - Choose between
  - Bisphosphonate (actonel, alendronate, didronel)
  - Protelos
  - HRT
  + calcium (1000mg) and Vit D (400 units) daily
PMR / TA

- Don’t miss the diagnosis
- Common diseases
- Educate to aid compliance with steroids (ACR leaflets)
- Use bone prophylaxis
- Attempt biopsy proof for TA
- Can sometimes develop as RA later
- Always carry a pineapple with you