Polymyalgia Rheumatica

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What is Polymyalgia Rheumatica (PMR)?

- First described in 1888
- Inflammatory disorder
- Pain and stiffness in neck, shoulders and pelvic girdle
Who is at risk for PMR?

- Almost exclusively in patients > 50 years
- Average age at diagnosis > 70 years
- Mainly seen in people of north European ancestry
- Women more frequently affected than men
- Relatively common: prevalence 700 per 100,000 persons > 50 years
What causes PMR?

• Cause is unknown
• Both genetic and environmental factors might be important
• Familial association rare
• Seasonal variation and differences in geographical distribution - ? possible environmental trigger
PMR - Presentation

• In some cases, onset can be sudden and dramatic
• In other cases, onset is gradual over a few weeks
• Presence of inflammation in body

➢ Blood tests: raised erythrocyte sedimentation rate (ESR) and/or C reactive protein (CRP)
How is PMR diagnosed?

• Clinical diagnosis - based on symptoms and examination
• No single test available for definite diagnosis
• Blood tests to confirm diagnosis and rule out other reasons for patient symptoms
• Additional tests may be ordered to exclude conditions that mimic PMR
Other conditions to consider in PMR

**Inflammatory disorders**
- Rheumatoid Arthritis
- Pseudogout (CPPD arthritis)
- Inflammatory muscle diseases (rare)

**Non inflammatory disorders**
- Frozen shoulder
- Osteoarthritis
- Infection
- Underactive thyroid
- Fibromyalgia (pain syndrome)
- Malignancy (rare)
How are PMR and Giant Cell (temporal) Arteritis (GCA) related?

- Headache, jaw pain on chewing, visual disturbance
- PMR: 2 to 3 more common than GCA
- 50% of GCA patients have PMR symptoms
- 10% of PMR patients go on to develop GCA
PMR - Treatment

• Corticosteroids are the only known effective treatment
• Anti-inflammatory may be used but usually not enough by itself
• Lack of complete response to recommended prednisone doses with atypical features – consider alternative diagnosis
PMR Treatment - steroids

• Monitor response
  ➢ Improvement in symptoms
  ➢ Symptoms and signs suggestive of alternative diagnosis
  ➢ Blood tests: CRP +/- ESR
  ➢ Side effects
PMR Treatment - steroids

- Urgency to start not necessary for PMR, unlike GCA
- Initial recommended prednisone dose 15 to 20mg daily
- If PMR, expect quick response within one week
- Expect inflammatory markers to normalise
- Prednisone dose is gradually tapered
- Most patients (33-50%) can discontinue steroids after two years
Adverse effects of steroids

• Long term and higher doses carry the most risk

- Skin changes; thinning and bruising
- Body composition changes; weight gain, fluid retention
- Cataracts/Glaucoma
- Irritation of stomach
- Osteoporosis
- Diabetes
- Cardiovascular disorders (high blood pressure, high lipids/cholesterol)
- Mood changes
Adverse events during long term steroid treatment in PMR (n = 222)

- 43% had at least one adverse event after being on steroids continuously for 2.5 years
- Osteoporosis (25%)
- Fracture from osteoporosis (14%)
- High blood pressure (12%)
- Diabetes (5%)
- Heart attack (4%)

Mazzantini M et al J Rheumatol 2012
PMR Treatment – steroids

• Never abruptly stop taking prednisone
• Doses may need to be increased when body is under stress e.g. after surgery, during illness
• Symptoms may recur when prednisone is reduced to lower doses
  ➢ Higher starting dose followed by rapid tapering rate – greatest risk of relapse
PMR Treatment – steroids

- 25-50% PMR patients relapse at some point, most likely over the first 18 months of treatment
- Increase in aches and pains during treatment may not necessarily mean a PMR flare
- Relapse can occur even after treatment discontinued
- 256 PMR patients – 40% relapses occurred 6 months after treatment stopped

Bogliolo Ann Rheum Dis 2009
PMR - Treatment

• Most PMR patients are diagnosed and managed by GPs
• Refer to rheumatologist if
  ➢ Atypical features
  ➢ Lack of response to therapy
Prevention and treatment of steroid induced osteoporosis

• Consider bone density scan
• Vitamin D supplements recommended
• Calcium supplements – dietary intake alone will suffice
• For high risk patients (reduced bone density or fragility fracture)
  ➢ Bisphosphonates (Fosamax/IV Aclasta) recommended
PMR Treatment
Steroid sparing agents
(not routinely used)

• Methotrexate

• Leflunomide (Arava)

Adizie T et al Int J Clin Pract 2012
What can I do?

• Balance activity and rest
• Exercise regularly (low impact) – physiotherapy is useful
• Nutrition – calcium in diet, healthy balanced diet
• Relaxation techniques
• Seek support
PMR – Update
American Rheumatology Conference Oct 2013

• PMR: Prioritisation of outcome measures – perspective of patients, GP and rheumatologists

• General symptoms (fatigue, malaise, sleep disturbance, depression/anxiety), functional status and quality of life more relevant to patients with PMR compared to rheumatologists

Singh Y et al Abstract #1662
<table>
<thead>
<tr>
<th>Clinical criteria for scoring algorithm*</th>
<th>Points</th>
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</thead>
<tbody>
<tr>
<td>Morning stiffness lasting more than 45 min</td>
<td>2</td>
</tr>
<tr>
<td>Hip pain or restricted range of motion</td>
<td>1</td>
</tr>
<tr>
<td>Absence of rheumatoid factor and antibody to cyclic citrullinated peptide</td>
<td>2</td>
</tr>
<tr>
<td>Absence of other joint involvement</td>
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</table>

<table>
<thead>
<tr>
<th>Ultrasound criteria for scoring algorithm*</th>
<th>Points</th>
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<tr>
<td>At least one shoulder with subdeltoid bursitis, biceps tenosynovitis, or glenohumeral synovitis; and at least one hip with synovitis or trochanteric bursitis</td>
<td>1</td>
</tr>
<tr>
<td>Both shoulders with subdeltoid bursitis, biceps tenosynovitis, or glenohumeral synovitis</td>
<td>1</td>
</tr>
</tbody>
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What is the outlook?

- Most patients with PMR lead productive active lives
- Duration of drug treatment differs for each patient
- In PMR, no increased risk of cardiovascular disease with corticosteroid treatment
  
  Maradit-Kremers Arthritis Rheum 2007

- Survival similar to general population
  
  Salvarani Arthritis Rheum 1995
  Gran Rheumatology 2001
PMR - Summary Points

• Common
• Affects the elderly
• Accurate diagnosis is key
• Early specialist opinion recommended if diagnosis uncertain
• Treatment (holistic approach) – emphasis: prevention and management of side effects of therapy
• Excellent long term prognosis - most resolve in 2-5 years