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# Polymyalgia rheumatica

## Diagnosis and management

### Background

Polymyalgia rheumatica (PMR) is a common inflammatory rheumatic disease and an indication for long term treatment with oral steroids. Its incidence rises progressively beyond the age of 50 years. For the most part, PMR is managed in primary care.

### Objective

This article highlights the main points in the British Society for Rheumatology and the British Health Professionals in Rheumatology guidelines that may be useful to general practitioners in the primary care setting.

### Discussion

Different levels of awareness of the condition between practitioners, and a lack of uniform diagnostic criteria may impede correct diagnosis and management of PMR. Updated international guidelines produced by the British Society for Rheumatology and the British Health Professionals in Rheumatology can aid diagnosis and direct treatment and disease monitoring.

**Keywords:** polymyalgia rheumatica; general practice; primary care

There are occasional familial cases and there is a clear north to south gradient with the highest prevalence in Scandinavian countries and in parts of the USA that have a population of a similar ethnic background, a lower prevalence in Mediterranean countries and the lowest prevalence in Arabian and Asian countries.<sup>7</sup> Studies report a cyclical yearly pattern of incidence and seasonal variations in incidence, which might suggest an environmental infectious aetiology.<sup>7</sup> Polymyalgia rheumatica is twice as common in women as in men and is related to giant cell arteritis (GCA), although the precise relationship between the two conditions remains unknown. Polymyalgia rheumatica is 2–3 times more common than GCA.<sup>7</sup>

### Clinical features

Polymyalgia rheumatica affects the shoulder and hip girdles causing aching and morning stiffness related to synovitis of proximal joints and inflammation of extra-articular synovial structures.<sup>8</sup> Patients may complain of difficulty getting out of bed or standing from the seated position. Typically, morning stiffness lasts for 30 minutes or more. The onset is often abrupt and symptoms are usually symmetrical. Pain can also involve the neck, upper arms, lower back and thighs. Distal musculoskeletal manifestations may include carpal tunnel syndrome and nonerosive, asymmetrical peripheral arthritis (affecting the knees and small joints of the hands and feet) and are seen in about half of patients.<sup>9</sup> Atypical presentations involving asymmetrical proximal joint symptoms or younger patients may occasionally occur.

Examination findings may be minimal and include painful restriction of active and passive movements of the shoulder and hip joints, usually without detectable proximal joint swelling. Muscle strength is usually normal, although interpretation may be difficult because of pain.

Polymyalgia rheumatica (PMR) is a common rheumatic disease that affects patients middle aged and older. Its incidence increases progressively beyond the age of 50 years.<sup>1</sup> The reported annual incidence in Europe and the United States of America varies between 1.3 and 11.3 per 10 000 individuals aged over 50 years.<sup>2–5</sup> This wide variation may reflect differing levels of awareness of the condition between practitioners, or a lack of uniform criteria used to make the diagnosis. A United Kingdom study<sup>6</sup> demonstrated that general practitioners do not always use established criteria to diagnose PMR. This may result in unnecessary further investigation and needlessly expose patients to the risks associated with long term steroid use.

While the aetiology of PMR remains elusive, both environmental and genetic risk factors are thought to contribute to its development.<sup>7</sup>

Systemic signs and symptoms such as low grade fever, depression, fatigue, anorexia, and weight loss may occur in up to 40% of patients.<sup>7</sup>

## Diagnosis

There are no specific diagnostic tests for polymyalgia rheumatica. The British Society for Rheumatology and the British Health Professionals in Rheumatology have published joint guidelines on the diagnosis and management of polymyalgia rheumatica.<sup>9</sup> Good scientific evidence is currently lacking and most of the recommendations for diagnosis and treatment included in these guidelines are graded at level B or C. The guidelines recommend a stepped diagnostic and management process with diagnosis involving identification of core inclusion criteria and exclusion of conditions that may mimic PMR – exclusion criteria (Table 1, 2). Inclusion and exclusion criteria should be documented in the patient’s medical record.

### Suspected giant cell arteritis

Regardless of the presence of PMR, if the acute phase reactants are elevated and this is associated with new headache or an unexplained pain located above the neck, it is important to consider a diagnosis of GCA. Features of this condition include headache, jaw claudication and visual symptoms. If GCA is suspected, commence treatment with high dose steroids immediately and refer the patient to a hospital emergency department without delay.<sup>10</sup> The Therapeutic Guidelines: rheumatology<sup>13</sup> suggest steroids and urgent ophthalmology referral.

## Investigations

So long as there are no features that suggest GCA, urgent institution of steroid therapy is not necessary, and treatment can be delayed to enable further investigation. Suggested initial laboratory investigations in PMR are listed in Table 3.

Laboratory test findings in PMR are nonspecific and usually indicate the presence of inflammation with elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). However, the presence of PMR can be established in the setting of a normal ESR if there is a classic clinical picture and a good response to steroids; this occurs in up to one-fifth of

**Table 1. Polymyalgia rheumatica: core inclusion criteria (all these criteria are required for a diagnosis)**

- Bilateral shoulder or pelvic girdle aching – or both
- Morning stiffness longer than 45 minutes
- Age older than 50 years
- Duration >2 weeks
- Evidence of an acute phase response (raised ESR/CRP)

**Table 2. Conditions that may mimic polymyalgia rheumatica**

- Active cancer (eg. multiple myeloma, lymphoma, leukaemia)
- Infections: viral or bacterial
- Giant cell arteritis
- Rheumatic diseases
  - late onset rheumatoid arthritis (symmetrical peripheral synovitis, positive rheumatoid factor and anticyclic citrullinated peptide antibodies, joint erosions, extra-articular manifestations)
  - remitting seronegative symmetrical synovitis with pitting oedema (RS3PE syndrome)
  - late onset spondyloarthritis (peripheral enthesitis, dactylitis, anterior uveitis, HLA-B27, and radiological evidence of sacroiliitis)
  - late onset systemic lupus erythematosus, other connective disease
  - polymyositis (proximal muscular weakness rather than pain, increased muscle with or without creatine kinase, myopathic electromyogram changes)
  - pseudogout
  - fibromyalgia (younger, absence of typical joint stiffness, normal inflammatory markers)
- Hypothyroidism
- Drug induced (eg. statin myopathy)
- Chronic pain syndrome (osteoarthritis of neck or shoulder, fibromyalgia)
- Local shoulder or hip pathology

Adapted from Gonzalez-Gay et al<sup>10</sup>

cases (range 7–20%).<sup>11</sup> C-reactive protein is a more sensitive indicator of disease activity than ESR in PMR as it is less affected by extraneous factors such as increasing age.<sup>11</sup> Patients in whom evidence of inflammation on blood testing is equivocal should be referred for specialist assessment.<sup>9</sup> Patients with PMR may have a normochromic, normocytic anaemia and raised alkaline phosphatase but creatinine kinase is invariably normal (in contrast to elevated levels in polymyositis, hypothyroidism and other muscle diseases).

The extent of further investigations will depend on the level of certainty of the diagnosis. Secondary investigations may include autoantibody screening, muscle biopsy and

imaging (eg. scintigraphy, magnetic resonance imaging, ultrasonography) to detect synovitis in proximal joints and periarticular structures.

## Management

### Corticosteroids

Once the diagnosis is established, the patient should be commenced on low dose steroid therapy. The dose is then gradually tapered over time. It is important to counsel the patient about potential risk factors associated with long term steroid therapy (Table 4).

Osteoporotic risk with long term steroid therapy can be minimised by using the minimum effective steroid dose. A recent meta-analysis

**Table 3. Suggested initial laboratory investigation in polymyalgia rheumatica**

- Full blood count
- ESR/plasma viscosity and/or CRP
- Urea and electrolytes
- Liver function tests
- Calcium, phosphate, albumin, alkaline phosphatase
- Protein electrophoresis (also consider urinary Bence Jones protein)
- Thyroid stimulating hormone
- Creatine kinase
- Rheumatoid factor (antinuclear antibody and anticyclic citrullinated peptide antibodies may be considered)
- Dipstick urinalysis

showed that starting prednisolone doses of 15 mg/day or lower was associated with lower cumulative steroid dosages than higher starting doses and that higher doses of corticosteroids were associated with more adverse effects.<sup>12</sup> In addition, higher doses may mask those conditions whose symptoms mimic PMR (*Table 2*).

There is no consistent evidence for an ideal steroid regimen that is suitable for all patients. Therefore, the approach to treatment must be flexible and tailored to the individual. Dose adjustment may be required for disease severity, comorbidity, side effects and patient wishes. The British Society for Rheumatology and the British Health Professionals in Rheumatology consensus regimen suggests using:<sup>9</sup>

- daily prednisolone: 15 mg for 3 weeks
- then 12.5 mg for 3 weeks
- then 10 mg for 4–6 weeks
- then reduction by 1 mg every 4–8 weeks or alternate day reductions (eg. 10/7.5 mg alternate days).

The Therapeutic Guidelines rheumatology group suggest a similar regimen:<sup>13</sup>

- 10–20 mg orally, daily in the morning initially for 2–4 weeks
- then reduce the daily dose by 2.5 mg every 2–4 weeks until the daily dose is less than 10 mg
- then decrease the daily dose by 1 mg every 4–8 weeks.

Intramuscular methylprednisolone (intramuscular depomedrone) may be used in milder cases and may reduce the risk of steroid related complications. Initial dose is 120 mg every 3–4 weeks, reducing by 20 mg every 2–3 months.<sup>14</sup>

The addition of nonsteroidal anti-inflammatory drugs to glucocorticoid regimens for the treatment of patients with PMR has shown no advantage over steroids alone in terms of duration of therapy or daily or cumulative prednisone doses, but produced more adverse events.<sup>15</sup> In specialist settings, glucocorticoid sparing agents such as methotrexate and other biological and nonbiological agents may be considered.

**Bone protection**

The decision about the use of a bone sparing agent (eg. oral biphosphonate) for bone protection when initiating steroids for PMR depends on the clinical risk of developing glucocorticoid induced osteoporosis. A recent review suggests that osteoporotic risk in PMR varies widely, from 3.6–27%, and from 58–91%.<sup>12</sup> However, all individuals with PMR should commence calcium and vitamin D supplementation and receive advice about weight bearing exercise. In patients deemed at high fracture risk clinically (eg. prior fragility fracture, high steroid dose) or following dual energy X-ray absorptiometry (DXA) scan, a bone sparing agent (eg. oral biphosphonate) should be considered. Medicare rebates are available for patients having DXA scan who are currently on prolonged glucocorticoid therapy (such as equivalent to or greater than 7.5 mg prednisolone daily for a period anticipated to last for at least 4 months).<sup>16</sup> Some bisphosphonates are available on the Pharmaceutical Benefits Scheme for: 'Treatment as the sole PBS subsidised anti-resorptive agent for corticosteroid induced osteoporosis in a patient currently on long term (at least 3 months), high dose (at least 7.5 mg per day prednisolone or equivalent) corticosteroid therapy with a bone mineral density (BMD) T-score of –1.5 or less.'<sup>17</sup>

**Follow up and monitoring**

Early follow up is vital to evaluate response to initial therapy and to confirm the diagnosis.<sup>18</sup> A patient reported global improvement of ≥70% within 1 week of commencing steroids is

**Table 4. Risks of long term steroid therapy**

- Osteoporosis
- Weight gain
- Skin bruising
- Raised blood pressure
- Impaired blood glucose
- Depression
- Cardiac failure
- Increased risk of infection
- Cataracts
- Glaucoma
- Cushing syndrome

**Table 5. Indications for referral in polymyalgia rheumatica**

**Atypical features**

- Age younger than 60 years
- Chronic onset more than 2 months
- Lack of shoulder involvement
- Lack of inflammatory stiffness
- Prominent systemic features, weight loss, night pain, neurological signs
- Features of other rheumatic disease
- Normal or extremely high acute phase response

**Treatment dilemmas**

- Incomplete, poorly sustained or nonresponse to corticosteroids
- Inability to reduce corticosteroids
- Contraindications to corticosteroid therapy
- Prolonged corticosteroid therapy (more than 2 years)

consistent with a diagnosis of PMR. Inflammatory markers usually normalise within 4 weeks. A lesser response should prompt a search for an alternative condition. If a relapse of PMR is suspected as the steroid dose is being tapered, the steroid dose can be increased to the previously effective dose level and dose reduction recommenced from that level.<sup>13</sup>

Each follow up encounter with the GP should try to explore the following:

- response to treatment – degree of reduction in proximal pain, fatigue and morning stiffness
- complications of disease including symptoms of GCA (eg. headaches, jaw claudication and large vessel disease)
- steroid related adverse events
- atypical features or other mimicking conditions
- laboratory monitoring (full blood count, ESR/CRP, urea and electrolytes, glucose) every 4–6 weeks.

Duration of treatment can vary from 1–3 years, and some patients will require small doses of steroids beyond 3 years.<sup>7</sup> Slow prednisolone dose tapering (<1 mg/month) is important as slower tapering regimens are associated with fewer relapses and earlier glucocorticoid treatment cessation than faster tapering regimens.<sup>12</sup> Steroids may be stopped when the patient is asymptomatic from their inflammatory symptoms. Isolated raised ESR or CRP is not an indication for continuing steroid therapy but may require further investigation and referral.

### When to refer?

The British Society for Rheumatology and the British Health Professionals in Rheumatology guidelines advise early specialist referral in cases with atypical features or when treatment dilemmas arise (grade C recommendation) (Table 5).<sup>9</sup>

### Summary

- Polymyalgia rheumatica is a chronic relapsing disease, managed for the most part in primary care. The extended use of steroids in this population group places extra emphasis on accurate diagnosis, safe management, ongoing monitoring of disease activity and prevention of complications.

- Suspect PMR in the elderly patient with bilateral shoulder ache and stiffness.
- Document inclusion and exclusion criteria in the medical record.
- Use the minimum effective steroid dose initially (15 mg/day prednisolone or less).
- Taper steroids gradually (<1 mg/month); relapses are common.
- Assess the need for bone sparing agents and institute osteoporosis prophylaxis early, if required.
- Atypical features and treatment dilemmas warrant referral.

### Resource

The stepped diagnostic and management process recommended by the British Society for Rheumatology and the British Health Professionals in Rheumatology is available at <http://rheumatology.oxfordjournals.org/content/49/1/186/F2.large.jpg>.

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