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Diagnosis and management of polymyalgia rheumatica

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INTRODUCTION

Polymyalgia rheumatica (PMR) is the commonest inflammatory rheumatic disorder affecting older people.[1] Patients typically present with bilateral shoulder pain, morning stiffness, raised inflammatory markers, and have a rapid response to low-dose corticosteroids. There is no gold standard diagnostic test and despite being first described in 1888, controversies still exist as to its defining characteristics. PMR carries a lifetime risk of 2.4% for females and 1.7% for males.[2] The incidence in the UK has been shown to be 8.42 per 10 000 person years.[3] In the UK, the majority of patients are managed exclusively in primary care[4] with an average full-time GP seeing five new cases of PMR per year.[5] Accurate diagnosis can be challenging even for specialists, but is essential as many serious illnesses can mimic PMR. Guidelines for the diagnosis and management of PMR have recently been published by the British Society of Rheumatologists (BSR) and British Health Professionals in Rheumatology (BHPR).[6]

DIAGNOSIS

Consider PMR in patients over the age of 50 years with:

- ≥2 weeks of bilateral shoulder and/or pelvic girdle ache;
- morning stiffness; and
- raised inflammatory markers.

Subsequent clinical assessment and investigations should be directed towards excluding disorders that can mimic PMR (Box 1). Suggested initial investigations include full blood count, renal, thyroid, and liver function, inflammatory markers (erythrocyte sedimentation rate [ESR]/C-reactive protein [CRP]), bone, protein electrophoresis, rheumatoid factor, urinary Bence Jones protein, creatinine kinase, and dipstick urinalysis. Additional investigations if clinically appropriate include antinuclear antibodies, anti-cyclic citrullinated peptide antibodies, and chest X-ray. Ultrasound of the shoulders and/or hips may show characteristic lesions such as sub-deltoid bursitis, bicipital tenosynovitis, and joint fluid.[7]

Giant cell arteritis (GCA) is a serious association of PMR. The latest guidance is summarised in an associated article.[8]

TREATMENT

A rapid response to low dose prednisolone (15 mg) is typical. However, a poor response should prompt further assessment for an alternative diagnosis or consideration for a specialist review. Patients taking long-term

Box 1. Disorders that can mimic polymyalgia rheumatica

Rheumatological disorders

Inflammatory

- Late-onset rheumatoid arthritis, spondylo-arthritides, psoriatic arthritis, systemic lupus erythematosus, scleroderma, Sjögren's syndrome, vasculitis, inflammatory myopathies

Non-inflammatory

- Osteoarthritis, rotator cuff disorders, frozen shoulder

Infection

- Tuberculosis, bacterial endocarditis, osteomyelitis, septic arthritis, other infections, for example, urinary tract infections

Malignancies

- Lymphoma, myeloma, and leukaemia. Solid tumours, and metastases, for example, prostate, bowel, lung, breast, and renal

Other

- Endocrine disorders (for example, hypo/hyperthyroidism, hyper/hypoparathyroidism)
- Drug induced myalgia (for example, statins)
- Parkinson's disease

corticosteroids are at high risk of developing osteoporosis. Current guidance suggests offering osteoporosis prophylaxis (bisphosphonate and calcium/vitamin D supplementation) to those who are at high risk of fracture (≥65 years or prior fragility fracture).[6] In other individuals calcium/vitamin D supplementation and a dual-emission X-ray absorptiometry scan is recommended.[6] Although not part of this latest guidance, gastric symptoms, are commonly reported among patients with PMR.[9] Gastric protection should therefore be strongly considered especially in at-risk patients.

ONGOING MANAGEMENT

Robust clinical evidence for corticosteroid tapering is lacking. An initial dose of 15 mg of prednisolone coupled with a slow reduction in dose is effective at maintaining remission.[10] Guidance suggests 15 mg of prednisolone for 3 weeks, followed by 12.5 mg for 3 weeks, then 10 mg for 4–6 weeks, and finally a reduction in dose of 1 mg every 4–8 weeks. After initial diagnosis, follow-up to assess response within 1 week is suggested. Subsequently a review of symptoms, progress, adverse side effects, complications of treatment, atypical features, and an assessment for GCA is suggested in weeks 3 and 6 and again, 3, 6, 9, and 12 months after diagnosis. This treatment and follow-up regimen serves only as a guide and should be modified according to individual patients' response and ongoing progress. Relapses should be assessed by clinical symptoms rather than being guided by laboratory results (such as ESR and CRP). Management of relapse should involve an increase of prednisolone to the previously higher dose that controlled symptoms, followed by reassessment.[6] Recurrent relapses (more than two) are an indication for specialist referral for consideration of steroid sparing agents such as methotrexate.

PATIENT EDUCATION AND SELF-MANAGEMENT

All patients should be provided with written information on PMR and corticosteroid treatments. They should also be given information on range-of-motion exercises for the shoulder and provided with contacts to their local Polymyalgia Rheumatica & Giant Cell Arteritis UK patient support group.[11]

REFERRAL

A wide range of illnesses can mimic PMR, some of which respond to corticosteroid therapy. Accurate diagnosis is therefore essential, ongoing management can usually continue in primary care once diagnosis has been confirmed. In cases of diagnostic uncertainty early referral for specialist review is essential. Some examples of indications for early referral are summarised in Box 2.

CONCLUSION

PMR is a commonly-seen disorder that is often managed exclusively in primary care. The BSR/BHPR guidance brings together the best evidence and extensive expert opinion to provide a much needed safe approach to the identification and ongoing management of this common inflammatory rheumatological disorder. Accurate diagnosis is key. Further early expert review should be sought in situations where diagnosis is uncertain. The guidance reinforces a more holistic approach to PMR emphasising the need to consider the prevention and management of potential side effects and complications of treatment. Dissemination of this guidance to general practice, where the majority of patients are managed, will hopefully facilitate accurate diagnosis and improve the ongoing management and, therefore, outcomes for patients with PMR.

Box 2. Indications for early referral to specialist

Atypical features

- Age <60 years
- Chronic onset
- Lack of shoulder involvement
- Lack of inflammatory stiffness
- Red flag features (prominent systemic features, weight loss, night pain, neurological signs)
- Features of peripheral arthritis, muscle disease, and other autoimmune/systemic diseases
- Very high or normal inflammatory markers

Treatment dilemmas

- Poor/incomplete response to corticosteroids
- Inability to reduce corticosteroid therapy
- Recurrent relapse
- Contraindications to corticosteroid therapy
- Prolonged treatment duration (>2 years)

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Provenance

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