Arthritis Research UK

Practical advice for GPs on management of rheumatic disease

Hands On

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Polymyalgia rheumatica

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Editorial

Polymyalgia rheumatica (PMR) is a condition that is commonly seen in older patients in primary care. It is known that there is wide variation in clinical practice with respect to diagnosis and management. The challenges include having no gold standard test for it, the possibility of atypical presentation and the existence of other conditions that can mimic it.

Having made the diagnosis it is important to balance treatment efficacy against potential side-effects. Patients vary in their response to steroids and the rate at which their treatment can be tapered.

Patients with PMR are likely to already have comorbidities or to be at risk of developing them due to steroid treatment. Primary care has a key role in screening and monitoring for hypertension, diabetes and bone health. Again it is known that there is often room for improvement here.

The authors of this report address these challenges with a very practical and useful guide to how assessment and management can be improved. This is definitely something to keep to hand as you see patients with PMR.

Simon Somerville

Introduction

Polymyalgia rheumatica (PMR) is the commonest inflammatory rheumatological disorder of older people, with an incidence of 8.4/10,000 person-years (95% CI 8.3 to 8.6) and a lifetime risk of 2.4% for women and 1.7% for men.[1,2] It is characterised by bilateral pain and stiffness of the hips and shoulders and is often associated with profound disability. The majority of patients with PMR are exclusively managed in the community, yet diagnosis can be difficult, especially for those with an atypical presentation. A recent analysis of GP consultation databases suggests that current primary care management may be sub-optimal and that patient care could be improved.[3] The aim of this edition of Hands On is to provide an evidence-based overview to the successful diagnosis and management of patients with PMR in general practice settings.

Making the diagnosis

PMR is uncommon in those under 60 years. For many patients, the onset of PMR is abrupt and may start with fevers or chills ('the flu that does not go away'). Patients complain of pain in the shoulders and hips that is associated with stiffness, especially in the morning. They often report that they rapidly deteriorate over a period of 1–2 weeks, becoming so disabled that they

are no longer able to get off the toilet without help or to turn over in bed. Patients usually have elevated inflammatory markers (e.g. ESR, CRP or where available plasma viscosity, PV) and may report systemic features such as malaise and fatigue. Treatment with low-dose glucocorticoids (e.g. prednisolone) produces a dramatic response in around 80–90% of patients.

The lack of a 'gold standard' (100% specific) diagnostic test makes diagnosing PMR challenging even for experts.[4] As such a thorough and systematic diagnostic work-up is essential in primary care to exclude other conditions that commonly present with a polymyalgic syndrome. These commonly include both rheumatological and non-rheumatological disorders, some of which may initially improve with glucocorticoid treatment, and so response to treatment is not diagnostic of PMR.[4] Therefore patients who do not have a rapid, complete response (see below for definition) warrant re-consideration of the diagnosis.

A number of 'core' investigations are recommended by the British Society for Rheumatology.[5] These are intended to help guide the diagnostic process and are presented in Box 1.

BOX 1. Recommended baseline investigations.

Blood tests

- Full blood count
- Inflammatory markers – e.g. ESR, CRP, PV
- Renal function
- Liver function
- Calcium and alkaline phosphatase
- Rheumatoid factor
- Thyroid function
- Glucose
- Protein electrophoresis
- Creatinine kinase

Urine tests

- Urine dip stick – e.g. glucose, blood, protein, nitrites
- Bence Jones protein

Specialist imaging such as musculoskeletal ultrasound of the shoulders and hips is now used in some hospitals to aid the diagnosis of rheumatic disease; however, its role in primary care remains to be defined. In some patients a chest x-ray is indicated if respiratory pathology is suspected or if the patient has prominent systemic symptoms.

TABLE 1. Conditions that can mimic PMR.

Category	Example
Inflammatory rheumatological disorders	Rheumatoid arthritis Giant cell arteritis Spondyloarthropathy Crystal arthropathy
Non-inflammatory rheumatological disorders	Osteoarthritis Shoulder pathology (e.g. frozen shoulder, rotator cuff disease) Fibromyalgia
Infection	Bacterial endocarditis, osteomyelitis, septic arthritis, tuberculosis and other infections
Malignancy	Leukaemia, lymphoma, myeloma Solid tumours (including prostate, renal, lung)
Endocrine	Diabetes Hypo/hyperthyroidism Hypo/hyperparathyroidism
Other disorders	Drug-induced (e.g. statins) Motor neurone disease Parkinson's disease

NB: This list illustrates some conditions to consider when evaluating patients with suspected PMR and is not intended to be exhaustive.

TABLE 2. Diagnosing PMR: the spectrum of classical vs atypical PMR.

	History, examination, investigations	Trial of glucocorticoids: dose	Response to treatment
A safe diagnosis of PMR in primary care can be made in presence of ALL THREE of these	Classical clinical features	15 mg prednisolone	'Magic', 'miracle', within 3 days
Consider other diagnoses if ANY of these are present (but can be 'atypical PMR')	Atypical features	Need for >15 mg prednisolone to relieve symptoms	Incomplete or delayed response

How to treat and monitor PMR

For most patients with PMR the mainstay of treatment is with glucocorticoids, usually oral prednisolone, although there is limited trial evidence to support the use of injectable glucocorticoids (such as intramuscular methylprednisolone) in patients with mild or localised symptoms. The mechanism of action of glucocorticoids in PMR is not fully understood, but high doses (30 mg prednisolone or more) should not be required. There is no role for the routine use of non-steroidal anti-inflammatory drugs.

The response to the initial glucocorticoid treatment could be viewed as an (admittedly imperfect) diagnostic test for PMR. This 'test' is most specific for PMR as patients feel completely better ('magic' or 'miracle' effects) after 3 days of 15 mg prednisolone. Sensitivity of the 'trial of steroids' is probably improved, at the cost of some loss in specificity, if patients are allowed longer (1–2 weeks) to achieve a 70% reduction in symptom scores, or if they are given higher doses (e.g. 20–25 mg prednisolone).

After the initial response, the glucocorticoid dose is tapered gradually. The average length of treatment in hospital-based cohorts is around 2 years but with wide variation. There is little evidence to help decide how to taper the dose. Some patients need a much slower taper than others, and some patients develop significant glucocorticoid toxicity. To reduce the risks of treatment, it is usually recommended to try the quicker taper first, but to slow this taper down if need be to keep the PMR symptoms under control. Individualised treatment and shared decision-making should be the rule rather than the exception.

The aim of PMR treatment is to achieve acceptable control of PMR symptoms while minimising the risks and side-effects of treatment. Therefore, if a patient feels their PMR is well controlled, there is no need to re-check inflammatory markers before reducing the dose. A transient (<1 week) increase in PMR-like symptoms after dose reduction is common and usually manageable if patients are forewarned.

A key role of the GP is to regularly monitor patients, checking for alternative diagnoses and assessing the risks and side-effects of glucocorticoids, which are common in PMR[6] and are a major consideration when making decisions on tapering rates. Risks and side-effects such as weight gain, skin fragility, changes in physical appearance, infections, glaucoma, steroid myopathy, osteoporosis/fracture, avascular necrosis, hypertension, diabetes, psychiatric morbidity, and peptic ulcers should all be considered as appropriate for each

BOX 2. Suggested steroid therapy regimen[4]
(Reproduced with permission from: B Dasgupta and Oxford University Press).

- 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 (e.g. 10/7.5 mg alternate days, etc.)

BOX 3. Items to potentially include in a PMR review.

- Discussion around the diagnosis, management, course and prognosis of PMR
- Provide written information on PMR
- Provide written information on GCA, including red flag signs
- Osteoporosis risk assessment
- Blood pressure monitoring
- Diabetes assessment
- Advice on keeping physically active
- Assessment of potential impact of glucocorticoids on comorbid conditions

individual patient. Consider adding calcium, vitamin D and perhaps bisphosphonates according to local guidelines. It may be wise to monitor blood glucose and blood pressure intermittently. Patients should be offered a 'steroid card' and access to support and information about their condition.

Rheumatologists often see the atypical cases, those with incomplete glucocorticoid response and those with difficulty in stopping glucocorticoids. Some rheumatologists use methotrexate or other drugs but most of the evidence comes from rheumatoid arthritis rather than PMR. If used, methotrexate also requires monitoring for potential toxicity.

Non-pharmacological treatments have not been formally evaluated although many patients self-manage pain and stiffness with heat packs and simple analgesia. Whilst physiotherapy interventions have not yet been formally investigated the maintenance of joint ranges around the shoulders and hips with gentle exercise is prudent and patients anecdotally report improvements in stiffness and pain as well as function. Additional strengthening exercises can be added to a daily programme to maximise general activities of daily living and mobility. Effective physiotherapy exercise can be enhanced by general advice relating to keeping active, optimal posture, diet, the use of heat, minimising the risk of falls, pacing strategies as well as being alert to headaches or other potential related symptoms.

Patient education forms an essential part of management for patients with PMR. Written information should be provided giving details of the natural history of the condition, along with information on treatment, side-effects and 'red flags' (including giant cell arteritis). In addition to online resources provided by Arthritis Research UK (http://www.arthritisresearchuk.org/arthritis-information/conditions/polymyalgia-rheumatica.aspx) PMRGCAuk (http://www.pmrgca.co.uk) is a registered charity that provides a range of services, including a telephone helpline, for patients and their families.

Do not miss giant cell arteritis

Perhaps 5–10% of patients with PMR are also diagnosed with giant cell arteritis (GCA); in some cases the GCA only appears later.[1] Untreated GCA can result in permanent visual loss or stroke, and as such is a 'must not miss' diagnosis. Tell patients with PMR to look out for headache, scalp tenderness, jaw pain/claudication and visual disturbance. GCA symptoms may need high glucocorticoid doses (usually at least 30–40 mg/day prednisolone) so the risk of steroid-associated side-effects is high. Patients suspected as having GCA should be urgently referred to local specialist services (usually rheumatology or ophthalmology, but this is dependent on local care pathways).

Specialist referral

Many patients with suspected PMR can be safely diagnosed and managed in general practice. Referral is usually indicated for one of two reasons: diagnostic uncertainty and lack of response to primary care treatment.

The British Society for Rheumatology recommends referral in the following situations:[4]

- younger age (usually less than 60 years)
- no shoulder involvement
- lack of inflammatory stiffness
- insidious onset
- normal or very high inflammatory markers
- red flag features (e.g. prominent systemic features, weight loss, night pain, neurological signs)
- suspicion of co-existing giant cell arteritis
- poor or incomplete response to glucocorticoids
- difficulty reducing steroids dose
- recurrent relapse
- contraindication to glucocorticoid treatment.

Key messages

- Do not start treatment with glucocorticoids until appropriate investigations have been completed to rule out other disorders.
- Do not miss giant cell arteritis.
- The majority of patients with PMR can be safely managed in primary care but GPs should have a low threshold for referral in cases of diagnostic uncertainty and lack of treatment success.
- Patient education is essential and should include information on potential treatment side-effects and on the symptoms of giant cell arteritis.
- Assess for comorbidities and for potential treatment risks and side effects during each patient encounter.

Continuing professional development (CPD) task

- Identify patients in the practice with a clinical diagnosis of PMR. Follow them up to see if they go on to get an alternative diagnosis (e.g. cancer, rheumatoid arthritis). If their diagnosis changes reflect on what could have been differently to ensure an accurate original diagnosis.
- Perform an audit to assess the proportion of patients with PMR who have received an osteoporosis risk assessment.
- Review your practice to assess the proportion of patients with a steroid card, having blood pressure and glucose checks, monitoring of common glucocorticoid side-effects.

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Further reading and useful resources

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Patient resources

PMRGCAUK: www.pmrgca.co.uk

Arthritis Research UK: http://www.arthritisresearchuk.org/arthritis-information/conditions/polymyalgia-rheumatica.aspx

Assessing and managing patients with joint pain

Practice nurses and others working in primary care may be interested in a NEW online degree module, 'Assessing and managing patients with joint pain'. The course has been developed by Education for Health and Arthritis Research UK, and is accredited by The Open University.

The six-month module is delivered using the latest eLearning features, combined with two study days, and backed up by email/phone support from clinical and student support staff at Education for Health.

The course will allow practice nurses, nurse practitioners and community nurses to increase their knowledge, skills and confidence to offer help and support to people with musculoskeletal conditions. A limited number of bursaries funded by Arthritis Research UK are available directly from Education for Health to reduce the cost of enrolment.

Find out more at: **http://www.arthritisresearchuk.org/efhcourse**

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