

Diagnosis and management of polymyalgia rheumatica

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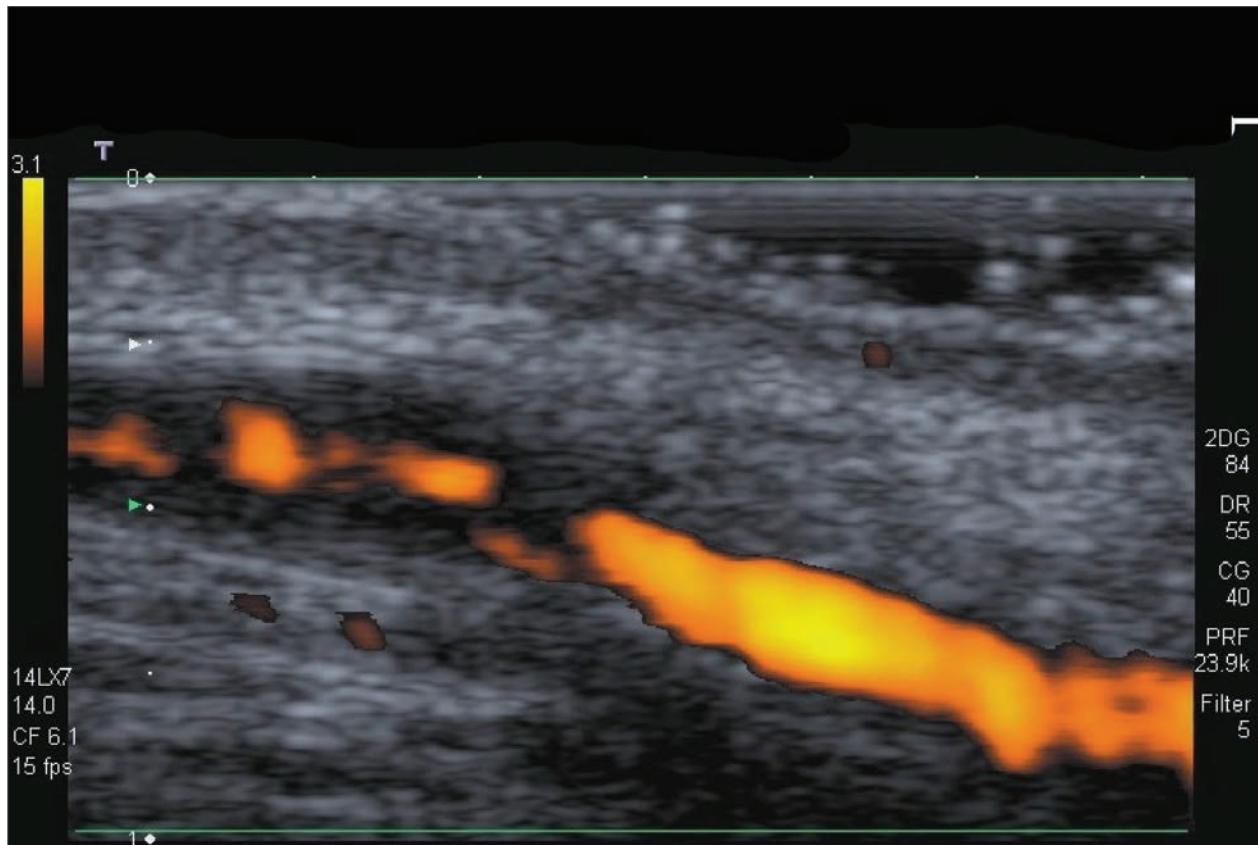
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FIGURE 1

Ultrasound halo sign in giant cell arteritis. In polymyalgia rheumatica always consider whether giant cell arteritis may be present



What are the presenting symptoms?

How should diagnosis be confirmed?

Which patients should be referred?



POLYMYALGIA RHEUMATICA (PMR) IS A COMMON CONDITION IN THE OLDER POPULATION.

The prevalence in the over 50s is approximately 1 in 133 with a female to male ratio of 2:1. PMR is an inflammatory condition, however, the exact aetiology is still unknown. Its association with giant cell arteritis (GCA), see figure 1, above, is well recognised. Half of patients with GCA will have some PMR symptoms and up to one fifth of patients with PMR will have clinical or histological evidence of GCA. If GCA is suspected, then assessment and treatment are urgent.¹

CLINICAL PRESENTATION

PMR symptoms develop over a matter of weeks. Typically patients will

experience bilateral shoulder or pelvic girdle pain and stiffness, that is worse in the mornings. As in inflammatory joint problems the symptoms improve with activity and deteriorate with rest, but there is no joint swelling. They can be associated with other non-specific symptoms, such as low-grade fever, malaise, fatigue, low mood, poor appetite and weight loss.²

DIAGNOSIS

There is no specific diagnostic test so the skills of the physician are paramount. It is important to make a thorough assessment before starting treatment. There is a range of other conditions that can mimic PMR (see table 1, right) that need to be considered and excluded through careful history, examination and

the use of simple investigations.¹

Diagnostic criteria have been proposed and vary slightly from one

Table 1

Conditions that can present in a similar way to polymyalgia rheumatica

- Rheumatic disease in the elderly e.g. rheumatoid arthritis
- Inflammatory muscle diseases
- Thyroid disease
- Malignancy
- Infection
- Bilateral shoulder capsulitis
- Osteoarthritis
- Parkinsonism
- Depressive illness

Table 2**Core inclusion and exclusion criteria in the diagnosis of polymyalgia rheumatica****Core inclusion criteria**

- Age > 50, duration > 2 weeks
- Bilateral shoulder/pelvic girdle ache (or both)
- Morning stiffness > 45 minutes
- Evidence of acute phase response (raised ESR & CRP)

Core exclusion criteria

- Active infection
- Active cancer
- Active giant cell arteritis

authority to another.³⁻⁶ The British Society of Rheumatology (BSR) has extracted the common points from each to form a set of core inclusion and exclusion criteria (see table 2, above).⁷ These can be used to help the clinician explore the likelihood of PMR being the diagnosis. These are purely for PMR; there are separate guidelines for GCA.⁸

'A small proportion of PMR patients will have normal inflammatory markers, but the usual pattern is a commensurate rise in CRP and ESR'

Simple investigations can exclude some of the mimics quite quickly. Serum markers for inflammation will help guide the diagnosis and are helpful later when tapering treatment.

Box 1**Investigations in suspected polymyalgia rheumatica**

- Full blood count
- Urea and electrolytes
- Liver function tests
- Bone profile
- CRP
- ESR
- Creatine kinase
- TSH
- Glucose/HbA_{1c}
- Myeloma screen (immunoglobulins, serum & urine electrophoresis in addition to above)
- Urine dipstick
- If appropriate consider antibodies, such as anti-CCP, rheumatoid factor and ANA
- Consider a chest X-ray, especially in smokers

It is important to recognise that a small proportion of PMR patients will have normal inflammatory markers, but the usual pattern is a commensurate rise in CRP and ESR. If only the ESR is elevated, then the clinician should consider conditions such as Sjögren's syndrome (which can also cause fatigue, but will have sicca symptoms — dryness of the eyes and/or mouth) or myeloma. As a baseline we would recommend the following investigations in all patients with suspected PMR (see box 1, below).

EVIDENCE-BASED MANAGEMENT

Unlike GCA there is no urgency to commence treatment for PMR. It is preferable to be clear whether there is an alternative more likely diagnosis first rather than potentially muddying the waters by rushing in with steroids, as it is usual for patients to feel better on steroids initially whatever the underlying problem.

If PMR is the most likely diagnosis then we recommend commencing oral prednisolone 15 mg daily with PPI cover and adding bone protection at first follow-up if indicated (see Complications of treatment, p15). Clinical and biochemical response should then be reassessed within 2-3 weeks. The classical response to a trial of steroids is 100% improvement within 24 hours.

If there is a suboptimal response, then referral to rheumatology is recommended, unless other localising symptoms flag a more appropriate specialty, for example, change of bowel habit or haemoptysis.

Box 2**BSR recommended regimen for dose reduction of steroids**

	Prednisolone dose
First dose	15 mg daily for 3 weeks
Followed by	12.5 mg daily for 3 weeks
Followed by	10 mg daily for 4-6 weeks

Then reduce dose by 1 mg every 4-8 weeks or alternate day reductions (e.g. 10 mg/7.5 mg alternate days)

The BSR guidelines for the management of PMR suggest the following regimen for dose reduction of steroids, see box 2, below.

This schedule is only a guide. In reality, the individual patient may need longer on each dose to allow for successful reduction.

In milder cases, or to reduce the risk of steroid-associated complications intramuscular steroids may be given instead. The guidelines suggest im depot methylprednisolone (Depot-Medrone) 120 mg every 3-4 weeks, reducing by 20 mg every 2-3 months. Although some have suggested the use of NSAIDs, especially in PMR without elevated inflammatory markers, this is not evidence-based and therefore the use of NSAIDs does not feature in the guidelines.

More recent recommendations for the management of PMR from a collaboration between the European League Against Rheumatism (EULAR) and American College of Rheumatology (ACR), were published in 2015. These suggest a broader range of starting doses of prednisolone.⁹ However, UK sources such as the BSR guidelines⁷ and NICE clinical knowledge summaries¹⁰ recommend 15 mg and our collective experience is that this should be sufficient in treating PMR, unless there is an overwhelming clinical indication for a higher dose (e.g. weight > 100 kg).

If symptoms return and the inflammatory markers rise in a manner consistent with a PMR flare then the dose of steroids should be increased back to the previous dose. It does not usually require more than this.

If a patient is having difficulty tapering steroids then a steroid-sparing agent may be considered. This would usually be a DMARD, such as methotrexate or azathioprine, which needs to be instigated by a rheumatologist.¹¹⁻¹⁵

Follow-up should be regular to assess response to treatment and monitor for complications of disease and treatment. We suggest follow-up at weeks 1-3, week 6, then months 3, 6, 9 and 12

key points

SELECTED BY

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Polymyalgia rheumatica (PMR) is a common inflammatory condition of unknown aetiology with a prevalence of 1 in 133 in the over 50s, and a female to male ratio of 2:1. Symptoms develop over a matter of weeks; typically bilateral shoulder or pelvic girdle pain and stiffness, that is worse in the mornings. Associated symptoms include low-grade fever, malaise, fatigue, low mood, poor appetite, and weight loss. There is no specific diagnostic test for PMR but the usual pattern is a commensurate rise in CRP and ESR. A small proportion of PMR patients will have normal inflammatory markers.

PMR is associated with giant cell arteritis (GCA).

Half of patients with GCA will have some PMR symptoms and up to one fifth of patients with PMR will have evidence of GCA. Other conditions that can mimic PMR include: rheumatic disease in the elderly e.g. rheumatoid arthritis; inflammatory muscle diseases; thyroid disease; malignancy; infection; bilateral shoulder capsulitis; osteoarthritis, Parkinsonism and depressive illness.

Unlike GCA there is no urgency to commence

treatment for PMR. It is preferable to be clear whether there is an alternative more likely diagnosis first rather than potentially muddying the waters by rushing in with steroids. If PMR is the most likely diagnosis commence oral prednisolone 15 mg daily. The classical response to a trial of steroids is 100% improvement within 24 hours. Clinical and biochemical response should then be reassessed within 2-3 weeks.

The many side effects of long-term steroid use are

widely known. It is important to discuss these with the patient prior to initiation of treatment. The important ones to mention are bone density loss which can lead to osteoporosis and the effect on blood sugar, which can result in new diabetes.

At diagnosis and each follow-up visit it is imperative to

consider the potential for associated GCA. The patient should be asked about headaches, jaw claudication and visual disturbance. If there is any suspicion of GCA, urgent discussion with the rheumatologist should take place that day.

Table 3

When to refer to secondary care

- Age < 60 years
- Onset > 2 months
- Lack of shoulder involvement
- Non-inflammatory stiffness
- Prominent systemic features, especially night pain, fever, weight loss, neurological features
- Features of another rheumatological disease
- Normal or extremely high inflammatory markers
- Incomplete/poorly sustained/no response to steroids
- Contraindications to steroid therapy
- Inability to reduce steroids
- Need for > 2 years of steroid treatment

NB If in doubt about whether referral is required individual cases should be discussed with the local on call rheumatologist

during the first year, in line with BSR guidance.⁷ Treatment duration should be 18 months on average.

REFERRAL

Patients with a classical presentation who respond to treatment (clinically and biochemically) can be managed in primary care.

Any atypical features increase the likelihood that the underlying diagnosis is something else. It is these cases that should have an early referral to secondary care.

Likewise, if treatment is not straightforward, patients should be managed under the supervision of a rheumatologist. Examples of these circumstances are outlined in table 3, above.⁷

COMPLICATIONS OF TREATMENT

The many side effects of long-term steroid use are widely known. It is important to discuss these with the patient prior to initiation of treatment. The important ones to mention are bone density loss which can lead to osteoporosis and the effect on blood sugar, which can result in new diabetes or difficulty controlling pre-existing diabetes. Follow-up blood tests should include glucose and/or HbA_{1c}. Tight control of blood sugar is required to minimise complications from diabetes.

Fracture risk should be considered in all patients who have a successful trial of

steroids (i.e. at the first review). That is the decision point when the patient is committing to more than 3 months of steroid therapy, so fracture risk should be calculated at that stage, taking into account use of steroids and the patient's other individual risk factors.

FRAX is a useful tool to guide risk stratification and can be used as part of the shared decision-making process.¹⁶ Individuals who have a high fracture risk (red zone) are recommended for treatment with bisphosphonates and vitamin D supplements, if required, without the need for a DEXA scan. Moderate risk patients (orange zone) should be referred for a DEXA scan and the FRAX score recalculated once the hip T score is available. Low-risk patients would only receive lifestyle advice and have FRAX recalculated if their risk factors change. Bisphosphonates, of course, have their own side effects and an open discussion with the individual will help both patient and doctor come to a decision about the optimal treatment strategy.

COMPLICATIONS OF DISEASE

As with other inflammatory conditions, PMR can flare. As mentioned above, flares can be managed by going back to the previous steroid dose and tapering at a slower rate. More than one flare may indicate the need for a steroid-sparing drug and therefore referral to a rheumatologist would be appropriate.

'At diagnosis and each follow-up visit it is imperative to consider the potential for associated giant cell arteritis'

At diagnosis and each follow-up visit it is imperative to consider the potential for associated GCA. The patient should be asked about headaches, jaw claudication and visual disturbance. If there is any suspicion of GCA, urgent discussion with the rheumatologist should take place that day. If there is visual loss this may require urgent discussion with the ophthalmologist, depending on the local trust protocol. Visual loss due to GCA should be treated with intravenous methylprednisolone. For more details on the management of GCA, refer to the BSR and BHPH guidelines.⁸ »

SYMPOSIUM

POLYMYALGIA RHEUMATICA

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Useful information

Polymyalgia Rheumatica & Giant Cell Arteritis UK (PMRGCAUK)

The charity offers support to sufferers and aims to raise awareness about the conditions and promote research in the field. There are local groups that meet regularly and are supported by local rheumatology departments.
www.pmrca.co.uk/content/home-page

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