Introduction

The diagnosis of polymyalgia rheumatica (PMR) can present a difficult challenge in primary care as there are no specific diagnostic tests and diagnosis depends on having a high index of suspicion supported by history, examination and raised inflammatory markers. On the other hand, PMR can be a very satisfying condition to treat in primary care as patients often have a very dramatic response to steroid therapy and are very grateful for the prompt relief of their symptoms.

Epidemiology of PMR

The epidemiology of PMR varies according to different studies with the incidence per 100,000 of the population aged over 50 reported between 13 and 68. The incidence increases with increasing age and there is an M:F ratio of 1:2.

Diagnostic criteria for PMR

Various authors have tried to set out diagnostic criteria for PMR, all of which include a combination of age, clinical features and a raised erythrocyte sedimentation rate (ESR). Bird et al list six criteria:

<table>
<thead>
<tr>
<th>TABLE 1. Criteria for diagnosis of polymyalgia rheumatica.1</th>
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<tbody>
<tr>
<td>1. Age over 65</td>
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<td>2. ESR over 40 mm/hr</td>
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<td>3. Bilateral upper arm tenderness</td>
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<td>4. Morning stiffness of more than 1 hour</td>
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<td>5. Onset of illness less than 2 weeks</td>
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<td>6. Depression and/or weight loss</td>
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The presence of any 3 of these criteria gives a sensitivity of 92% and a specificity of 80% for the diagnosis of PMR. If an additional criterion of a rapid response to oral steroid therapy is added, this increases the sensitivity to 99%.2 The Bird criteria are very useful and easy to apply in primary care, although it should be noted that they do not yet have universal acceptance in the rheumatological community as the definitive set of criteria.3

Clinical features of PMR

Clinical features include:
- stiffness and pain in shoulder and pelvic girdles
- raised ESR
- often systemic features of debility, weight loss, tiredness and low-grade fever
- either: dramatic onset of pain and stiffness
  or: insidious onset with less stiffness and pain and more systemic features.

Stiffness is usually the main feature. Affected muscles may be tender but the joints themselves are not usually affected.

The diagnosis can be straightforward in patients who present with a sudden onset of symptoms associated with a raised ESR. Some patients, however, present with a more insidious onset. In these patients systemic features may be more prominent, making the diagnosis much more difficult, and in these cases a wide range of differential diagnoses may need to be considered. These include:
- multiple myeloma
- other malignancy
- rheumatoid arthritis
- connective tissue disease
- osteoarthritis
- myopathy
- myositis
- hypothyroidism

ESR erythrocyte sedimentation rate; PMR polymyalgia rheumatica
• osteomalacia
• fibromyalgia
• shoulder problems (e.g. capsulitis or tears of the rotator cuff).

Investigations for suspected PMR

There is no specific test for PMR, although most patients will have a significantly raised ESR. If you suspect PMR it is worthwhile doing the following investigations:

<table>
<thead>
<tr>
<th>Investigation</th>
<th>Result</th>
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<tr>
<td>ESR</td>
<td>Usually raised</td>
</tr>
<tr>
<td>FBC</td>
<td>May be normocytic anaemia</td>
</tr>
<tr>
<td>LFT</td>
<td>May be raised alkaline phosphatase</td>
</tr>
<tr>
<td>Thyroid function tests</td>
<td>Hypothyroidism is associated with PMR</td>
</tr>
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</table>

Although the ESR is usually significantly raised in PMR there are some patients in whom the ESR is normal or only a little raised. In these patients and in those presenting with systemic features other causes of illness should be excluded.

Rheumatoid arthritis in an older patient may present with symptoms of PMR with shoulder girdle pain and stiffness. Be aware of the possibility of a polymyalgic presentation of rheumatoid arthritis if:

• there are peripheral joint signs such as synovitis
• there is a failure to respond to an adequate dose of steroids
• the initial dose of steroid cannot be reduced without exacerbating the symptoms.

If the diagnosis remains unclear, re-examine the patient thoroughly with other diagnoses in mind. At this stage other investigations may be useful. Consider the following:

<table>
<thead>
<tr>
<th>Investigation</th>
<th>Purpose</th>
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<tr>
<td>Protein electrophoresis</td>
<td>To exclude multiple myeloma</td>
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<tr>
<td>Rheumatoid factor</td>
<td>To support diagnosis of rheumatoid arthritis</td>
</tr>
<tr>
<td>ANA</td>
<td>To support diagnosis of connective tissue disease</td>
</tr>
<tr>
<td>X-ray of affected joint</td>
<td>To indicate osteoarthritis</td>
</tr>
<tr>
<td>CPK</td>
<td>To indicate myositis</td>
</tr>
<tr>
<td>Bone profile</td>
<td>To indicate osteomalacia</td>
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Remember that steroid therapy has risks and side-effects including:

• risk of osteoporosis – bone protection therapy should be initiated at the time of starting steroid therapy. Bone is lost rapidly in the early stages of steroid therapy so treatment should not be delayed while awaiting a dual-energy x-ray absorptiometry (DEXA) scan. In those patients aged over 65 and in those with a previous fracture a DEXA scan is not necessary as these patients are at considerable increased risk of osteoporosis and subsequent fracture and require continued bone protection therapy.

• gastric irritation – gastro-protection may be required.

Relapses

Relapses are common in PMR and should be diagnosed on clinical grounds rather than relying on ESR. They are usually caused by an overly rapid decrease in the dose of corticosteroid and should be treated by increasing the dose again and then titrating down more slowly. If the steroid dose cannot be reduced without exacerbating the symptoms in PMR, or if the patient suffers several relapses, consider another diagnosis – especially rheumatoid arthritis.

Relapses are commoner in the first 6–12 months of treatment and relapse rates are between 30 and 60%.

Prognosis

The long-term prognosis of PMR is good. Many patients can stop their steroid treatment at around 2 years, although some patients may require a small dose of steroids over several years. This is more likely to occur in females, and in those who were older and with a higher ESR at diagnosis.
Referral to secondary care

Most straightforward cases of PMR can be managed within primary care. In others however diagnosis and management may be difficult and these cases should be referred to a rheumatologist. Such patients may include those:

- who have an inadequate response to steroids
- in whom it is difficult to reduce the steroid dose without causing a relapse of symptoms

REFERENCES


COMMENT

The cardinal symptoms of PMR – prolonged morning stiffness in the shoulder and pelvic girdles with an elevated ESR – are generally straightforward. However PMR is essentially a diagnosis of exclusion. As the differential diagnosis is extensive it is important to exclude more sinister conditions at the outset with screening blood investigations. PMR is exceedingly rare below the age of 50 and occasionally the ESR is misleading, being normal or only slightly raised.

Of particular importance is the link between PMR and giant cell arteritis (also known as temporal arteritis). Many authorities believe the two represent opposite ends of the spectrum of the same disease. In any patient who presents with PMR it is mandatory to ask about headache, scalp tenderness and jaw claudication, as the catastrophic sequel of untreated GCA is irreversible blindness. In such cases higher doses of prednisolone are required with possible referral for temporal artery biopsy.

Education is paramount in PMR and patients should be warned that treatment is usually necessary for at least 2 years, with some requiring long-term maintenance therapy. In prednisolone-resistant cases response to steroid-sparing agents has been disappointing.

If diagnosis is proving difficult referral to a rheumatologist is prudent. However the treatment of a classic case of PMR is highly effective and one of the most satisfying in medicine.

PMR case study

- Mrs X, aged 88, presented with a 2-week history of pain and stiffness in her neck and shoulders and to a lesser extent around her hips. On examination she had a reasonable range of movement of her neck but some difficulty in raising her arms above her head. Past history included osteoarthritis of the spine and hip with a hip replacement 10 years previously.
- At this stage it was thought that she probably had PMR and investigations of ESR, CRP, FBC and rheumatoid factor were undertaken. While awaiting the ESR result she was treated with analgesics rather than anti-inflammatory drugs because of a history of hiatus hernia and oesophageal stricture.
- Initial results showed a normal ESR of 5, raised CRP of 32 with a normal FBC, with rheumatoid factor still awaited.
- At this stage the patient’s stiffness and pain worsened to the extent that she was unable to dress herself and spent her nights sitting in a chair as she felt unable to lie down in bed because of her symptoms. She complained of feeling generally unwell, with loss of appetite.
- The differential diagnosis remained between PMR and RA. Signs and symptoms seemed more in favour of PMR but ESR was only 5. At this stage we received the rheumatoid factor result which was 382 (normal 0–22). Although careful examination of the patient showed no evidence of synovitis it was possible that this was a case of RA with a polymyalgic presentation.
- This patient was miserable and needed treatment to alleviate her symptoms. Should we treat her with oral steroids, perhaps starting at a dose of 15 mg daily on the presumptive diagnosis of PMR? On the other hand if the diagnosis turned out to be RA perhaps oral steroids would not be the treatment of choice.
- Following discussion with a consultant rheumatologist colleague we decided that the best option was to give a dose of intramuscular steroid and review at hospital outpatient.
- Because of her on-going symptoms we also added a COX-2 inhibitor and amitriptyline to her medication at this stage.
- She responded well to the steroid injection with resultant decrease in stiffness and pain allowing her to attend her grandson’s wedding.
- Subsequent ESR was reported as 99 and to date the patient has not developed any signs of synovitis. She is still awaiting her clinic appointment.

This case study illustrates the difficulty we often have in Primary Care in making a diagnosis between PMR and early RA.

Confounding factors in this instance were:
- the original ESR of 5
- high level of rheumatoid factor
- no signs of synovitis.

Often the situation only clarifies as time goes on.

Perhaps this patient should have been treated with oral steroids at presentation.
Suggested PMR audit

It is a useful exercise to take a retrospective look at the care and clinical course of patients within your practice with a diagnosis of PMR. Useful data to collect includes:

- age at diagnosis
- presenting symptoms and signs
- initial ESR
- starting dose of prednisolone
- whether there was a dramatic response to treatment
- whether patients received osteoporosis advice or treatment
- number of relapses
- total length of treatment with steroid therapy
- referrals to secondary care
- any laboratory tests such as thyroid function and rheumatoid factor.

An audit of PMR was undertaken in 1993 by GP members of the Primary Care Rheumatology Society. Data from 47 patients in primary care were analysed retrospectively. The M:F ratio was 17:30 and the mean age at diagnosis was 69.6 years.

<table>
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<tr>
<th>Questionnaire</th>
<th>Results</th>
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<tr>
<td>Most common presenting symptoms</td>
<td>Shoulder pain and aching 25 (53%)</td>
</tr>
<tr>
<td>Initial ESR</td>
<td>Mean 67 range 5–126 (SD28)</td>
</tr>
<tr>
<td>Relapses</td>
<td>• 32 (68%) suffered at least 1 relapse</td>
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<td></td>
<td>• 18 (38%) suffered 2 or more (1 patient had 6)</td>
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<tr>
<td>Mean initial starting dose of prednisolone</td>
<td>30 mg range 10–60 (SD12)</td>
</tr>
<tr>
<td>Osteoporosis advice or treatment</td>
<td>18 (38%)</td>
</tr>
<tr>
<td>Initial dramatic response to treatment</td>
<td>35 (75%); 10 (21%) referred</td>
</tr>
<tr>
<td>Non-dramatic response to treatment</td>
<td>12 (25%); 36 (77%) referred</td>
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Audits undertaken today should show a lower initial starting dose of prednisolone (around 15 mg) and a much higher percentage of patients counselled regarding, or treated for, osteoporosis. Only 38% of patients in the audit above were noted to have been given treatment or counselling about the risk of osteoporosis. The new Guidelines on corticosteroid induced osteoporosis advise starting bone protective therapy at the same time as starting steroid therapy in all patients aged over 65 or with a history of fragility fracture.

REFERENCE