Red Whale

In association with VERSUS ARTHRITIS

# Polymyalgia rheumatica

Polymyalgia rheumatica (PMR) is predominantly diagnosed and managed in primary care. It can be very satisfying to diagnose and manage because we have an effective treatment that works.

But there are some challenges for us too: misdiagnosis rates could be as high as 50%, and follow-up and management of the consequences of long-term steroids, e.g. osteoporosis, diabetes and hypertension, may not be adequate.

There is also some diagnostic overlap with giant cell arteritis, and you may also find that article useful.

## How are we doing in primary care?

A study of UK general practitioners highlighted the following issues in the primary care diagnosis and management of PMR (BJGP 2018;68(676):e783):

- Features that made us consider the diagnosis were in line with current guidance – but we found diagnosis challenging, largely relying on response to steroids.
- We are not routinely requesting all the investigations recommended by the British Society for Rheumatology and EULAR to rule out other conditions.
- GPs *reported* starting bone protection in about 80% of cases and gastroprotection in 70% of cases. Studies auditing patient records suggest lower levels of initiation of bone protection and PPIs.

The current evidence and guidance for PMR was reviewed in a seminar in the Lancet (Lancet 2017;390:1700). There is no NICE or SIGN guidance, but the British Society for Rheumatology produced guidance in 2009 and EULAR in 2015.

## Facts and stats

- The lifetime risk of developing PMR is 2.4% for women and 1.7% for men.
- Incidence peaks between age 70–80y; it rarely occurs under the age of 50y (so, think of other things).
- The incidence is highest in those of Northern European descent.

## Aetiology and pathology

PMR is an inflammatory disorder causing proximal muscle pain and stiffness. Like many rheumatological conditions, we neither know the cause of PMR, nor fully understand what causes the symptoms. Research imaging studies (PET/CT, MRI) show that the inflammation is predominantly outside the joints, including the peritendinous areas (Rheumatology 2018; 57(2):345).

There are many theories of possible triggers but none at the moment are of much practical help to primary care clinicians.

*Let's look at potential pitfalls in diagnosing and managing PMR in primary care…*

## Making the correct diagnosis

In research studies, between 25 and 50% of people given a diagnosis of PMR are subsequently found to have a different diagnosis, most commonly late-onset rheumatoid arthritis or giant cell arteritis (GCA).

Diagnosis of PMR is primarily based on a thorough clinical history and physical examination, supported by elevated inflammatory markers.

The symptoms are very characteristic and are dominated by:

- Painful stiffness in the:
  - Upper arms.
  - Lateral hips.

    - Buttocks.
    - Thighs.
- Pain eases off over the first few hours of the morning.
- Some patients describe all-over stiffness that lasts all day.
- Typical functional impact of difficulty getting out of bed, turning in bed and getting up from a chair due to stiffness (interestingly, some qualitative studies suggest that patients may describe symptoms in terms of disability, rather than localised symptoms).
- Feeling systemically unwell.

Physical examination is directed at excluding other diagnoses – particularly infection or cancer. If examined in the morning, patients may be very stiff on mobilising.

**The most important thing if suspecting PMR is to evaluate at regular intervals for GCA as this may often accompany PMR or appear weeks, months or years later.**

If the diagnosis is unclear, refer to rheumatology.

## Differential diagnoses

Other conditions we should consider include:

- Active cancer.
- Infection.
- GCA.
- Inflammatory arthropathies, e.g. rheumatoid.
- SLE, myopathy, connective tissue disease.
- Local shoulder/hip conditions.
- Fibromyalgia/pain syndromes.
- Hypovitaminosis D: knee and thigh pain alone?
- Drug-induced myopathy – statins.

*Ask yourself, are any of these more likely?*

## Investigations

*There has been great store set in a number of primary care journals over recent years in our 'failure' to collect the 'minimum data set' recommended in the 2015 EULAR guidelines (BJGP 2018;68(676):e783). However, interestingly, this is not a concept familiar to many rheumatologists; apparently, if we mentioned the 'minimum data set' in our referral letters, we would not be talking a common language!*

So, instead, we should use our clinical judgement to determine whether all of these tests are appropriate for the particular patient in front of us.

They are a pragmatic guide to screen for alternative explanations of symptoms and potential comorbidities that are important when we are considering long-term steroids:

| **Investigations recommended by EULAR 2015** |
|---|
| After a full examination, do:<br>• Bloods: FBC, ESR, CRP, U&E, LFT, calcium, glucose/HbA1c, RhF, anti-CCP *(consider TFTs, creatine kinase, myeloma screen, vitamin D).*<br>• Urinalysis.<br>• Consider a CXR (if prominent systemic symptoms/smoker, etc.).<br>This is considered a 'minimum dataset' and should be documented at diagnosis. |

## PMR vs. GCA overlap: think PMR, think GCA

Giant cell arteritis (GCA) is a large vessel vasculitis and a 'never miss' diagnosis because early treatment can prevent sight loss.

GCA and PMR are often concurrent and have some symptomatic overlap, and may be different facets of the same disease – but they are different in that they need different doses of steroids to achieve remission of symptoms:

- 50% of people with GCA exhibit PMR symptoms.
- 20% of people initially diagnosed with PMR have GCA.
- Imaging studies of patients with relapsing PMR frequently find evidence of GCA that is being partially masked by the steroid therapy.

So, each time we think 'could this be PMR?', we also need to think 'could this be GCA?' *(You may find the GCA article helpful).*

Specifically, check for:

| Symptoms suggestive of GCA | Signs suggestive of GCA |
|---|---|
| • New-onset headache.<br>• Scalp tenderness?<br>• Jaw/tongue claudication (pain on chewing).<br>• Visual disturbance.<br>• Fever/weight loss/fatigue.<br>• Limb claudication. | • Tender/thickened temporal artery.<br>• Scalp tenderness.<br>• Visual field defect.<br>• Asymmetry of pulses. |

**No single sign, symptom or test is sufficient to rule GCA in or out, so if you suspect it clinically:**

- **Admit/refer immediately** people with possible GCA and visual disturbance for ophthalmology review – IV steroids may be required.
- **Discuss same day** (usually with the rheumatology team) all those in whom we consider GCA a possible diagnosis.

## Who needs referral?

The British Society for Rheumatology and RCP Guidelines (2010) and EULAR 2015 guidance recommend we should consider referral in people with suspected PMR if they:

- Are younger (<60y).
- Have a more chronic or atypical onset.
- Have an absence of typical symptoms.
- Have a normal or very high ESR/CRP (>100) – *may suggest GCA or other pathology, e.g. late-onset rheumatoid*.
- Have incomplete/non-response to steroids.
- Have difficulty withdrawing steroids.

We should also discuss/refer as above if there are overlapping GCA symptoms.

## Treatment of PMR

Treatment is with oral corticosteroids, usually prednisolone. There will usually be a good symptomatic response within 24–72h.

**Prior to starting steroids**

- Assess for comorbidities that may increase the risk of glucocorticoid-associated side-effects, e.g. hypertension, diabetes/prediabetes, obesity, osteoporosis, cataract or glaucoma.
- Review medications for drugs that may increase the risk of side-effects or toxicity from the steroids.

Presence of multiple comorbidities or interactions would lead us to trial a lower dose of prednisolone in the first instance, and to have a lower threshold for referral in the event that higher doses of steroids were needed to maintain remission or if relapses occur (for early consideration of steroid-sparing therapy).

**Starting steroids**

There is no 'best regimen' due to an absence of evidence. This is the regimen recommended by the EULAR 2015 guidance (Lancet 2017;390:1700):

- Start with 12.5–25mg prednisolone daily – use the minimum effective dose.
- Maintain for 2–4 weeks.
- Taper gradually by 2.5mg every 4w until 10mg.
- After 1m of 10mg, reduce thereafter by 1mg each month.
- 3-monthly FBC, ESR/CRP, U&E and glucose are recommended *(a combination of HbA1c and a random glucose may be most useful; fasting glucose will miss a significant proportion as prednisolone will tend to impact on glycaemic processing of meals)*.

Give the patient a steroid card to show if they receive any medical care/buy medications. This can be used as a shared record of the tapering regimen. They should also carry a steroid emergency card to warn of the potential of iatrogenic adrenal insufficiency (this is now recommended for all taking ≥5mg prednisolone daily) (Clinical Medicine 2020;20 (4):371).

**Assessing response**

Patients will usually have a good symptomatic response within 24–72h.

If higher doses of steroids (>25mg) are needed to get a clinical response, re-evaluate the diagnosis – could this be GCA?

## Follow-up

**The EULAR guidance recommends follow-up visits every 4–8 weeks in the first year of treatment and every 8–12 weeks in the second year.**

**At these visits, we should:**

- Assess clinical remission (symptoms/signs), including checking FBC, ESR/CRP, U&E, glucose.
- Check for new symptoms/signs that may suggest GCA, and ensure patient is aware of GCA symptoms that should prompt seeking urgent medical advice.
- Check for side-effects/complications of glucocorticoid therapy, e.g. monitor weight and BP.

## Protecting people from the impact of prolonged glucocorticoids

Consider bone and gastroprotection:

| Drug | Dose | Notes |
|---|---|---|
| Proton pump inhibitor, e.g. omeprazole or lansoprazole | Standard dose | For gastroprotection for duration of steroids. |
| Bone protection: usually a bisphosphonate + calcium/vitamin D if needed | Standard dose for preparation | All people starting long-term steroids should have a fracture risk assessment using either FRAX or QFracture, ideally as soon as they start their steroids as bone loss occurs quickly. See article on *Osteoporosis* for more details on how to interpret this. The majority of people will need bone protection.<br>Risk returns to baseline about 12 months after stopping treatment (DTB 2010;48:98). |

## Relapses or difficulty weaning steroids

About 50% of patients will relapse during treatment, requiring steroids to be stepped up again for a brief period before tapering more slowly. The median duration of steroid treatment is between 1 and 2 years, but up to 40% will need more than 2 years of treatment, with 20–33% needing 5 years or more (Rheumatology 2010; (4):716-22).

Patients with persistent symptoms or multiple relapses should have the diagnosis reviewed. It may indicate undiagnosed GCA or another inflammatory arthritis – refer for further assessment.

DMARDs, usually methotrexate, are sometimes used in this group. This is a secondary care decision.

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|---|---|
|  | **Polymyalgia rheumatica**<br>• Have a logical approach to the diagnosis of PMR; look for red flags and consider alternatives.<br>• Always think, could this be GCA?<br>• Ensure patient is educated about potential symptoms of GCA which should prompt seeking urgent (same-day) medical advice.<br>• Be ready to review diagnosis if there are atypical features or a poor response to treatment.<br>• Protect the patient's bones and stomach. |
|  | Review your last 2 diagnoses of PMR. Did they have all the recommended baseline investigations prior to a diagnosis being made?<br>Review any patients on long-term steroids for PMR. Do they have:<br>• A written tapering regimen/steroid card?<br>• Gastroprotection?<br>• Bone protection?<br>• Evidence of assessment for the complications of long-term steroids?<br>Would developing a practice/PCN-wide protocol and template to support long-term prescribing of steroids be helpful? Do you have a practice pharmacist who could help with this? |
|  | **Steroid emergency cards can be ordered here:**<br>https://secure.pcse.england.nhs.uk/_forms/pcsssignin.aspx<br>**Patient information:**<br>https://tinyurl.com/Red-Whale-PMR-leaflet<br>**Treatment:**<br>Steroids \| Side-effects, uses, time to work (versusarthritis.org)<br>**Living well:**<br>Managing fatigue \| Causes, self-help, support (versusarthritis.org)<br>Get help \| Helpline, online community, arthritis virtual assistant (versusarthritis.org)<br>Exercising with arthritis \| Top tips, specific exercises (versusarthritis.org)<br>Let's Move with Leon (versusarthritis.org)<br>**Patient support group:**<br>http://www.pmrgca.co.uk/content/home-page |
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