Referral of patients with suspected polymyalgia rheumatica: how complete is our view of ‘planet PMR’?

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Polymyalgia rheumatica (PMR) is the second most common inflammatory rheumatic disease after rheumatoid arthritis in persons older than 50 years, reaching an estimated prevalence of up to 1.5% in this age group. Despite its frequency, many uncertainties and misconceptions still exist about how PMR should be diagnosed and managed. In many countries, primary care appears the practical and appealing setting for the triage and treatment of patients with PMR. Patients present with abrupt onset of symptoms that are usually accompanied by elevated inflammatory markers, making the initiation of glucocorticoid (GC) treatment a tempting and apparently easy choice for the general practitioner (GP).

Despite this seemingly straightforward scenario, experience over the years has repeatedly demonstrated that at least one-third of patients initially classified as having PMR eventually receive another diagnosis, usually elderly onset inflammatory arthritis, but also any number of others including infection and malignancy. In primary care, alternative conditions in ‘typical’ cases are often not extensively considered, and the response of symptoms to GC administration is extensively considered, and the response of symptoms to GC administration is regarded as confirmation of the diagnosis. A international online survey of 394 GPs and 937 rheumatologists showed that only 25% of patients with suspected PMR are referred from primary care to rheumatologists, with substantial variation between countries, ranging from 10% in the UK and the Netherlands to 60% in Italy and Romania, and up to 100% in Colombia (with wide variation in the percentage of referral also within countries). A median of 50% (IQR 15%–73%) of patients were seen by a rheumatologist after starting treatment in the primary care setting.

Considerable differences are evident in the diagnostic approach taken by GPs and rheumatologists. More than 15% of GPs do not always request C reactive protein in patients with suspected PMR, more than 40% never or rarely check the presence of anticitrullinated protein antibodies, and about two-thirds never or rarely ask for any imaging examination. The approach to treatment by GPs and rheumatologists is also discrepant. GPs prescribe a median prednisone starting dose of 20 mg instead of the median dose of 15 mg prescribed by rheumatologists. Notably, more than 30% of GP respondents used starting prednisone dosages higher than 25 mg per day, diverging from the starting dosage between 12.5 and 25 mg currently recommended by American College of Rheumatology and European Alliance of Associations for Rheumatology.

In Annals of the Rheumatic Diseases, Keller et al propose the first consensus-based recommendations for early referral of individuals with suspected PMR. An initial systematic literature review informed the proposed population, intervention, control and outcomes format questions underlying the recommendations. The two overarching principles are patient-focused, stating that management of patients with suspected PMR should be based on shared decisions between the patient and the healthcare provider and highlighting the necessity of informing patients of the potential overlap with GCA, a dangerous and insidious companion of PMR.

We briefly review the five recommendations, discussing the first together with the fifth. Recommendation 2 advocates for a thorough history and clinical examination, combined with laboratory examinations, before specialist referral. While concurring, we suggest that practical application of this recommendation may be hampered by the lack of specific features of PMR. No data are available on the diagnostic predictive value of each manifestation of PMR, in contrast, for example, to limb and jaw claudication, which show a high likelihood ratio for the diagnosis of GCA, at least its cranial form. Despite the several sets of classification and diagnostic criteria that have been proposed for PMR, none have optimal performance. As a consequence, the identification of PMR still relies on ‘clinical gestalt’.

Two of the fundamental conundrums faced by clinicians evaluating patients with polymyalgic symptoms are the possibilities of concomitant GCA and underlying malignancy. If cranial manifestations typical of GCA are present, the diagnosis may be clear, leading the GP to initiate GC treatment and rapid specialist referral. Conversely, large vessel involvement may present with subtle and non-specific manifestations, such as fatigue and fever, complicating GP decision-making. Moreover, clinical predictive factors of the presence of large vessel vasculitis, in patients with apparently isolated PMR, are lacking, as is evidence whether such patients require more aggressive treatment. In the context of this recommendation, then, should the GP also be responsible for screening large vessel involvement in patients with PMR?

Some cancers may elicit or mimic PMR symptoms. A cohort study using the UK General Practice Research Database revealed a higher likelihood of malignancy detection in the first 6 months following a diagnosis of PMR. Evaluation of the patient for cancer should preferably be performed by GPs, noting that cancer screening initiatives are often part of primary care practice. However, the infrequent request for imaging studies in

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patients with suspected PMR, highlighted in the aforementioned survey, may raise concern about the extent of malignancy investigation.15 Recommendation 3 advocates that patients with severe symptoms be referred via rapid access pathways. Although this approach is reasonable and preliminary data suggest that PMR fast-track clinics may reduce outpatient visits and even hospitalisation, the totality of evidence on this topic is still scanty18 and it should be considered in the context of the global healthcare workforce shortage.24 Recommendation 4 suggests that GC therapy should be deferred for those patients referred to rheumatologists via fast-track pathways. In contrast to GCA, for which immediate initiation of GC is recommended to avert potential sight loss, PMR does not pose imminent organ-threatening risks. Initiation of treatment before the diagnosis is established may blur the clinical picture and reduce the diagnostic performance of vascular imaging undertaken to assess for possible GCA.23 The pain and stiffness of PMR may be unbearable, leaving GPs with little option than to prescribe GC ahead of referral.

Recommendation 5 considers a return of the patients to the primary care setting following specialist evaluation, provided that there has been a good initial response to GC and that the patient has a low risk for treatment-related adverse events. This statement is necessarily tied to recommendation 1 that ‘each individual with suspected or recently diagnosed PMR should be considered for specialist evaluation’, and to the leitmotif of the recommendation effort, namely that every patient with PMR should be managed, at least initially, in the rheumatology specialist setting. The possibility that the patient with PMR may have underlying active GCA requiring specialist attention and advanced therapies is an important concern driving the recommendations, but are they realistic and feasible?

The proportion of the general population taking GC for any reason ranges from 1.2% to 1.7%.26,37 In 2008, 0.75% of the UK general population was receiving long-term (ie, ≥3 months) oral GC, with asthma, PMR/GCA and chronic obstructive pulmonary disease representing the most frequent indications, accounting respectively for 18.7%, 13.2% and 13.4% of total long-term GC prescriptions.28 Another study from Sweden identified PMR and GCA as the most frequent reasons (29.5% of all conditions) for long-term use of GC, defined in this case as consumption of more than 300 tablets

Available data suggest that GPs tend to treat PMR with higher GC dosages for shorter periods compared with rheumatologists,15 an approach associated with a higher relapse rate.16 However, it is probable that this mare magnum of GC prescription stems mainly from primary care, where many, and perhaps most, patients with PMR are diagnosed and treated.

The Earth and the Moon both rotate, but, due to the synchronisation of their respective spins, the same side of the moon is always seen from the Earth, a phenomenon called ‘tidal locking’.26 Similar, while the rheumatologists on the task force have deep knowledge and experience with PMR, and can reflect what we as a rheumatology community think we know about PMR and how it is seen in the wider world of general practice, it nevertheless may be a single view of what the ‘planet PMR’ as a whole actually is. The four GPs on the task force developing these recommendations may also have a view of PMR that is different than that of their rheumatology colleagues, and also not fully representative of the vast primary care universe in which more than 80% of patients with PMR may be exclusively managed, on the dark side of the ‘PMR planet’, unseen by the rheumatologist.15

Despite these potential limitations, the current recommendations for referral of subjects with suspected PMR represent significant progress towards increasing awareness for PMR, implementing the standardisation of treatment and improving the management of these patients.

**REFERENCES**

Editorial


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