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Review

Current diagnosis and treatment of polymyalgia rheumatica

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ABSTRACT

The diagnostic and therapeutic strategies for polymyalgia rheumatica (PMR) have changed substantially in recent years. Rather than a single disease entity, PMR has emerged as a syndrome produced by a variety of conditions. The diagnostic criteria that have been used for several decades are inadequate. These facts support a new and broader pathological concept, polymyalgic syndrome, and a standardized diagnostic and therapeutic approach designed to rule out diseases with misleading presentations and to identify the limited number of patients with polymyalgic syndrome who have PMR. Criteria for both polymyalgic syndrome and PMR were developed recently but remain to be validated. These criteria are discussed, as well as the suggested diagnostic approach and treatment strategy. In contrast, studies on pathophysiological models, inflammatory mechanisms, and genetic factors are not considered herein, as they were conducted in heterogeneous populations of patients who did not meet the new criteria. Current data indicate that polymyalgic syndrome is a mode of onset of inflammatory joint disease in individuals older than 50 years of age and not (in most cases) a disease entity.

1. Introduction

In 1953, Forestier and Certonciny [1] described an apparently inflammatory condition involving only the limb girdles and affecting older individuals, which they designated “pseudo-polyarthrite rhizomélique” “polymyalgia rheumatica” (PMR). Links were then recognized between PMR and giant-cell arteritis (GCA), and the term polymyalgia arteritica was suggested in the 1960s to designate both diseases [2,3]. The main flaw of this concept is that it views PMR and GCA as two expressions of the same disease entity, whereas most patients who present with symptoms suggesting PMR, i.e., with polymyalgic syndrome, do not have GCA.

Recent studies support the view that PMR and GCA are two distinct conditions that overlap to some extent [4,5]. The misconception that PMR and GCA are the same disease has led to the inclusion in several clinical and therapeutic studies of patients with either condition. Recent work supports the polymyalgic syndrome concept and indicates a pressing need to look for the cause of polymyalgic syndrome elsewhere than in GCA.

Advances in the field of PMR can be credited to an ACR-EULAR task force [6] that has been working for 15 years to dissect the clinical, diagnostic, and therapeutic aspects of PMR and to develop management guidelines, as well as criteria sets for diagnosing polymyalgic syndrome and PMR. Their work does not presume to resolve all the issues raised by PMR but has nevertheless resulted in substantial progress.

2. Background

PMR is a clinical condition defined as the presence of inflammatory pain in the limb girdles [6]. Range of motion is decreased at the shoulder and pelvic girdles, and the pain radiates to the buttocks and cervical spine. By definition, these symptoms persist for at least 1 month and are combined with incapacitating morning stiffness, a decline in general health, and an elevated acute-phase response. PMR affects individuals older than 50 years of age and is the most common inflammatory rheumatic disease of the elderly. According to the new concept, PMR is a benign and highly steroid-sensitive disease that should resolve fully with low-dose glucocorticoid therapy for 12–24 months, although one or two relapses may occur. By definition, conditions that fail to meet this description constitute polymyalgic syndrome and not PMR.

Thus, polymyalgic syndrome is a set of manifestations that mimic PMR at presentation but are ascribed based on subsequent information to another disease. In contrast, PMR is the condition described by Forestier, whose evaluation may benefit from the new diagnostic criteria. However, PMR continues to raise many unresolved issues.

3. Unresolved issues

At present, the management of PMR is affected by six unresolved issues [6–9]: (a) the definition of PMR rests only on clinical criteria; (b) no specific diagnostic test is available; (c) no treatment objective has been defined; (d) the clinical course is highly variable; (e) no definitions exist for PMR flare or relapse; and (f) no consensus has been developed regarding the treatment of PMR.

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The absence of specific markers for PMR and extraordinary variability in the clinical manifestations mandates the use of diagnostic criteria in clinical practice [6,7]. The disparate criteria suggested in the past had good sensitivity but low specificity for PMR. As discussed below, even in experienced hands these criteria produce the wrong diagnosis in half the cases. In other terms, they allow the diagnosis of polymyalgic syndrome but not of PMR.

PMR is the leading reason for long-term glucocorticoid therapy in individuals older than 60 years of age. Some patients remain on glucocorticoid therapy for years (and probably have polymyalgic syndrome), whereas current data indicate that patients with PMR can be taken off glucocorticoid therapy after 1 or 2 years [6–9].

In rheumatoid arthritis (RA), the treatment objective is to obtain a remission. No treatment objective has been clearly defined in PMR [6,7,9]. Possible objectives include absence of pain, weaning off glucocorticoid therapy, functional recovery, absence of relapses, and resolution of the systemic inflammation.

The treatment of PMR is not standardized [6–9]. A nonsteroidal antiinflammatory drug or low-dose glucocorticoid therapy (< 0.3 mg/kg/day) has been suggested. Studies found no benefits from the use of hydroxychloroquine, azathioprine, methotrexate, or TNFα antagonists, although these negative results may be ascribable to the heterogeneity of the patient populations.

Finally, when used as a diagnostic criterion, a rapid and dramatic response to glucocorticoid therapy results in an erroneous diagnosis in half the cases (for the same reason). In several studies [6,9], after 3 weeks of glucocorticoid therapy, 35% of patients on average had persistent systemic inflammation (22% with an ESR > 30 mm/h and 32% with CRP > 10 mg/L), 45% a complete clinical response, 44% a partial clinical response, and 11% no response. A poor response was associated with a higher risk of relapse within the first year (27%, 33%, and 37%, respectively). After 3 weeks, one-third of patients still reported limb girdle pain and morning stiffness.

4. Suggested broadened definition of polymyalgic syndrome

The ACR/EULAR task force has suggested the concept of polymyalgic syndrome, which can indicate PMR or other diseases [6]. According to this concept, the diagnostic strategy must be expanded considerably. Then, exclusion criteria must be ruled out. Finally, standardized treatment with 15 mg/day of prednisone is given. Thus, the definite diagnosis of PMR can be established only at the end of follow-up, usually after 12–18 months of treatment, when the patient is fully recovered and off glucocorticoid therapy. This strategy can be likened to an obstacle course during which the patient and rheumatologist rule out conditions that mimic PMR. As shown in Table 1, these criteria are considerably simpler than the previous ones and allow the inclusion of a larger number of patients.

The task force suggests the following stepped diagnostic process:

- evaluate for inclusion criteria;
- evaluate for exclusion criteria;
- prescribe standardized low- or moderate-dose glucocorticoid therapy;
- evaluate the response;
- and confirm the diagnosis on follow-up and treatment discontinuation.

Table 1
Core criteria for polymyalgic syndrome.

Core inclusion criteria
Age > 50 years
Bilateral shoulder or pelvic girdle aching, or both
Morning stiffness duration of > 45 minutes
Evidence of an acute-phase response (e.g., raised ESR or CRP)

Table 2
Categories of diseases that may present as polymyalgic syndrome.

Common diagnoses (strong similarities with PMR)
Active infection
Cancer
Giant-cell arteritis (temporal headache, visual disturbances, jaw, tongue, or limb claudication. . .)
Rare diagnoses (limited similarities with PMR)
Chronic inflammatory joint diseases (rheumatoid arthritis, spondyloarthritis, psoriatic arthritis, lupus. . .)
Crystal deposition disease
Chronic multifocal pain syndromes
Myopathies (metabolic, inflammatory, drug-induced. . .)
Endocrinopathies
Neurological conditions (e.g., Parkinson's disease)

5. Exclusion criteria

The criteria used for years to diagnose PMR were neither selective nor specific [10–14]. By giving clinicians a false sense of confidence in the diagnosis, they carried a risk of unnecessary and/or prolonged exposure of patients to glucocorticoids. The diagnostic error was often discovered only late in the course of the disease.

The new criteria expand the diagnostic field (Table 1), thereby inducing a risk of poor specificity. To improve specificity, the new approach involves a list of exclusion criteria and a standardized treatment regimen [6].

The exclusion criteria are divided into frequent and rare conditions, depending on the likelihood that their presentation may mimic PMR (Table 2). An exhaustive list of conditions that must be sought is provided (Table 3). The conditions at the top of Table 2 are considered closely similar to PMR in terms of the clinical picture, and those in the lower part of the table as less likely to mimic PMR. This classification is partly subjective, as no detailed epidemiological studies are available. My own experience suggests that polymyalgic syndrome is more likely to result from elderly-onset RA or spondyloarthritis than from infective endocarditis [15–18]. However, rather than the contents of the list, the important point is the emphasis on looking for possible causes to polymyalgic syndrome other than PMR (Table 3) [17,19–28].

The usefulness of looking for exclusion criteria is well illustrated by a prospective study of 249 patients with suspected PMR (mean age, 71 years; 60% of women) seen at eight rheumatology clinics in England [29]. Although the patients were evaluated by experienced rheumatologists, diagnostic errors were common. Fulfilment of Jones and Hazelman criteria for PMR was an inclusion criterion, but no exclusion criteria were applied routinely and all patients received 15 mg/day of oral prednisolone tapered according to a predefined schedule. Six months later, 120/249 patients had incontrovertible evidence of another disease and after 1 year 10 additional patients received other diagnoses (e.g., cancer or RA). Thus, overall, 130/249 (52.2%) patients were initially given a mistaken diagnosis of PMR.

6. Routine laboratory tests

To facilitate the initial workup and follow-up, the task force recommends a minimal set of laboratory investigations (Table 4). These investigations contribute to the initial diagnostic strategy and serve as a reference data set in the event the initial diagnosis is subsequently called into question. This list of investigations is