REVIEW

Polymyalgia rheumatica and temporal arteritis: evidence and guidelines for diagnosis and management in older people

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Abstract

Polymyalgia rheumatica and temporal arteritis commonly present for the first time in older people. Guidelines for diagnosis, investigation and management are available but have largely been developed by rheumatologists from studies where older people have been excluded. It is not clear whether geriatricians care for a group of patients with different clinical presentations compared to those under the care of other clinicians. There is a need for further prospective studies and randomised controlled trials to clarify a host of outstanding issues to improve the care of older people with polymyalgia rheumatica and temporal arteritis.

Keywords: polymyalgia rheumatica, temporal arteritis, steroids, older people

Introduction

Polymyalgia rheumatica (PMR) and temporal arteritis (TA) are two common related disorders affecting older people. The term polymyalgia rheumatica was introduced by Barber in 1957 but the syndrome had been recognised in the late nineteenth century [1]. The syndrome is characterised by pain and stiffness in proximal muscles. Temporal arteritis was first described by Hutchinson in 1890 [2] and typically presents with headache and scalp tenderness. Since 1965, the two conditions have been regarded as part of the same disease spectrum [3].

The mean age of onset of these conditions is 70 years [4], with the majority of cases occurring between 60 and 75 years. Both TA and PMR are rare in people less than 50 years of age and incidence rates are probably highest in those over age 80 [5–7]. However, this age distribution may not reflect the experience of geriatricians who may manage the oldest individuals with these conditions.

PMR/TA is a chronic disease and the majority of patients with PMR/TA are cared for by rheumatologists and general practitioners but as diseases of older people, the conditions are frequently managed by geriatricians. It is not clear whether geriatricians care for a group of patients with different clinical presentations compared to those under the care of other clinicians. Accurate diagnosis is important to ensure that patients with the conditions are recognised and that others are not given treatment, with its associated risks, unnecessarily. The correct dosage and regime of steroid treatment is important to avoid undertreatment resulting in increased risk of arteritic complications, but also overtreatment which may result in an increased risk of steroid-associated side effects.

Guidelines for diagnosis and management of PMR/TA have been developed by rheumatologists and in many studies older people have been excluded. Therefore much of the evidence used in clinical practice is extrapolated from studies in younger age groups. Review of the current literature, particularly the various guidelines and recommendations for diagnosis and treatment of these conditions reveals a dearth of evidence specific to geriatricians.
Guidelines for diagnosis

The diagnostic process for PMR/TA can be divided into three components. Firstly the assessment of the clinical features which may be grouped into myalgic, arteritic and systemic. Secondly, information gained from supportive tests such as inflammatory markers or temporal artery biopsy. Thirdly, the exclusion of the other conditions giving similar clinical features [8]. This final aspect is particularly pertinent in polymyalgia where the symptoms may be largely non-specific. From the conditions listed in Table 1, there is potential for adverse consequences if steroids are used injudiciously.

Attempts have been made to provide evidence-based frameworks for diagnosis. However, as a gold standard diagnostic investigation does not exist for either PMR or TA, the validity of any specificity and sensitivity data has to be viewed with this in mind.

Polymyalgia rheumatica

In 1978, Bird described clinical features and investigations in 146 patients assessed retrospectively by rheumatologists to be suffering unequivocally from PMR compared to 253 patients suffering from conditions that frequently mimic PMR [9]. The seven most discriminatory criteria for diagnosis were established from sensitivity and specificity calculations (Figure 1). If three of seven criteria were satisfied a 92% sensitivity and 78% specificity for a diagnosis of PMR was achieved, and this was suggested for use in clinical practice.

The control population for this study was obtained from new referrals to a rheumatology unit and later validated on a group of new referrals to a general medical unit. The mean age of the patients recruited into the study was not stated and no comment was made as to the influence of increasing age on clinical features. Age > 65 years and ESR > 40 mm/hour are two of the stated criteria for diagnosis but these may not be sufficiently discriminating for the caseload seen in geriatric practice. We reviewed new cases presenting in 2000 to our geriatric unit and found that 9/10 (90%) did satisfy the criteria [10].Whilst this may represent high sensitivity, it was not possible to review specificity from our data, which would be of concern. Overall, Bird’s criteria for diagnosis comprise the largest series from the best available evidence base, in an area where evidence is scanty.

The diagnostic criteria for both PMR and TA frequently cited from Jones and Hazleman [11] were developed purely as inclusion criteria for clinical trials but it is important to recognise that though they are used extensively they were not validated or necessarily intended for use in everyday clinical practice.

Temporal arteritis

The American College of Rheumatology in 1990 [12] derived diagnostic criteria for TA from a retrospective study of over 214 cases. The mean age at onset was 69 years and only 17 cases were over 80 years of age. These guidelines have sensitivity and specificity values of over 90%. However the control group used were 593 patients suffering from ‘other forms of vasculitis’ making their use in a geriatric setting less clear. These guidelines, in contrast to others, are more applicable to clinical practice as a temporal artery biopsy does not need to be performed in all cases and the response to steroid therapy is not specified as a criterion.

Investigation

There is general agreement in the literature of the investigations that should be performed (Figure 1). Although some of these are used to screen for other conditions, these investigations may be abnormal in PMR/TA per se reflecting the systemic acute phase response resulting in a normochromic normocytic anaemia and abnormal liver function tests [13]. In general, increased viscosity, ESR and CRP are almost invariably present. However it is not clear how increasing age modulates this inflammatory response. It is also important to note that relapses in PMR/TA are common and that in 50% of clinical relapses the ESR or CRP is not raised [14, 15].

The role of temporal artery biopsy is controversial with numerous factors influencing the positivity rates. In cases of ‘pure’ polymyalgia rheumatica the rates of positive biopsies are low, between 15 and 20%. In ‘pure’ temporal arteritis the rate is between 60 and 80% [16]. The influence of steroid therapy is strong. In a study of 132 cases of TA [17], 82% of biopsies were positive pre-steroid therapy decreasing to 60% after one week of treatment and to 10% after two weeks. Taking into account these various factors, Hazleman [6] recommends a pragmatic approach to temporal artery biopsy:

- Perform biopsy if diagnosis is in doubt, particularly if systemic symptoms predominate.
- Biopsy is most useful within 24 h of starting treatment, but do not delay treatment for the sake of biopsy.
- A negative biopsy does not exclude temporal arteritis.
A positive result helps to prevent later doubts about diagnosis, particularly if treatment causes complications.

**Treatment**

**Principles of treatment**

The mainstay of treatment in PMR/TA is that of oral corticosteroid therapy. The dose of steroid may be regarded as a balance between the risk of relapse and complications of the disease process itself versus the risk of steroid-induced side effects. As the risk of arteritic complications is highest in temporal arteritis then these patients should receive higher initial doses of steroid. Cases of temporal arteritis giving rise to visual symptoms have the highest risk of permanent blindness and it is not recommended that geriatricians attempt to manage these patients, who should be referred immediately to an ophthalmology consultant. The further management of these cases will not be discussed further in this review.

There are numerous issues surrounding corticosteroid therapy, including the correct starting dose and duration of treatment. Unfortunately there is only limited trial evidence to resolve these issues with no large prospective controlled studies.
Starting doses
A starting dose of between 10 and 20 mg of prednisolone for polymyalgia rheumatica is recommended [6, 16, 17]. A retrospective study of 132 patients advocated the use of 10 mg, whereas a prospective study of 49 patients found higher relapse rates over the first 2 months with 10 mg compared with 20 mg [18]. The guidelines that have included available evidence were published in 1993 by the Drugs and Therapeutic Bulletin (DTB) [19], and recommended a dose of 15 mg for the first month.

Similarly, only a small amount of evidence exists for the management of TA. There are two retrospective and two prospective studies included in the DTB guidelines that suggest a starting dose of 20–40 mg prednisolone for 8 weeks in TA without visual involvement. However, in clinical practice, particularly in those with visual impairment, higher starting doses (40–60 mg) of steroids are generally advocated.

Continuing steroid therapy
The continuing management of steroid therapy is based principally on clinical experience alone, with no studies studying the detail of steroid dosages beyond the first 2 months. The DTB provides a clear suggested regime and divides treatment into four phases: initiation, initial reduction, maintenance and final reduction. Prospective evaluation of these recommendations in an appropriate clinical setting with patients who are managed by rheumatologists and/or geriatricians is required.

Duration of therapy
There is some evidence from observational and immunological studies to suggest duration of therapy. Around 50% of patients with either PMR or TA are able to discontinue steroids around 2 years, whereas some need treatment for 4 years and a minority need to continue low doses of prednisolone on a long term basis [14, 20].

Steroid side effects
Whilst steroid treatment effectively suppresses the inflammatory process, the incidence of serious steroid-induced side effects has been reported to be as high as between 20 and 50% [19]. The risk of side effects has been shown to increase with high initial dose, high maintenance dose, high cumulative dose and prolonged therapy [15]. Older individuals, frequently with comorbidity, may be at particular risk. Specifically, The National Osteoporosis Guidelines state that individuals on > 7.5 mg of prednisolone for more than 6 months should be given osteoporosis prophylaxis [21]. The steroid doses recommended in the guidelines for both PMR and TA would indicate that all patients should be given osteoporosis prophylaxis at the onset of treatment.

Conclusion
It is not clear from guidelines which clinicians should best manage PMR/TA. Geriatricians are not mentioned specifically. However, the DTB guidelines suggest that uncomplicated PMR may be managed by general practitioners [19]. It is suggested referral to a rheumatologist be considered in cases of temporal arteritis, especially where there are atypical features, doubt surrounding the diagnosis or problems with steroid therapy or side effects.

Figure 1 shows a suggested management algorithm for diagnosis and management of PMR/TA specifically applicable to geriatricians, and attempts to amalgamate the best available evidence, even though this is scanty. There is much need for further prospective investigation and large randomised controlled trials to fully clarify a host of outstanding issues to improve the care of older people with these conditions.

Key points
• Much of the evidence for diagnosis and management of polymyalgia rheumatica and temporal arteritis comes from studies in younger age groups.
• Rheumatologists have predominantly developed available guidelines and whether they are appropriate to the patients seen by geriatricians requires further study.

References


Received 7 March 2002; accepted in revised form 11 December 2002