Distal musculoskeletal manifestations in polymyalgia rheumatica

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ABSTRACT

In polymyalgia rheumatica (PMR) the marked and distinctive symptoms of proximal aching and stiffness have tended to draw attention away from the distal musculoskeletal manifestations which also occur in this syndrome. Peripheral manifestations are present in about half of all cases of PMR and include joint synovitis, diffuse swelling of the distal extremities with or without pitting edema, tenosynovitis and carpal tunnel syndrome. Awareness of these findings will help to facilitate the proper diagnosis and institution of appropriate therapy for this disease.

The hallmark for the diagnosis of polymyalgia rheumatica (PMR) is the proximal involvement characterized by marked aching and morning stiffness in the shoulder and pelvic girdles and in the neck (1-3). The prominence assigned to these proximal symptoms has probably overshadowed the less well characterized and more variable distal musculoskeletal manifestations. However, Bruce in the first published description of PMR in 1888, noted distal findings in 2 of his 5 patients (4). Case I had symptoms "affecting his hands and feet as well as his wrists and ankles". Case IV had symptoms that laid "hold of his arms, shoulders, and hands which swelled and stiffened, and lost so much strength that he could not put his hand to his mouth and had to be fed at meals like a child".
Successively, Bengt Hamrin accurately described in 1972 the presence of distal extremity swelling with pitting edema in 25/93 (27%) PMR patients (5).
Despite these descriptions, distal symptoms have been underestimated and often a diagnosis of rheumatoid arthritis (RA) or other syndromes is made to explain these findings when they occur.
In a prospective follow-up study on 177 patients we determined the frequency and the characteristics of distal musculoskeletal manifestations in PMR (6). Distal findings occurred in approximately half of these cases of PMR. They were characterized by inflammatory involvement of the distal articular and tenosynovial structures.
A non-erosive, self limited, asymmetric peripheral arthritis (predominantly affecting the knee and wrist) was found in 25% of the patients. These data were in keeping with the results of previous studies which demonstrated the presence of peripheral synovitis in a percentage of PMR patients oscillating between 20-30% (7). The occurrence of peripheral arthritis, particularly in both hands, may create some difficulties in the differential diagnosis between PMR and elderly onset seronegative RA (EORA). Some patients with EORA present with a benign symmetric synovitis characterized by a prompt and complete response to corticosteroids (CS) and a clinical course similar to PMR (8). Healey noted patients who developed episodes of PMR and EORA at different times during their follow-up (9). These patients have a clinical condition quite different from "classic" RA that most closely resembles PMR.
A recent study from the Mayo Clinic examined the musculoskeletal manifestations in a population-based cohort of patients with giant cell arteritis (GCA) followed over 42-year period (10). Six of 128 GCA patients identified during the study period satisfied the criteria for RA. Four of them were seronegative for rheumatoid factor and never developed joint erosions or extra-articular features of RA during their follow-up over a number of years. They experienced multiple separate episodes of symmetrical arthritis, proximal symptoms of PMR, and distal extremity swelling with pitting edema. All of these manifestations responded rapidly to CS without any disease progression. A diagnosis of PMR or RA could therefore be made in the same patient at different times depending on the clinical expression of the disease. However, we feel that these manifestations represent part of the broader clinical spectrum of PMR.
In addition to peripheral joint synovitis,

an acute carpal tunnel syndrome with clinical evidence of wrist flexor tenosynovitis was observed in 14% of the patients (6). Moreover, a well-defined distal tenosynovitis was recorded in 3%. The sites of tenosynovial inflammation were the extensor hand tendons, peroneal tendons and posterior tibial tendon.

Distal extremity swelling with pitting edema over the dorsum of the hands and wrists, the ankles, and the tops of the feet was present in 12% (6). It might be either unilateral or bilateral with predominant involvement of the upper limbs. The distribution of edema was prevalently along the course of the tenosynovial structures (11). Hand and foot magnetic resonance imaging (MRI) confirmed the clinical impression that inflammation of the tenosynovial sheaths is the hallmark of this condition. Joint synovitis was more difficult to appreciate at clinical examination due to the extensive swelling involving the hands and feet. However, hand MRI through the metacarpophalangeal joints also showed the presence of joint effusion (Fig. 1) (7,12).

All distal symptoms responded promptly to corticosteroids. No evidence of joint deformities, erosions, or the development of RA was observed during the 39-month follow-up period.

Distal swelling and edema observed in PMR patients are similar to those observed in patients with remitting, seronegative, symmetrical synovitis with pitting edema (RS3PE) syndrome. This condition was described by McCarty in 1985 (13) as being characterized by an acute onset bilateral symmetrical synovitis involving predominantly the wrist, the carpus, the small hand joints, and the flexor digitorum sheaths, associated with marked pitting edema of the dorsum of the hand. Patients are persistently seronegative for rheumatoid factor and the articular complaints respond rapidly to small doses of corticosteroids.

We performed a 5-year prospective follow up study to investigate the relationship between "pure" RS3PE syndrome, and PMR with and without distal pitting edema (14). No significant differences in the demographic, clinical, immunogenetic and MRI features were found between 23 patients with "pure" RS3PE, 156 patients with PMR without pitting edema and 21 patients with PMR with pitting edema.

These similarities and the concurrence of the two syndromes suggest that these conditions may constitute part of the same disease and that the diagnostic labels of PMR and RS3PE syndrome may not indicate a real difference.

Therefore, the involvement of extra-articular synovial structures is a predominant part of the distal inflammatory process of PMR. This distinctive pattern of extensive, self-limiting inflammation of extra-articular synovial membranes may represent the common link between PMR and RS3PE syndrome and some cases of seronegative RA in the elderly, and may have important implications for a reclassification of inflammatory arthritis in older persons.

In summary, distal musculoskeletal manifestations are common in PMR. Peripheral synovitis is most frequent, but distal symptoms of diverse types such as carpal tunnel syndrome, isolated tenosynovitis and distal pitting edema often occur. Physicians need to be aware of these various manifestations in order to make a prompt and correct diagnosis and institute early the appropriate therapy.

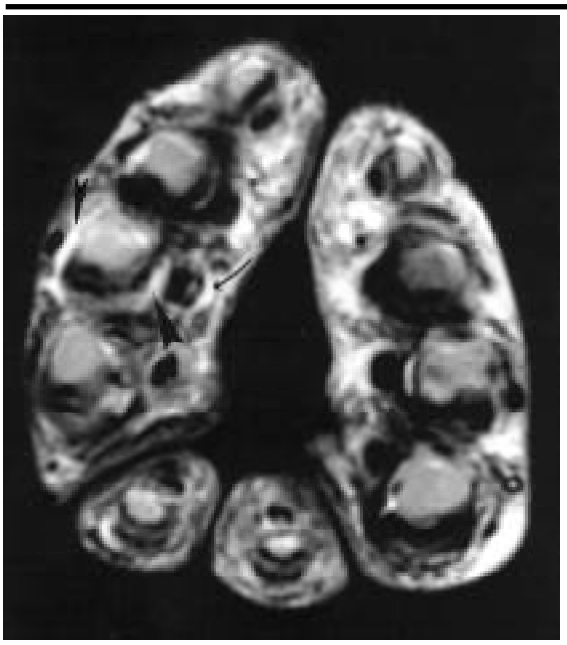

Fig. 1. An axial T2 weighted section through the metacarpophalangeal joints shows fluid collection in the flexor synovial sheaths of the third digit of the right hand (curved arrow) and joint effusion of the corresponding joint (large and small arrowheads).

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