Case Report

Development of Polymyositis After Long-Standing Rheumatoid Arthritis

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ABSTRACT. Rheumatoid arthritis (RA) and polymyositis (PM) are distinct clinical syndromes. The concurrent diagnoses of RA and PM in the same patient are rare. We describe a patient who developed outright PM after 16 years of well established RA, review the literature, and highlight the need to consider a broad base of differentials including PM in the diagnosis of muscle weakness in RA. (J Rheumatol 2006;33:362–3)

Key Indexing Terms:
RHEUMATOID ARTHRITIS POLYMYOSITIS

Rheumatoid arthritis (RA) and polymyositis (PM) are distinct clinical syndromes[1,2]. When myositis occurs in RA, it is frequently secondary to therapy with steroids or disease modifying medication[3]. The concurrent diagnoses of RA and PM in the same patient are rare. We describe such a case and review the current literature.

CASE REPORT

In 1986, a 26-year-old black woman developed an inflammatory arthritis of the small joints of the hands and positive serology (rheumatoid factor 99.4 IU). Based on clinical, radiographic, and laboratory findings, she was diagnosed with RA.

Treatment included corticosteroids, high dose aspirin, and subsequently intramuscular gold injections, which were discontinued due to adverse effects. Penicillamine and chloroquine were also tried, but the patient discontinued each medication due to "malaise." Her RA continued to be active, requiring 2 hospitalizations.

The patient came to our attention in 1988. She had 35 tender and 12 swollen joints, and at least 2 deformities. She was treated with local and systemic oral corticosteroid and began weekly oral methotrexate. Unfortunately, she was not compliant, and had frequent and severe flares of disease. In 1995, azathioprine was initiated.

In 2002 she complained of severe neck pain. Radiographs showed instability at C1/C2 level, with flexion-extension views demonstrating 12 mm of excursion. Neural foramina were intact. Magnetic resonance imaging showed an atlantodental interval of 5 mm. She was urgently referred to neurosurgery, but refused fusion.

In October 2002 she complained of bilateral hand, arm, and leg muscle weakness. She also described an insidious decrease in exercise tolerance, leg spasms, and difficulty initiating swallowing. She was noted to have profound proximal weakness and "Velcro" rales at the lung bases. No rash was detected.

Her creatinine kinase (CK) was 797. Extractable nuclear antigen (ENA) testing was positive for anti-Ro antibodies, but all other ENA were negative, including anti-Jo-1, anti-RNP, anti-Sm, and anti-Scl70. Review of her antinuclear antibody levels showed a variance between 1:40 (homogenous) and 1:160 (speckled), although her most recent testing was reported as inconclusive. Electromyographic studies showed a diffuse myopathic process without associated neuropathic changes. A muscle biopsy was performed, and confirmed the typical pathologic changes of PM. A high resolution computed tomography (CT) scan of the lungs showed bilateral peripheral interstitial disease with honeycombing and ground glass.

The corticosteroid dosage was increased, achieving symptomatic improvement and a decrease of CK to normal. She continues to refuse treatment with steroid-sparing agents.

DISCUSSION

This case represents a very unusual clinical evolution from long-standing seropositive RA to biopsy-proven inflammatory PM. The concurrence of RA and PM is uncommon, although some patients have an "overlap" syndrome with features of both diseases[4]. In our case, the patient had well established seropositive erosive RA for 16 years before developing PM.

There are few published case reports detailing the development of PM in patients with long-standing RA. Pitkeathly, *et al*[5] describe 3 cases of PM in RA, of which 2 patients developed PM after RA of several years' duration. Miro, *et al*[6] found biopsy evidence of PM in 1/21 patients with RA and symptoms of weakness, suggesting an incidence rate of about 5%. Agrawal, *et al*[7] described 2 of 23 patients with RA who had muscle biopsy findings consistent with PM.

Patients with anti-Jo-1 associated myositis have been reported to have inflammatory and subluxing joint changes, mimicking RA. O'Neill and Maddison[8] described a patient positive for anti-Jo-1 antibody who developed myositis after several years' duration of RA. These patients are usually classified as having "overlap" connective tissue disease. Anti-Jo-1 antibodies are also associated with pulmonary inflammation and fibrosis. Our patient developed both

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myositis and pulmonary fibrosis in the absence of anti-Jo-1, suggesting they were not part of an overlap syndrome.

Medications used in the treatment of RA may precipitate myositis, including d-penicillamine and corticosteroids[3]. Our patient received d-penicillamine briefly, although at least 15 years before the onset of myositis. She used corticosteroids as the main form of control for her RA, but her biopsy excluded steroid myopathy.

Despite the well known systemic nature of RA, characterization of muscle pathology remains enigmatic. Many patients with RA have muscular complaints, but it is difficult to separate disuse or atrophy symptoms from other muscle pathology. Muscle weakness in RA is most commonly attributable to rheumatoid cachexia, likely from the catabolic effects of circulating inflammatory cytokines[9]. Additionally, rheumatoid vasculitis can affect muscle strength.

Halla, *et al* characterized the clinical and histological features of rheumatoid myositis in 1984[10]. Their study suggested that rheumatoid myositis was a distinct clinical entity consisting of muscle fiber necrosis and mononuclear cell infiltration. This entity appeared to occur in patients with an erythrocyte sedimentation rate (ESR) disproportionately higher than their disease activity.

It has been noted that patients with RA often have low levels of serum CK on laboratory testing, often below the normal range[7]. This suggests that patients with even minor rises in CK levels merit investigation for the presence of myositis.

There have been few studies that have analyzed muscle biopsy specimens from patients with RA. The estimated prevalence of symptomatic muscle involvement has varied widely, from 6% to greater than 70%[7,10].

Little is known regarding the survival of patients with idiopathic inflammatory myopathies. A recent study by Danko, *et al*[11] reviewed 162 cases of idiopathic inflammatory myopathy diagnosed between 1976 and 1997. In this study, the most frequent causes of death were cardiac and pulmonary complications. Global survival rates were 95% at one year, and 92% and 89% for 5 and 10 years, respectively. Mortality was higher in the first year after diagnosis than in subsequent years. The authors noted that they found higher survival rates than previously reported, and suggested that this was due to newer cases, fewer paraneoplastic cases, improved diagnosis, and improved therapy. We have not found prognostic information for patients who develop PM after long-standing RA.

Our case illustrates a rare phenomenon in rheumatology: the development of outright polymyositis after 16 years of well established RA. Electromyelography and muscle biopsy were particularly useful in our case. Muscle weakness in RA is often multifactorial, but our case reminds the clinician of the need to consider a broad base of differentials, including PM.

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