Systemic lupus erythematosus presenting as polymyalgia rheumatica in the elderly

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SUMMARY Three patients who developed systemic lupus erythematosus (SLE) over the age of 60 are described. All patients presented with a clinical syndrome typical of polymyalgia rheumatica. In two cases there was an underlying myositis.

Key word: myositis.

Variation in the clinical presentation is a striking characteristic of SLE. This depends on the spectrum of organ involvement and fluctuation of disease activity. Its features blur with those of other connective tissue diseases, so that diagnosis may be difficult. SLE in the elderly is particularly variable in its clinical pattern and may differ from the disease seen in younger patients.¹ Its presentation may therefore be very atypical. Whether this is because it is a different disease process in the elderly or a modification by age of the underlying disease is unknown. Recognition of unusual presentations of connective tissue diseases in the elderly is important to allow appropriate effective therapy. We describe three cases of SLE presenting with a polymyalgia rheumatica-like syndrome, two of whom were found to have an underlying myositis.

Case reports

CASE 1

A 76 year old Caucasian man presented with proximal muscle pain and stiffness associated with marked weight loss which had developed over four months. He had had mild Raynaud's disease two years previously. Examination was unremarkable, and in particular there was no evidence of muscle weakness. Laboratory investigations showed a raised plasma viscosity of 1.94 (normal range 1.54–1.72) and a normochromic normocytic anaemia. Serology showed a weakly positive anti-nuclear antibody (ANA) titre (40 IU) but normal DNA binding. The serum creatine phosphokinase

(CPK) and transaminase levels were normal. The diagnosis of polymyalgia rheumatica was made, and he was treated with prednisolone 15 mg a day initially, which was subsequently reduced to a maintenance dose of 5 mg a day. Despite initial dramatic improvement in his muscle stiffness and pain he became insidiously weaker and was readmitted two years later with profound muscle weakness and wasting, predominantly affecting the proximal and bulbar muscles. Laboratory investigations showed a viscosity of 1.92. Haemoglobin was 126 g/l. He had a polymorphonuclear leucocytosis of $19.9 \times 10^9/l$ (normal range $4-11 \times 10^9/l$) but a lymphopenia of $0.6 \times 10^9/l$ (normal range $1.5-4.0 \times 10^9/l$). C reactive protein was raised at 0.16 g/l (normal <0.01 g/l). Creatine phosphokinase and transaminases were normal. Serology showed a raised ANA titre of 640 IU, DNA binding of 30% (normal range $<20\%$), but no precipitating antibodies to soluble non-histone antigens were detected. Raised anticholinergic receptor antibody titres (46.7, normal <2) were noted, but both neostigmine and pyridostigmine stimulation tests were negative. Muscle biopsy showed marked atrophy of all fibre types, with replacement fibrosis and a mild inflammatory mononuclear cell infiltrate. A skin biopsy specimen from uninvolved skin from the extensor aspect of the forearm showed granular deposition of IgM and C3 at the dermatoepidermal junction.

A diagnosis of myositis complicating SLE was made. Prednisolone was increased to 30 mg a day and methotrexate added. After a single 5 mg dose of methotrexate he developed an aplastic anaemia which was complicated by septicaemia, and he died. Postmortem examination confirmed the extensive muscle involvement (Fig. 1).

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Fig. 1 Striated muscle specimen taken after death showing extensive fibrosis and muscle fibre atrophy. (Haematoxylin and eosin).

CASE 2

A 62 year old Caucasian woman developed severe pain and stiffness in the upper and lower limb girdle overnight. A diagnosis of polymyalgia rheumatica was made, and the symptoms resolved over one month with treatment with flurbiprofen 50 mg three times a day. Three months later the symptoms recurred, now associated with pain in the small muscles of the hand, dyspnoea, and pleuritic chest pain. On examination she had mild proximal muscle weakness but no wasting, synovitis of the second and third metacarpophalangeal joints, proximal interphalangeal joints, both wrists, and right knee, and bilateral coarse crepitations at both lung bases. Investigation showed a raised viscosity of 1.87, and normochromic normocytic anaemia, a leucopenia with a white cell count of $2.4 \times 10^9/l$, of which $0.4 \times 10^9/l$ were lymphocytes. Serology showed a positive ANA titre (164 IU), precipitating antibodies to the cytoplasmic ribonuclear protein, Ro (SSA), but DNA binding was normal. Schirmer's test was positive. Chest x ray showed pulmonary fibrosis, and pulmonary function test demonstrated a diminished transfer factor KCO of 1.4 (expected value 1.65). The serum CPK and transaminase levels were normal, but an electromyogram showed polyphasic potentials, and a muscle biopsy confirmed an interstitial inflammatory infiltrate diagnosing an underlying myositis (Fig 2). There was no evidence of renal, neurological, or skin disease. The diagnosis of myositis complicating another

connective tissue disease, possibly systemic lupus erythematosus, was made. She was treated with prednisolone 30 mg a day, with immediate clinical improvement.

CASE 3

A 73 year old Caucasian woman presented with sudden onset of severe muscle stiffness, myalgia, and weight loss. She had become increasingly dyspnoeic and had developed Raynaud's phenomenon in association with gritty eyes and marked hair loss over the previous year. Ten years previously she had had a similar illness, diagnosed as polymyalgia rheumatica and treated for a year with prednisolone. This had gone into remission but recurred five years previously and was then associated with a positive ANA (titre 1 in 40), in addition to a high erythrocyte sedimentation rate and a normochromic normocytic anaemia. Again she had responded well to corticosteroid therapy.

On admission she had diffuse alopecia, basal pulmonary crepitations, but there was no evidence of muscle weakness, wasting, or skin rash. Laboratory investigation showed a raised plasma viscosity of 1.95 with a normochromic normocytic anaemia. CPK and transaminase levels were normal. Her ANA titre was 40 IU, with a deoxyribonucleic acid binding of 30%. She had precipitating antibodies to Ro(SSA) and La(SSB). A cold test confirmed she had Raynaud's disease, and Schirmer's test was positive. Salivary gland biopsy confirmed Sjögren's syndrome. Chest x ray showed diffuse pulmonary

Fig. 2 Striated muscle biopsy specimen showing cellular infiltration and muscle degeneration (H and E).

infiltration consistent with fibrosis. The gallium scan was negative, bronchial lavage was hypocellular, and pulmonary function test showed a diminished KCO of 0.4 (predicted value 1.56).

She was treated with prednisolone 30 mg a day, which produced a dramatic improvement in her symptoms and enabled her to be maintained on a level of 5 mg a day. She has remained well for 18 months.

Discussion

These three cases illustrate how polymyalgia rheumatica can be the presenting feature of a connective tissue disease and in two patients showed how this can mask an underlying myositis.

Systemic lupus erythematosus is increasingly recognised as presenting in the elderly. A number of studies show that between 10 and 20% of patients present over the age of 55.¹⁻⁷ These studies have also recognised that the clinical disease pattern is different in the elderly than in younger patients. Its onset is more insidious, and there is less cutaneous, renal, and central nervous system disease. Lung disease is often very prominent, but its course appears to be more benign. This pattern of disease overlaps Sjögren's syndrome. Indeed, although one case we describe fully satisfies the revised American Rheumatism Association diagnostic criteria for systemic lupus erythematosus⁸ and the other two have characteristic clinical and serological markers, they do show features of Sjögren's syndrome. Two patients had keratoconjunctivitis sicca and antibodies to the cytoplasmic protein Ro(SSA). These antibodies are found in systemic lupus erythematosus in a frequency varying between 25 and 40%⁹⁻¹¹ and also in up to 70% of patients with primary Sjögren's syndrome^{12 13} but are rare in other conditions.

Muscle disease is common in systemic lupus erythematosus.¹⁴ In myositis, myopathy and myoasthenic syndrome have been described. Foad *et al* first described five elderly patients in whom a polymyalgia rheumatica-like syndrome had been the presentation of systemic lupus erythematosus.¹ In none of those patients, however, was there underlying muscle disease. Myopathy presenting with marked weaknesses also has been described as a presentation of elderly patients with Sjögren's syndrome,¹⁵ but polymyalgia rheumatica has not been described as a presenting feature of this myopathy. In the three cases we describe all present with a polymyalgia rheumatica syndrome. In one case there was no evidence of underlying muscle disease, but one subsequently developed a severe myositis, which was possibly present earlier but masked by the features of polymyalgia rheumatica.

The third had evidence of myositis at the time when she presented with clinical syndrome of polymyalgia rheumatica. In one case described here these antibodies were present, but the failure of the patient to respond to either neostigmine or pyridostigmine testing makes it unlikely they had any clinical significance.

Polymyalgia rheumatica is typically associated with giant cell arteritis¹⁶ and may be related to other connective tissue diseases, in particular rheumatoid arthritis, but this relationship remains uncertain.¹⁷ It has been described in association with a positive antinuclear antibody titre but no other clinical evidence of SLE.¹⁸ The cases we describe, together with those described by Foad, suggest that the syndrome of polymyalgia rheumatica may also be associated with the development of systemic lupus erythematosus. Two of the cases show that a polymyalgia rheumatica-like syndrome may be obscuring an underlying myositis. So with polymyalgia rheumatica, particularly if there are any features of a connective tissue disorder, or a poor response to treatment, the possibility of systemic lupus erythematosus should be considered. It is particularly important because an unrecognised underlying myositis may develop which will require appropriate steroid and immunosuppressive therapy.

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