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Case Report

Anti Jo-1 Myositis. 'Mechanic's hands' and interstitial lung disease

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The presence of myositis specific antibodies allows classification of polymyositis into clinically important subsets. Anti Jo-1 autoantibodies are associated with 'mechanic's hands' and interstitial lung disease.

We report three patients who presented with painful weak muscles, a violaccous rash on the extensor aspect of their hands, elevated muscle enzymes, electromyographic and muscle biopsy findings typical of polymyositis. All three patients had anti Jo-1 autoantibodies, 'mechanic's hands' and evidence of interstitial lung disease on CT scan. An incomplete response to corticosteroids was observed in all three patients and additional immunosuppression was required.

CASE REPORT 1

A 41-year -old unemployed man was referred by his general practitioner to a general physician. He presented with shoulder muscle stiffness and weakness. On questioning he was found to have breathlessness on walking two hundred yards. He also described symptoms of Raynaud's phenomenon. On clinical examination he had a heliotrope skin rash over the extensor aspect of his metacarpal-phalangeal and interphalangeal joints. Fissuring and cracks were noted on the distal digital skin pads on his thumbs and fingers. A proximal muscle weakness of grade 4/5, was noted in his upper limbs. Respiratory examination was reported as unremarkable and without signs of cardiac insufficiency.

Serum creatine kinase was elevated at 1467 IU/L (normal range 0-180 IU/L). Subsequent electromyography and histological findings on muscle biopsy showed typical features of inflammatory myopathy. The patient was commenced on 40mg of prednisolone. An incomplete response was noted in terms of symptoms and muscle enzymes (creatine kinase ranged from 539-928 IU/L).

The patient was subsequently referred to a consultant rheumatologist where anti Jo-1 autoantibody testing was found to be positive and CT scan of chest revealed predominantly mid and lower zone interstitial pattern markings with some 'ground glass' appearances. On pulmonary function testing, transfer factor per litre of alveolar volume (KI.CO) was found to be 1.58 m mol min^{-1} kpa^{-1} (94% predicted). Azathioprine was commenced in addition to corticosteroids.

CASE REPORT 2

A 40-year-old housewife was referred by her general practitioner with a gradual onset of neck, shoulder and thigh pains. She noticed while playing badminton that her serve 'wasn't as strong'. Over this period of time she also described breathlessness. On examination, she had a heliotrope rash over the extensor aspect of her metacarpal-phalangeal/inter-phalangeal joints and 'mechanic's hands' (Figure 1). She had mild proximal muscle weakness, graded 4+/5 in her shoulder and pelvic girdles. Fine bibasal early

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inspiratory crepitations were heard on auscultation.

Serum creatine kinase was 1518 IU/L (normal range 0-180 IU/L). Anti Jo-1 autoantibody testing was positive. Subsequently an electromyogram revealed mild myopathic changes and muscle biopsy revealed features in keeping with inflammatory myopathy. Early interstitial fibrosis was seen on CT scan of chest. K1.CO was measured at 2.01 m mol min^{-1} kpa^{-1} (113% of predicted).

This patient was commenced on 40mg of prednisolone daily. A moderate symptomatic improvement was observed but her serum muscle enzymes remained elevated. The addition of azathioprine coincided with a symptomatic improvement and normalisation of muscle strength and enzymes. Her corticosteroid therapy was slowly reduced to 7.5mg of prednisolone daily, without relapse.

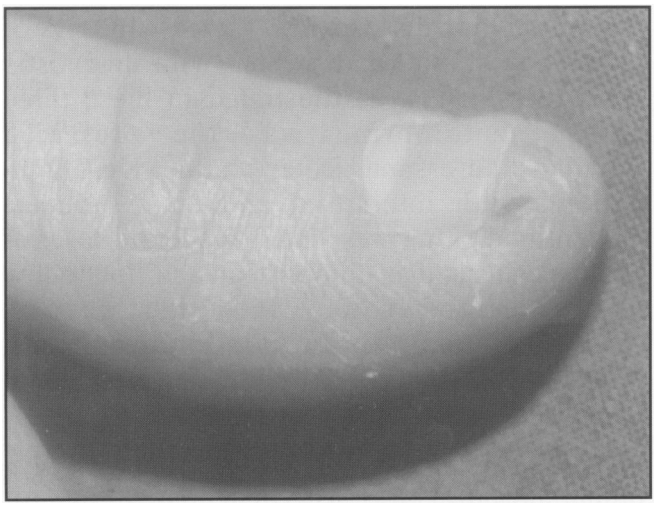

Fig 1. 'Mechanic's hands' – characterized by scaling and hyperkeratosis of the distal skin pad and lateral aspect of the fingers.

CASE REPORT 3

A 30-year-old school teacher was referred by his general practitioner. He reported an acute history of painful shoulders and 'roughening' of the skin at his fingertips. During this period he noted himself to be 'breathless' on exertion. On examination fissuring and cracking of the skin pads in his distal fingers, typical of 'mechanic's hands' was observed. A typical dermatomyositis rash was present on the extensor aspect of his hands. Muscle weakness was not observed. Fine bibasal early inspiratory crepitations were heard on auscultation of his chest.

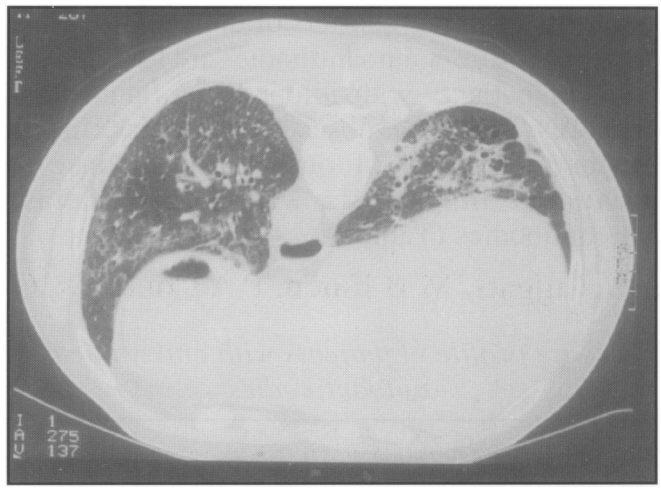

Fig 2. CT scan of chest revealing bibasal interstitial changes in a patient with anti Jo-1 autoantibodies.

Initial serum creatine kinase was mildly elevated at 296 IU/L. Other muscle enzymes were mildly elevated AST 51 IU/L (normal range 10-42 IU/L) and Aldolase 8.2 U/L (Normal range 0.5-3.12 U/L). Anti Jo-1 antibody testing was positive, confirming the clinical suspicion of antisynthetase syndrome. Mild myopathic changes were noted on his electromyogram. CT scan of his chest revealed bibasal interstitial changes (figure 2). K1.CO was 1.3 m mol min^{-1} kpa^{-1} (76% of predicted). An incomplete response to 40-60mg of prednisolone was observed. A clinical improvement was noted in terms of breathlessness and muscle enzymes with the addition of azathioprone. His corticosteroid therapy was reduced to 5mg of prednisolone daily, without relapse.

DISCUSSION

These cases illustrate that the presence of 'mechanic's hands' and serum antibodies to anti Jo-1, should alert physicians to the possible presence of interstitial lung disease.

The annual incidence of polymyositis/dermatomyositis has been estimated at 6 cases per million per year, with peak age of onset at 45-55 years.[1] Bohan and Peter's classification[2] of patients with idiopathic inflammatory myopathies, defined subgroups with specific clinical and pathological criteria, More recently, the detection of myositis specific autoantibodies, has been found to aid classification of defined subgroups with regard to clinical features, response to treatment and prognosis.[3]

Antisynthetase autoantibodies are the most common myositis-specific autoantibody. Anti Jo-1

accounts for three-quarters of the antisynthetase family. Anti Jo-1 autoantibodies are found in 20-30% of patients with idiopathic inflammatory myositis. Love *et al* observed particular clinical features, rapidity of onset and response to treatment in patients with antisynthetase autoantibodies (Table).[3]

TABLE

Disease profile of patients with antisynthetase autoantibodies

Sex ratio – Female/Male	1.7
Age at diagnosis	4th decade
Onset	Acute
Major clinical features	Interstitial lung disease Fever Raynauds phenomenon 'mechanic's hands'
Steroid response	Moderate
Mortality (%)	20

Anti Jo-1 myositis usually has an acute onset in the spring. Patients with antisynthetase autoantibodies, compared to patients without these autoantibodies, have significantly more frequent distinctive clinical features – 'Mechanic's hands', non-erosive arthritis, fever, Raynaud's and interstitial lung disease. As illustrated in *fig. 1*, 'Mechanics hands' is characterised by scaling, fissuring and hyperkeratosis of the distal skin pad and lateral aspect of the fingers. 'Mechanic's hands' are present in approx. 70% of patients with antisynthetase autoantibodies.

Interstitial lung disease has been reported in approximately 90% of patients with antisynthetasc autoantibodies [3] and is an important cause of mortality. Asymptomatic patients may have radiographic manifestation of interstitial lung disease and breathlessness may be due to non-pulmonary problems. Respiratory muscle weakness, cardiac disease and aspiration pneumonia may also cause breathlessness in patients with inflammatory muscle disease.[9, 10, 11] However, the presence of 'mechanic's hands' and anti Jo-1 autoantibodies suggests that interstitial lung disease is the likely cause of breathlessness on exertion. Sauty *et al* reported four cases of patients who presented with interstitial lung disease without muscle involvement and whose serum was positive for anti Jo-1 autoantibodies.[4]

Patients with antisynthetase syndrome tend to have a moderate response to corticosterolds. Sixty percent of patients will experience a flare-up of their myositis during tapering of corticosteroid therapy.[3] Late relapses after initial remission appear to be more frequent in patients with antisynthetase syndrome.[5] Therefore corticosteroid therapy should be tapered slowly, and close surveillance is necessary to monitor for disease exacerbation. Because of the poor response to corticosterolds, immunosuppressive agents should be considered early in patients with antisynthetase syndrome. This may have benefits in terms of reducing steroid side-effects and possibly improved response.[6] A report by Joffe *et al* suggests that patients with antisynthetase autoantibodies may respond more favourably to methotrexate than to azathioprine.[7] Although there is no evidence that methotrexate pulmonary toxicity is more common in patients with interstitial lung disease, the development of this complication in a myositis patient with interstitial lung disease, may increase the risk of respiratory failure and present with diagnostic difficulties. Thus, as illustrated in these three cases, azathioprine was the preferred immunosuppressant. Deteriorating K1.CO in patients with interstitial and antisynthetase autoantibodies may respond to moderate doses of cyclosporine and azathioprine in addition to low dose corticosteroid.[4] In other reports cyclophosphamide in association with corticosteroids has shown a clinical and functional response.[8]

Patients with antisynthetase syndrome have a higher mortality rate (21%) than myositis overall (7%),[3] which is probably related to interstitial lung disease.

The physical sign of 'mechanics hand' is a useful sign for identifying patients with anti Jo-1 myositis. The presence of anti Jo-1 autoantibodies is particularly important because it alerts the clinician to interstitial lung disease and the fact that additional immunosuppression may be required.

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