Case report

Keratoderma blenorrhagica, arthritis, and polymyositis with cardiopulmonary complications

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SUMMARY A patient is described who had polymyositis with arthritis, keratoderma blenorrhagica, pulmonary fibrosis, and cardiac failure with a right bundle branch block. The cutaneous lesions on his palms and soles, considered to be specific for Reiter's syndrome, pointed to an overlapping of polymyositis with features of this syndrome. Findings typical of myositis were present. In addition a muscle biopsy showed sarcolemmal and endomysial deposits of IgG and IgM, and the serum level of IgM was elevated. Because skin lesions appeared at the height of symptoms in other organs, the possibility is raised that both might stem from a common immune process.

The usual cutaneous manifestation of polymyositis/dermatomyositis is localised or diffuse erythema over the face, shoulders, and arms. In children a pathognomonic 'heliotrope' rash is sometimes seen. Occasionally, raised and erythematous lesions, which later become atrophic and scaly, appear over elbows, knuckles, and medial malleoli.¹

We report a case of polymyositis with keratoderma blenorrhagica, an association which to our knowledge has not been described before. It is possible that this case represents an overlapping of polymyositis and features of Reiter's syndrome.

Case report

Until December 1979 a 53-year-old farmer had been in good health and had had no history of rheumatic, respiratory, neurological, or skin disease. In 1969 the patient had given up a 20-year habit of smoking a pack of cigarettes a day. In January 1979 a mass screening chest roentgenograph had been normal. In December 1979 swelling of the wrists developed. During the following month the joint symptoms grew worse and were accompanied by some fever, cough, and dyspnoea. Up to this time there had been no history of muscle weakness. In February 1980 he was

admitted to hospital because of symptoms and because a chest roentgenograph showed basal infiltrates (Fig. 1).

On admission the patient's condition was poor. He had fever (up to 39°C) and suffered from dyspnoea on even the slightest physical activity. Blood pressure was 160/90 mmHg and pulse rate 100/min. The heart sounds were normal and there were no frank signs of cardiac failure. Dry rales were heard, particularly

Fig. 1 Chest roentgenograph. Mainly basal, interstitial infiltrates consistent with pulmonary fibrosis.

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over the basal zones. His ankles, wrists, and hands were swollen and the joints generally painful. On admission his skin was healthy, but within a few days sharply demarcated florid lesions developed on the palms and soles, and to a slight extent on the neck. These lesions were indistinguishable from the keratoderma blennorrhagica of Reiter's syndrome (Fig. 2). Fig. 3 summarises the symptoms, some laboratory results, and corticosteroid treatment.

In February 1980 the erythrocyte sedimentation rate (ESR) was 26 mm/h, haemoglobin 13.9 g/l, and leucocytes $10.8 \times 10^9/l$ with some shift to the left in the differential count. Urine analysis showed 8–10 leucocytes per field; *Escherichia coli* was cultured in the first urine sample but not in the follow-up sample. Serum creatine kinase was markedly raised (2716 U/l), as were lactate dehydrogenase (2171 U/l), aldolase (70 U/l), aspartate aminotransferase (367 U/l), and alanine aminotransferase (268 U/l). Serum alkaline phosphatase was normal. Serum total proteins were diminished (49 g/l) and there was slight hypoalbuminaemia (20 g/l). The gammaglobulin level was slightly above normal, with the IgM level raised (3.3 g/l) and IgG, IgA, and IgE levels normal. Serum concentrations of C3 and C4 were normal.

The Waaler-Rose test for rheumatoid factor was negative (titre <32), as were tests for LE cells, anti-nuclear antibodies, and antibodies to yersinia and chlamydia. Sputum samples were negative for bacteria, fungi, and tubercle bacilli, and the Mantoux test (10 TU) was negative. Tests for kidney and thyroid function revealed no abnormality. Liver biopsy showed only fatty changes. Lung function tests, however, disclosed restriction and severe impairment of ventilation, the diffusion capacity for CO being only half normal (16.9 ml/min/mmHg). The patient was negative for HLA B27 but positive for B8.

Roentgenographs of hands, wrists, knees, and ankles showed only soft-tissue swelling. No active bony changes were seen, and none developed during follow-up.

In February 1980 the patient was put on oral prednisolone (20 mg daily) and indomethacin. Fever disappeared but respiratory and articular symptoms responded poorly. The lesions on the palms and soles waned slowly over the following months.

In April 1980 cardiac failure developed and the electrocardiogram (ECG) showed a right bundle branch block. The decompensation was controlled with digoxin and diuretics.

Fig. 2 Sharply demarcated erythematous, keratotic lesions (keratoderma blennorrhagica) on palms and soles.

Fig. 3 Summary of symptoms, serum enzyme levels, and dosages of peroral corticosteroids.

In May 1980 the patient complained of proximal muscle weakness which prevented him from lifting heavy objects and walking uphill. Towards the autumn joint symptoms again grew worse. Not only his hands but also his knees were now swollen, his hip joints ached, and bilateral Baker's cysts developed. Synovial fluid drawn from the right knee contained 2000×10^6 leucocytes/l, of which 94% were mononuclear.

Towards the end of 1980 the patient began to suffer from Raynaud's symptoms. In spring 1981 muscle enzyme levels started to rise. A needle electromyogram (EMG) taken in May 1981 showed abundant fibrillation and a bizarre series of potentials with extremely polyphasic and long units, mainly in the proximal muscles. Nerve conduction velocities were within normal limits. The findings were consistent with subacute polymyositis.

A biopsy was taken from the left anterior tibial muscle (Fig. 4) in May 1981. The size of the fibres was abnormally variable, with both round and flattened atrophic fibres concentrated along the periphery of the fascicles. Internal nuclei were numerous, and the degenerating fibres showed abnormalities of internal structure, such as 'moth-

eaten' changes and whorling of the intermyofibrillary network. There was moderate necrosis, phagocytosis, and regeneration. A scanty inflammatory infiltrate appeared in the endomysium. These changes were pathognomonic of polymyositis. Immunohistochemical studies with the peroxidase-antiperoxidase technique² revealed a striking accumulation of IgM on the muscle cell surface, in the endomysium, and along vessel walls. Increased amounts of IgG showed a similar distribution. Indirect immunofluorescence revealed deposits of IgG, scant deposits of IgM, but no complement C3.

In May 1981 the steroid dosage was increased to 32 mg of methylprednisolone daily, which quickly brought the muscle enzyme levels down to normal. In a follow-up EMG in September 1981 the unit potential abnormalities were unchanged, but the fibrillation potentials had virtually disappeared.

The patient has now been followed-up for more than 2 years. He is on a sickness pension because of dyspnoea due to pulmonary fibrosis and heart disease. He continues to suffer from Raynaud's symptoms but not from pain or swelling of the joints. Muscle weakness is minimal. Medication is digoxin, diuretics, and 8 mg of methylprednisolone daily.

Fig. 4 Muscle biopsy. (a) Abnormal variability of fibre size with perifascicular atrophy (routine ATPase at pH 9.4; $\times 50$). (b) Necrosis, phagocytosis, regeneration, and inflammation (haematoxylin-eosin; $\times 130$). (c) Cell surface/endomysial deposits of IgM as revealed by the peroxidase-anti-peroxidase technique. Inset: anti-IgM stained normal muscle ($\times 85$).

Discussion

The patient's disease fulfilled the criteria of definite polymyositis.^{3,4} He had (a) symmetrical proximal muscle weakness, although this was not the presenting symptom; (b) evidence of muscle degeneration with pathognomonic perifascicular fibre atrophy, inflammation, and regeneration; (c) a high serum level of creatine kinase; and (d) EMG findings consistent with myopathy.

The prominent extramuscular symptoms are probably also manifestations of polymyositis. Pulmonary fibrosis is estimated to affect up to 9% of patients with polymyositis.⁵ Cardiac involvement, due to inflammation or to fibrosis of the myocardium or both, is present in 10 to 69% if the electrocardiogram is used as the criterion. The commonest findings are arrhythmias and conduction blocks. Cardiac failure is uncommon.⁶ From 25 to 50% of patients with polymyositis have arthritis. It is usually mild, non-erosive, and typically confined to hands, wrists, and knees. Schumacher *et al.*⁷ have suggested that, as arthritis and pulmonary fibrosis often occur together, this combination of symptoms may represent a distinctive subset of myositis.

The presence of keratoderma blenorrhagica raises the possibility of an incomplete manifestation of Reiter's syndrome. Arthritis and typical skin manifestations were present, but there was no convincing

evidence of urethritis, no balanitis, and no conjunctivitis. Furthermore, unlike most patients with Reiter's syndrome, our patient was negative for HLA B27, but he was positive for B8, which is known to correlate positively with polymyositis.⁸ Overlap syndromes between polymyositis and other connective tissue diseases do occur, but we know of no reported overlapping between polymyositis and Reiter's syndrome.

A differential diagnosis of the skin involvement must also take psoriasis into consideration. The rupioid form of psoriasis in particular may be both clinically and histologically indistinguishable from the cutaneous manifestations of Reiter's syndrome. Cases have been reported in which a typical Reiter's syndrome with keratoderma blenorrhagica has progressed to typical psoriasis with either rheumatoid-like or psoriatic arthritis.⁹ Indeed, psoriasis and Reiter's syndrome may be related conditions. However, whereas Reiter's syndrome may be complicated by carditis, neither Reiter's syndrome nor psoriasis alone would explain the myositis in our patient.

In polymyositis, skin manifestations rarely extend to the hands. Stahl *et al.*¹⁰ have reported, however, on 8 patients with mixed connective tissue disease, dermatomyositis, or systemic lupus erythematosus (SLE), in whom myositis was associated with a non-pruritic, hyperkeratotic eruption accompanied by scaling, fissuring, and a hyperpigmentation that gave

the appearance of 'mechanic's hands'. The rash occurred symmetrically along the ulnar aspect of the thumb and radial aspect of the fingers. Involvement of palms and soles seems to be rare. In his view on the skin manifestations of dermatomyositis, Keil¹¹ cites the cases of Crevald (1929; large red spots on the palms), Pick (1935; bluish-violet noncircumscribed spots on the palms), and Wharf and Wilnes (1936; 'dermatitis' of the palms), but stresses that palm/sole involvement speaks in general for SLE rather than for polymyositis. The most similar to our patient seems to be a young man reported by Keil¹¹ who presented with a 'punctate type of keratoderma' on the palms—superficial, small, erythematous lesions marked by adherent scales and in some areas by atrophy. However, whereas Keil's patient had many features of polymyositis/dermatomyositis, a history of both topical and systemic exposure to lead compounds and the lack of objective evidence of muscle degeneration bring his taxonomic position into question.

The pathogenesis of the unusual skin condition in our patient can only be speculated on. The muscle biopsy showed evidence of an IgG and IgM mediated humoral immune response (compare Whitaker and Engel¹²), and the serum level of IgM was raised. Such immune aberrations are not a recognised pathogenetic feature of keratoderma blenorrhagica. The appearance of skin lesions at the height of symptoms in other organs does suggest, however, that both stem from a common immune process.

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