Acute dermatomyositis associated with a staphylococcal infection

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Summary

A case is reported of dermatomyositis developing in a woman with staphylococcal osteomyelitis. After treatment for the infection, the dermatomyositis resolved and no other cause for it has been found. Dermatomyositis has not previously been described in conjunction with a staphylococcal infection.

Introduction

Polymyositis and dermatomyositis may be associated with autoimmune phenomena and with malignancy. The initiating factors are unclear; viral infections have been proposed (Pearson and Bohan, 1977) and a few cases have been associated with bacterial and parasitic infections (Naidoo and Chan, 1975; Kegen, Kimball and Christian, 1974; Greenlee *et al.*, 1975; Samuels and Rietschel, 1976). Staphylococcal infection is recognized as causing pyomyositis by direct involvement of skeletal muscle, particularly in the tropics (Levin, Gardner and Waldvogel, 1971). No link has been drawn between staphylococcal infection and polymyositis or dermatomyositis. A case is reported of dermatomyositis associated with a septicaemia due to *Staphylococcus aureus* and osteomyelitis of the cervical spine.

Case history

A 49-year-old teacher was admitted with progressive pain and stiffness of the neck for 10 days. Four days before admission she had developed generalized stiffness of the joints and pain and weakness of the muscles. The left elbow had become red. The day before admission the legs became generally swollen, she was too weak to get out of bed without help and was breathless. Penicillin had been prescribed without effect.

Past medical history, social and family histories were not contributory. She was on no other drugs and there were no known allergies.

On examination she was unwell, flushed, dehydrated and dyspnoeic at rest, with a temperature of 37.6°C. The limbs were swollen with slight

pitting oedema but there was no facial oedema. Erythematous patches were noted over the face, neck, chest and limbs. The joints, particularly the elbows and knees, were swollen and tender on passive movement. There was marked generalized tenderness and weakness of the muscles particularly in the proximal groups. Muscles of the neck, tongue, jaw and respiratory muscles were involved and there was dysphagia and dysarthria. The reflexes were symmetrically diminished and the plantars were flexor; sensation was normal. Visual fields were full and there was no diplopia nor nystagmus. Fundoscopy was normal. The pulse was 120/min and regular, BP 160/70 mmHg, jugular venous pressure was not elevated, and a fourth heart sound was heard. In the chest there were crackles at both lung bases and in the left mid zone. Abdominal, rectal and pelvic examinations were normal, as were the breasts, the thyroid and the reticuloendothelial system. A clinical diagnosis of dermatomyositis was made.

Investigations showed the urine to be dark, cloudy, with red cells $200 \times 10^6/l$, WBC $20 \times 10^9/l$, protein +, sugar negative and granular casts; culture was sterile. Hb 11.8 g/dl, WBC $10.3 \times 10^9/l$ with a neutrophil leucocytosis, ESR was 119 mm/hr. Serum sodium was 130 mmol/l, potassium 5.62 mmol/l, creatinine 90 $\mu\text{mol/l}$, and blood sugar 3.8 mmol/l. Pco_2 was 3.46 kPa; Po_2 was 10.64 kPa, PEFR was reduced at 200 l/min (predicted 400-500 l/min). Chest X-ray showed patchy shadowing in both lung bases and mid zones. An ECG showed sinus tachycardia. The CPK was 340 i.u./l (normal 25-200 i.u./l); hydroxybutyrate dehydrogenase 440 i.u./l (normal 150-350 i.u./l). There was no urinary myoglobin. Total serum proteins were 53 g/l, albumin 19 g/l, plasma protein electrophoresis showed non-specific inflammatory change, and immunoglobulins were normal. Liver function tests were abnormal with gamma glutamyl transpeptidase 143 i.u./l, alanine aminotransferase 53 i.u./l, alkaline phosphatase 633 i.u./l, bilirubin 21 $\mu\text{mol/l}$. Four successive blood cultures grew *S.*

aureus resistant to penicillin but sensitive to flucloxacillin and fusidic acid. Phage typing was not performed because the organism did not fall into the groups usually used for typing. Staphylococcal antibodies measured 48 hr after admission were: anti- α haemolysin 16 i.u./ml (normal ≤ 2 i.u./ml), anti- γ haemolysin 32 i.u./ml (normal ≤ 4 i.u./ml), anti-micrococcal nuclease 16 i.u./ml (normal up to 4 i.u./ml). Viral studies were unremarkable. Barium meal and enema were normal and ANA, antibodies to extractable RNA and rheumatoid factors were negative. DNA and complement concentrations were normal.

Muscle biopsy from the left triceps showed changes on light microscopy of a myositis. Electron microscopy showed grossly abnormal fibrils with total disorientation of Z, A and F lines; myofibrils were indistinguishable. The sarcolemmal cells showed occasional myelin figures and fat globules were quite numerous. The changes were consistent with a myositis. An electromyogram performed after treatment was begun showed patchy abnormalities also consistent with a myositis.

The patient was given i.v. fluids and with rehydration the blood urea and urine microscopy returned to normal. Flucloxacillin was given i.v., and prednisolone, 60 mg daily was started orally. Within one week the temperature was normal, the patient was able to get out of bed unaided and by 2 weeks she could walk unaided. The peak flow rate was 400 l/min. The ESR was 23 mm/1 hr, the muscle and liver enzymes were normal and the anti-staphylococcal antibodies had fallen. The chest X-ray changes resolved, and the antibiotics were stopped after 15 days and the steroids halved.

Two weeks later, the neck was again painful. X-rays showed distortion of C4, 5, 6 and tomography suggested osteomyelitis. Anti-staphylococcal antibodies and the ESR were again elevated. Flucloxacillin 250 mg orally and fusidic acid 500 mg thrice daily orally were given for 6 months. Steroid therapy was reduced further to 10 mg on alternate days and stopped after 2 further weeks. At the end of 6 months she was well. The cervical X-rays had improved, power was normal in all muscle groups, the ESR was 15 mm/1 hr and the anti-staphylococcal antibodies had returned to normal limits. Muscle and liver enzymes remained normal.

After more than 2 years' follow-up there is no evidence of an underlying malignancy nor an autoimmune process.

Discussion

Polymyositis is regarded as an autoimmune inflammatory disease of skeletal muscle, dermatomyositis being a similar condition with skin involve-

ment (Bohan and Peter, 1975). The initiating factor may be an infection which then initiates a hypersensitivity response which becomes directed at skeletal muscle (Pearson and Bohan, 1977). In cases associated with malignancy both the myositis and malignancy may be independent effects of a single cause. Alternatively, an immune response directed against tumour cell antigens may cross-react with muscle fibres.

Viral infections have been implicated in this process, virus-like particles having been identified on electron microscopy of affected muscles taken from patients with myositis (Chou 1968; Ben-Bassat and Machtey, 1972; Greco, Askenase and Kashgarian, 1977).

Acute myositis has been associated with influenza (Mejlszenkier *et al.*, 1973; Barton and Chahub, 1975; Mason and Keller, 1975; Dietzman *et al.*, 1976) and influenza B infections (Mason and Keller, 1975). An initial direct toxic effect of the virus has been suggested and a host immune response may then develop, directed against skeletal muscle. A link with Coxsackie B infections is suggested (Travers *et al.*, 1977) and also typhoid fever (Naidoo and Chan, 1975) and toxoplasmosis (Kegen *et al.*, 1974; Greenlee *et al.*, 1975; Samuels and Rietschel, 1976).

The present case seems to be the first in which staphylococci have been implicated in polymyositis although they cause pyomyositis by direct involvement of striated muscle (Levin *et al.*, 1971).

The clinical evidence of dermatomyositis was strong with an acute onset of generalized muscle weakness, and tenderness involved particularly the proximal muscles and the neck. The rash was typical of dermatomyositis and there was evidence of joint involvement. There was no evidence of an underlying malignancy.

CPK and hydroxybutyrate phosphokinase concentrations were only mildly elevated but they may be normal in 10% of cases (Pearson and Bohan, 1977). Results of electromyographic studies performed during the recovery phase were compatible with a myositic process without evidence of a nerve conduction defect. Histology confirmed a myositis. There was no laboratory evidence of viral infection or an autoimmune process but demonstration of auto-antibodies to muscle tissue was not undertaken. Clinical and laboratory findings thus strongly support the diagnosis of dermatomyositis although without evidence of underlying malignancy or autoimmune disease. The findings of a staphylococcal septicaemia, the later discovery of osteomyelitis of the cervical spine and elevated staphylococcal antibodies were all evidence of a significant and deep-seated staphylococcal infection.

Rapid clinical improvement followed treatment

with loss of muscle tenderness, return of muscle power and subsidence of the erythematous rash. This was accompanied by sterilization of the blood, return of the staphylococcal antibodies to normal and the later healing of the cervical osteomyelitis.

Cases of acute post viral myositis seem to be self limiting (Mejlszenkier *et al.*, 1973; Barton and Chalhub, 1975; Mason and Keller, 1975; Dietzman *et al.*, 1976) and treatment of the infection in typhoid and toxoplasma polymyositis seems to improve the muscle weakness.

The association of dermatomyositis and staphylococcal infection in this patient raises the possibility (although this is difficult to prove conclusively) that the latter was the cause of the former. Response to anti-staphylococcal treatment alone would have been helpful in this respect but this was not thought to be justifiable. However, there was no relapse as the steroids were withdrawn.

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