CASE REPORTS

Dermatomyositis following chronic staphylococcal joint sepsis

Sally Lane, Michael Doherty, Richard J Powell

Abstract

A young man is reported with recurrent *Staphylococcus aureus* joint sepsis associated with dermatomyositis. His dermatomyositis failed to resolve on treatment with antimicrobial agents alone, indicating that if staphylococcal infection was the triggering event for the dermatomyositis then the subsequent process was apparently self perpetuating, requiring cytotoxic agents for its control. This case can be interpreted as possible further evidence for the triggering of autoimmune disease by infective agents.

The dermato-polymyositis complex is not a single disease entity and this heterogeneity presents problems not only of prognosis and treatment, but more importantly in the determination of the cause of the condition. Dermatomyositis or polymyositis may occur alone or in connection with a spectrum of connective tissue diseases: a smaller number of cases are associated with various malignancies. We present a young man with classical dermatomyositis, which apparently followed chronic staphylococcal joint sepsis.

Case report

In 1984 a previously fit 16 year old boy was admitted with a three week history of progressive right shoulder pain and fever. Examination showed right glenohumeral synovitis and mild Raynaud's phenomenon. Investigations included haemoglobin 103 g/l, white blood cell count (WBC) $11.1 \times 10^9/l$ (lymphocytes $2.0 \times 10^9/l$), erythrocyte sedimentation rate (ESR) 60 mm/h, and *Staphylococcus aureus* was cultured from the blood and the right shoulder aspirate. The shoulder was treated by surgical drainage and parenteral flucloxacillin and sodium fucidate. Ten days later he developed a tense haematoma of the shoulder requiring surgical evacuation; subsequent arteriography showed a false aneurysm of the posterior circumflex humeral artery. *S. aureus* sensitive to flucloxacillin was again grown from the joint and oral flucloxacillin was continued for three further weeks. At discharge his absolute lymphocyte count had fallen to $0.7 \times 10^9/l$, the wound still required daily dressings, and a bluish discoloration had developed over the dorsum of his right hand. Over the next few months he became generally worse, was unable to take part in active sport, and at times had difficulty in getting out of chairs.

Four months after initial presentation he was

readmitted with recurrence of his septic right shoulder and a strange affect, considered to be part of a reactive depression. A full blood count showed haemoglobin 119 g/l, WBC $6.3 \times 10^9/l$ (lymphocytes $0.8 \times 10^9/l$), ESR 70 mm/h. The right shoulder was re-explored, and a bleeding hole in the false aneurysm on the posterior circumflex artery sutured; treatment with flucloxacillin, tobramycin, and ceftazidime was started. Flucloxacillin was continued orally for a further three months, at which time his full blood count showed haemoglobin 101 g/l, WBC $6 \times 10^9/l$ (lymphocytes $0.4 \times 10^9/l$), and C reactive protein <20 mg/l.

Three years later he was referred to our unit with a swollen, painful left elbow, stiffness of hands and knees, and marked symptomatic Raynaud's phenomenon. Examination showed Gottron's papules over the metacarpophalangeal joints, a heliotrope rash, a swollen non-tender right proximal interphalangeal joint, and a left olecranon bursitis. *S. aureus* was aspirated from the left olecranon bursa. Investigations included haemoglobin 146 g/l, WBC $7.7 \times 10^9/l$ (lymphocytes $1.2 \times 10^9/l$), ESR 42 mm/h, C reactive protein 27 mg/l, creatine phosphokinase 127 IU/l (normal range (NR) 24-170); serum immunoglobulins measured IgG 30.1 g/l, IgA 7.7 g/l, IgM 1.3 g/l, (NR IgG 5-16, IgA 1.25-4.25, IgM 0.5-1.7) and IgG subclasses were normally represented. The autoantibody screen was negative, and, in particular, rheumatoid factors and antibodies to Jo 1 were not identified. Serum C3 and C4 were normal, but the plasma C3 conversion product (C3dg) was grossly increased at 26 units/ml (NR 5-13). A needle biopsy specimen from the quadriceps showed established perifascicular atrophy with two foci of interstitial inflammation: degenerate fibres and regenerating fibres were noted around the periphery of the fascicles. ATPase staining did not show type specific atrophy. This was considered consistent with dermatomyositis, though lymphoid infiltration was sparse. Radiographs of hands and feet showed isolated calcinosis in the right thumb soft tissues (figure). Viral screen, including antibodies to HIV were negative. Investigations for possible immunodeficiency showed normal numbers of peripheral blood T and B lymphocytes and T cell subsets; phagocytosis, killing, and opsonisation were likewise normal. There was no history of or evidence of any drug abuse.

The olecranon bursitis resolved on treatment with flucloxacillin. No spontaneous improvement in his dermatomyositis was noted over the next four months, and treatment was therefore started with azathioprine 150 mg daily, increas-

Department of Immunology,
Queen's Medical Centre,
Nottingham NG7 2UH
S Lane
R J Powell

Rheumatology Unit,
City Hospital,
Nottingham
M Doherty

Correspondence to:
Dr Powell.

Accepted for publication
25 July 1989

Calcinosis in the right thumb soft tissues.

ing to 250 mg daily. After three months a definite clinical improvement, both in general wellbeing and muscle strength, was recorded, and this improvement has been sustained. His asymmetrical small joint arthritis required local steroid injections.

Discussion

This case of classical dermatomyositis seems to have followed a chronic, partially treated staphylococcal septic arthritis. Such dermatomyositis must be distinguished from the acute focal suppurative myositis in topical areas consequent upon direct invasion of skeletal muscle by staphylococci. Only a single previous case of dermatomyositis associated with *S aureus* infection has been published, in which the septic focus, osteomyelitis, was treated with steroids in addition to antimicrobial agents from the outset, and the possible outcome from antibiotics alone is therefore unknown.¹ In our case steroids were not prescribed, and the introduction of cytotoxic drugs was delayed for three months after diagnosis. (In retrospect the delay might have been three years after the onset of dermatomyositis.) No improvement in muscle

strength occurred during prolonged antibiotic treatment: however, since the introduction of cytotoxic drugs there has been a very significant clinical improvement. If indeed *S aureus* infection was a triggering event in the development of polymyositis then a self-perpetuating mechanism had supervened.

Acute skeletal muscle inflammation may occur after a variety of acute viral infections, such as rubella² and influenza,³ and in association with infections by bacteria—for example, staphylococci⁴ and streptococci⁵; these are usually self-limiting or improve with appropriate antimicrobial drug treatment. The rarity of primary joint sepsis and connective tissue disease in a boy of this age suggests more than just chance concurrence. Although mild Raynaud's phenomenon antedated onset of joint sepsis in our patient, there were no other symptoms or signs to suggest a pre-existing connective tissue disease; it is noteworthy that lymphopenia was not present at the first admission but developed subsequently. One interpretation is that sepsis triggered autoimmune disease in a predisposed subject. The debate about possible infective agents triggering dermatomyositis has been strengthened recently by the demonstration of sequence homology between the amino acid sequences of *E coli*, histidyl-tRNA synthetase, and alanyl-tRNA and two proteins identified as autoantigens in polymyositis.^{6,7} The sequences are long enough to function as epitopes, and this molecular mimicry with infective agents might suggest a rationale for the skeletal muscle target in polymyositis.

Our patient failed to show any antibodies to cellular/cytoplasmic antigens, in particular Jo 1, but it remains a possibility that the *S aureus* organism triggered this autoimmune muscle disease by a related mechanism. Probably, both environmental and genetic factors govern autoantibody specificity, and this case is further evidence for the triggering of autoimmune disease by infective agents.

- 1 Haegerty A M, Byrom N P, Cookson J B. Acute dermatomyositis associated with a staphylococcal infection. *Postgrad Med J* 1981; 57: 796-8.
- 2 Landry M, Winkelmann R K. Tubular cytoplasmic inclusion in dermatomyositis. *Mayo Clinic Proc* 1972; 47: 479-92.
- 3 Middleton P J, Alexander R M, Szymanski M T. Severe myositis during recovery from influenza. *Lancet* 1970; ii: 533-5.
- 4 Altrocchi P H. Spontaneous bacterial myositis. *JAMA* 1971; 217: 819-20.
- 5 Barrett A M, Gresham G A. Acute streptococcal myositis. *Lancet* 1958; i: 347-51.
- 6 Walker E J, Jeffrey P. Polymyositis and molecular mimicry, a mechanism of autoimmunity. *Lancet* 1986; ii: 605-7.
- 7 Mathews M B, Bernstein R M. Myositis autoantibody inhibits histidyl-tRNA synthetase: a model for autoimmunity. *Nature* 1983; 304: 177-9.