

Short reports

Dermatomyositis and rapidly progressive fibrosing alveolitis

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Fibrosing alveolitis is a rare complication of dermatomyositis.¹ Since the association between the two conditions was first described² about 40 cases have been published. Most patients with fibrosing alveolitis associated with dermatomyositis respond well to corticosteroids. A recent report³ suggests that early, potentially reversible lesions occur in the lungs, which if not treated at an early stage are followed by irreversible, steroid-resistant fibrosis. We report two patients with dermatomyositis who developed rapidly progressive lung disease despite early immunosuppressive therapy.

Case reports

Patient 1 A 58-year-old civil engineer was referred to a rheumatology clinic in August 1979 with a six-week history of pain and swelling of the small joints of the hands. He had also noticed a red, scaly rash on the dorsum of both hands and purple discoloration and swelling of his eyelids. Clinical examination confirmed a symmetrical inflammatory polyarthropathy of the small joints. His chest radiograph was normal. A presumptive clinical diagnosis of dermatomyositis was made. Three weeks later he developed dyspnoea on exertion and fine crackles were heard at both bases. At this stage his chest radiograph (fig 1) showed only early bilateral interstitial changes. Lung volumes were decreased and his carbon monoxide transfer factor was $2.05 \text{ ml min}^{-1} \text{ mm Hg}^{-1}$ (predicted normal 9.3 ± 1.7). The serum lactate dehydrogenase level was raised to 559 units/l (normal range 72-395) but creatine kinase was normal at 58 units/l.

A muscle biopsy showed evidence of a mild focal myositis. Haemolytic complement (CH_{50}) was reduced at 32% (normal range 50-150% normal human pool) but circulating immune complexes were not detected. A diagnosis of dermatomyositis with fibrosing alveolitis was made and he was discharged at the end of October 1979 on 40 mg of prednisolone a day. Three weeks later he was readmitted with considerable deterioration of his dyspnoea. Arteritic lesions were present on his elbows and fingers. A chest radiograph showed progression of the bilateral basal shadowing. Arterial blood gas analysis (during the breathing of air) confirmed hypoxic respiratory failure (Po_2 5.58 kPa, Pco_2 3.73 kPa, H^+ 30 nmol/l). There was further

reduction in CH_{50} levels and circulating immune complexes were detected by Cl_q binding assay. A decision was made to increase immunosuppressive treatment by raising the dose of prednisolone to 100 mg per day and introducing azathioprine 100 mg daily. The patient's condition deteriorated and plasma exchange was performed on three occasions. After the third treatment the circulating immune complexes became undetectable. Despite these measures his symptoms, arterial blood gases, and chest radiograph (fig 2) worsened and he died in respiratory failure on 8 December 1979. At necropsy fibrosing alveolitis affecting all the lobes of the lungs was found, with no evidence of pulmonary infection or neoplasm.

Patient 2 A 76-year-old housewife presented in April 1979 with a two-month history of progressive proximal muscle weakness and a pruritic, erythematous and scaly rash on her face and limbs. In the week before admission she had noticed dyspnoea on exertion. On examination she had an extensive exfoliative rash over most of her body and obvious symmetrical proximal muscle weakness. Auscultation of her chest indicated bilateral basal crackles. Her

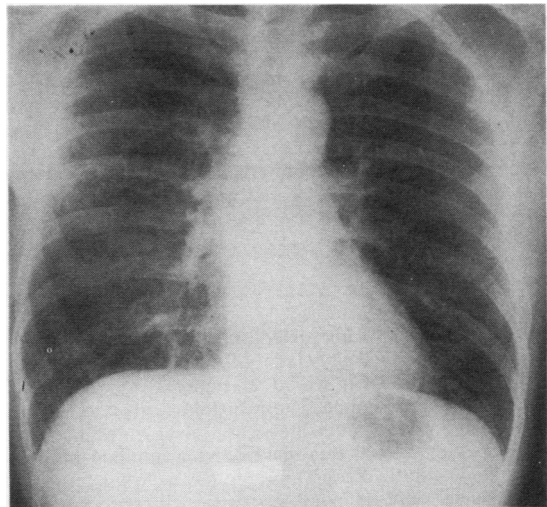


Fig 1 Patient 1: chest radiograph after onset of dyspnoea showing early interstitial changes.

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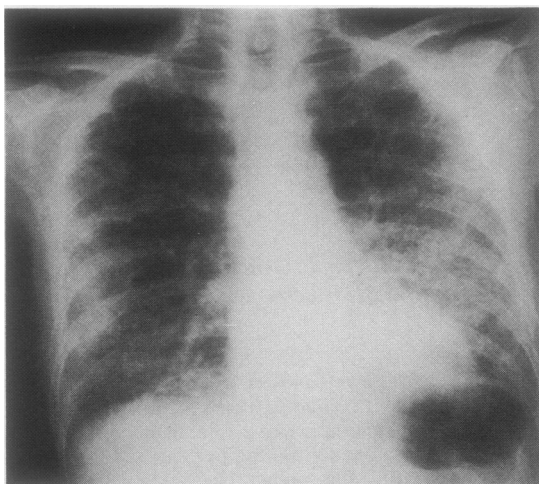


Fig 2 Patient 1: chest radiograph (68 days after fig 1) showing progression of bilateral changes.

chest radiograph showed shadowing in both lower zones. The serum lactate hydrogenase level was raised to 598 units/l (normal range 72–395) but creatine kinase was normal. A muscle biopsy showed marked myopathic features. Serum complement levels were reduced (C3 125 mg/dl (normal range 134–156); C4 26 mg/dl (normal range 30–51 mg/dl)) and circulating immune complexes were detected by a complement-fixation test. A diagnosis of dermatomyositis with fibrosing alveolitis was made and she was treated with prednisolone 80 mg daily. There was considerable improvement in her muscle power and skin rash and she was discharged taking prednisolone 20 mg daily on 22 May 1979. She was readmitted in August 1979 with a two-month history of progressive dyspnoea despite an increase in her prednisolone dosage to 30 mg per day. No new findings emerged from examination though her rash had improved. A chest radiograph showed deterioration and arterial blood gas analysis indicated hypoxia. Immunosuppressive treatment was immediately increased by raising the dose of prednisolone to 80 mg per day and adding cyclophosphamide 50 mg three times a day. Despite these measures her clinical condition, arterial blood gases, and chest radiograph deteriorated and she died in respiratory failure on 15 October 1979. A postmortem examination showed severe fibrosing alveolitis with no evidence of infection.

Discussion

Patients with fibrosing alveolitis associated with dermatomyositis are usually classified into two groups.^{4,5} The first comprises those in whom the pulmonary disease runs an acute course, and in whom histologically there is an acute inflammatory infiltrate in the alveolar wall, which resolves with steroid treatment leaving minimal fibrosis. The patients in the second group follow a chronic progressive course with a poor response to corticosteroids. Schwarz *et al* described 10 such “steroid-resistant” patients in a review of the published reports.⁵ Eight of the group had died at the time of reporting, the mean survival period

being 40 months. Only two patients, however, died of respiratory failure. The remaining two patients in the “non-responder” group were alive six and seven years after diagnosis. The pulmonary disease in our cases did not respond to steroid treatment and both died in respiratory failure within six months of the onset of symptoms.

The existence of circulating immune complexes in patients with fibrosing alveolitis is well documented. Dreisin *et al* failed to find them in patients with established pulmonary fibrosis but showed that their presence correlates well with active alveolitis and with a good response to corticosteroids.⁶ Our patients both had circulating immune complexes with low serum complement levels, and yet they did not respond to corticosteroids, cytotoxic drugs, or (in one case) plasma exchange. The use of plasma exchange in adult dermatomyositis has not been reported. This treatment was tried in the first patient because he had evidence of an acute immunological process causing arteritic lesions but was not responding to conventional immunosuppressive treatment. His failure to respond may have been due to prior progression of his pulmonary disease to irreversible fibrosis.

Fernandes and Goodwill described a patient who presented with the classical cutaneous features of dermatomyositis and who developed fibrosing alveolitis during corticosteroid treatment, dying in respiratory failure two months later.⁷ This patient was similar to the two we describe, like them developing rapidly progressive, fatal pulmonary disease despite the early introduction of adequate corticosteroid treatment. They may constitute hitherto unrecognised subgroup of patients with dermatomyositis and fibrosing alveolitis.

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References

- Bohan A, Peter JB, Bowman RL, Pearson CM. A computer-assisted analysis of 153 patients with polymyositis and dermatomyositis. *Medicine (Baltimore)* 1977;**56**:255–86.
- Mills ES, Matthews WH. Interstitial pneumonitis in dermatomyositis. *JAMA* 1956;**160**:1467–70.
- Songcharden S, Raju SF, Pennebaker JB. Interstitial lung disease in polymyositis and dermatomyositis. *J Rheumatol* 1980;**7**:353–60.
- Duncan PE, Griffin JP, Garcia A, Kaplan SB. Fibrosing alveolitis in polymyositis. *Am J Med* 1974;**57**:621–6.
- Schwarz MI, Matthey RA, Sahn SA, Marmorstein BC, Scheinhorn DJ. Interstitial lung disease in polymyositis and dermatomyositis. *Medicine (Baltimore)* 1976;**55**:89–104.
- Dreisin RB, Schwarz MI, Theofilopoulos AN, Stanford RE. Circulating immune complexes in the idiopathic interstitial pneumonias. *N Engl J Med* 1978;**298**:353–7.
- Fernandes L, Goodwill CJ. Dermatomyositis without apparent myositis complicated by fibrosing alveolitis. *J R Soc Med* 1979;**72**:777–9.