(Dubowitz & Dubowitz 1964, Sandbank *et al.* 1966, Camp *et al.* 1972) and polymyositis (Thompson & Mackay 1970, Olsen & Swenson 1972, Duncan *et al.* 1974). Frazier & Miller (1974) reported an incidence of this lung complication in 5% of patients with polymyositis – dermatomyositis. In their review of 31 cases in the literature, Schwarz *et al.* (1976) indicated that in 39% of these patients the pulmonary manifestations were predominant and preceded the characteristic features of the underlying muscle disease by as long as three years. Duncan *et al.* (1974) stressed that those patients with extensive pulmonary fibrosis histologically had a poor prognosis despite corticosteroid therapy, a finding similar to that in diffuse idiopathic fibrosing alveolitis (Scadding & Hinson 1967).

A recent report (Plowman & Stableforth 1977) described an encouraging response to treatment with cyclophosphamide in a single patient with dermatomyositis and fibrosing alveolitis, though the place of this therapy in the pulmonary lesions of DM needs further evaluation. It might be argued that our patient could have fared better on immunosuppressive treatment; however this is unlikely because his lung biopsy showed severe interstitial fibrosis. He died from this complication just ten months after he came under observation and before unequivocal muscle involvement.

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Eosinophilic fasciitis with megakaryocyte aplasia[1]

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Since Shulman (1974) initially described an acute scleroderma-like syndrome, later termed eosinophilic fasciitis by Rodnan *et al.* (1975), a number of cases have been reported which conform to the original description or have additional features widening the possible clinical picture. The patient reported here had a megakaryocyte aplasia in addition to scleroderma-like illness, the latter resolving with steroid treatment and being very similar to the cases of eosinophilic fasciitis already described in the literature. However, marrow problems have not previously been reported in this condition and it is not known whether any connection indeed exists.

Case report

Mr R W, a 55-year-old sales manager, was first seen in October 1974, with an exacerbation of his long-standing Raynaud's phenomenon. Investigations revealed an erythrocyte sedimen-

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tation rate (ESR) of 50 mm/hour (Westergren) and a white cell count of 8300/mm^3 with a 24% eosinophilia. He was found to be thrombocytopenic with platelet counts ranging from 45 000 to 62 000/mm^3. Other investigations, including liver function tests, creatine phosphokinase, rheumatoid factor and autoantibodies were normal. A muscle biopsy taken from the trapezius showed normal muscle fibres but the presence of septal oedema and perivascular lymphohistiocytic infiltration with some plasma cells and eosinophils. Electromyography was normal.

He was thought to have polymyositis and commenced on 40 mg prednisolone per day, which was stopped after three weeks because of abdominal pain. He had had a previous duodenal ulcer in 1960.

In February 1975, he complained of malaise, persisting Raynaud's phenomenon and the onset of severe diarrhoea. A barium enema was normal. In March 1975, he was troubled by severe aching around his shoulder girdle, present at rest and aggravated by exercise, coming on first after mowing the lawn. The skin of his upper arms was noted to be abnormally tight and by April the trunk was also involved, together with flexion contractures of both elbows. The skin was shiny and oedematous and tightly bound down to the underlying tissues. Investigations showed that the ESR had fallen to 25 mm/hour, his blood film showed an eosinophilia on one occasion and the muscle enzymes were normal. A skin biopsy from an affected area showed a mainly lymphohistiocytic perivascular infiltrate with dermal fibrosis and normal skin appendages. These changes were thought unlikely to be secondary to scleroderma. A barium swallow and meal were normal.

Another severe bout of watery diarrhoea occurred in May 1975, accompanied by fresh bleeding and mucus together with abdominal pain. A rectal biopsy and barium enema were normal. He was started on salazopyrine. Two weeks following this he was noted to have a petechial rash on his trunk and a platelet count revealed a thrombocytopenia of 30 000/mm^3, there being no previous knowledge at that time of the original low count in 1974. The salazopyrine was immediately stopped and he was started on 60 mg of prednisolone per day. A bone marrow showed a megakaryocyte hypoplasia with almost absent iron stores but normal erythropoiesis and granulopoiesis.

Following a further drop of his platelet count in August 1975, a repeat bone marrow showed dyserythropoietic features suggesting involvement of the red cell precursors. Oxymethalone 50 mg three times per day was started and the prednisolone was increased to 150 mg per day. On discharge at the end of August 1975, his prednisolone had been reduced to 15 mg per day and in the next five months his thrombocytopenia remained between 10 000/mm^3 and 25 000/mm^3 although his haemoglobin reverted to normal on oral iron.

In February 1976, he became icteric, having developed an acute intrahepatic cholestasis due to oxymethalone. This drug was stopped and over the following six months his general condition improved. He had complete relief of his upper girdle pain, the skin loosened and the elbow movements returned to normal. He returned to full-time employment and his prednisolone was reduced to 5 mg daily by August 1976.

No further joint or skin problems developed. His ESR remained normal. The thrombocytopenia became increasingly important and failed to respond to increased doses of steroids or vincristine. A further bone marrow showed a hyperplastic granulopoiesis with an excess of early forms, blast cells forming 5% of the nucleated cells and it was possible that a slowly evolving acute myeloid leukaemia was occurring. He needed platelet transfusions initially every three weeks, to reverse severe crippling pain in the legs and feet secondary to subperiosteal and muscle bleeds, but terminally the transfusions lasted for a few days only and finally he became resistant altogether, dying in April 1978 after a cerebral haemorrhage. An autopsy was not performed.

Discussion

The acute scleroderma-like syndrome has many of the features described by Shulman in 1974 and termed 'eosinophilic fasciitis' by Rodnan *et al.* (1975). Raynaud's phenomenon has been described in one patient only (Bennett *et al.* 1977) and this is not typical of the condition. The

acute onset following exercise, the tight puckered skin of the limbs and trunk sparing the face and joint contractures are typical features. Laboratory findings have shown a raised ESR, transient eosinophilia and hypergammaglobulinaemia. The striking feature of all previously reported cases has been the marked thickening of the deep fascia between the fat and muscle with perivascular lymphohistiocytic infiltration in the superficial muscle layer and the skin. Scattered eosinophils in the dermis and muscle have been seen by some observers (Rodnan *et al.* 1975, Caperton & Hathaway 1975). A full thickness biopsy was not carried out in this patient on account of the severe thrombocytopenia.

The patients previously described have improved remarkably on steroid treatment. The histology available in this patient together with lack of elevation of muscle enzymes and normal EMG findings would make a diagnosis of scleroderma or polymyositis appear unlikely and spontaneous remission in scleroderma is rare.

Thrombocytopenia has been described in scleroderma (Carcassonne & Gastaut 1976), but haematological abnormalities other than those described have not been a feature of this disease and it would appear that this patient had eosinophilic fasciitis with a coincidental megakaryocyte aplasia.

Addendum

Since presentation of this case, a report of a patient with diffuse fasciitis and aplastic anaemia has been published (Hoffman *et al.* 1979).

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Ehlers-Danlos syndrome with surgical repair of eventration of diaphragm and torsion of stomach[1]

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A 63-year-old female, who presented in 1970 with vomiting, was found to have type I Ehlers-Danlos syndrome which is the classic severe type inherited as an autosomal dominant trait. When she was first reported to the Royal Society of Medicine in 1974 (Linnemann 1975), there was a history of previous poor wound healing and life-long kyphoscoliosis (Figure 1) with the development of premature osteoarthrosis. She had skin hyperextensibility, calcified subcutaneous spheroids, kyphoscoliosis (with later development of C4–5 subluxation), genu valgum, eventration of the left diaphragm (Figures 1 & 2) and organo-axial torsion of the stomach (Figure 3).

Despite radiologically demonstrable persistence of the gastric torsion, she recovered from a total of nine episodes of vomiting treated conservatively over a seven-year period, but in 1977, when aged 71 years, operative intervention became imperative. In view of the tissue

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