Evans' syndrome associated with dermatomyositis

E M Hay, M Makris, J Winfield, D A Winfield

Abstract

Autoimmune haematological complications in dermatomyositis are very uncommon. This case report describes autoimmune haemolytic anaemia and thrombocytopenia in a patient with dermatomyositis and pulmonary fibrosis.

Case report

A 77 year old woman presented with increasing breathlessness, angina pectoris, and bruising of her legs. Her past history was unremarkable, except for spinal osteoporosis complicated by vertebral collapse. A widespread purpuric rash was noted over the legs on examination. She was dyspnoeic and inspiratory crackles were heard throughout the lung fields. Investigations showed: haemoglobin 90 g/l, white cell count $8.5 \times 10^9/l$, platelets $25 \times 10^9/l$, mean cell volume 88 fl, and erythrocyte sedimentation rate 100 mm/h. The peripheral blood film showed numerous microspherocytes and polychromasia, in keeping with an autoimmune haemolytic anaemia. This diagnosis was supported by a reticulocytosis of $216 \times 10^9/l$ and a strongly positive Coombs' test of IgG specificity. Red cell bound IgG measured by radioimmunoassay was 5290 mol/red cell (normal <100) and red cell eluate yielded antibodies of IgG1 and IgG2 subclass with no apparent specificity. Bone marrow examination showed active erythropoiesis with plentiful megakaryocytes, implying platelet consumption rather than failure of production. Platelet autoantibodies were demonstrated by immunofluorescence. As the coagulation screen was normal, and there was no evidence of infection, immune thrombocytopenia was the most likely diagnosis.

Immunological tests, including tests for anti-nuclear antibody, antibodies to soluble cellular antigens, anti-dsDNA, and IgM rheumatoid factor, were all negative. Diffuse interstitial shadowing was present on the chest x ray, and a restrictive ventilatory defect with reduced diffusion capacity to carbon monoxide was shown by pulmonary function tests. Diagnoses of autoimmune haemolytic anaemia, immune thrombocytopenia, and pulmonary fibrosis were therefore made. Evidence of pulmonary fibrosis was present on a chest x ray from four years previously. There was a satisfactory response to treatment with 40 mg of prednisolone daily, with normalisation of the haemoglobin and platelet count. The corticosteroid was subsequently reduced to a maintenance dose of 5 mg/day.

The patient remained well for eight months, when she presented again with a florid erythe-

matous scaling eruption over the light exposed areas of the face, arms, and chest, and profound muscle weakness. A clinical diagnosis of dermatomyositis was made. There were no other features of connective tissue disease—in particular, no arthritis or Raynaud's phenomenon. Serum levels of muscle enzymes were raised: creatine kinase 308 U/l (normal 24–170), aspartate transaminase 62 U/l (normal 7–40), alanine transaminase 32 U/l (normal 7–45). A skin biopsy specimen obtained from an affected area showed mononuclear inflammatory infiltration in the upper dermis which disrupted the vascular lumen. A deltoid muscle biopsy specimen showed variation in fibre size, with many atrophic ('moth eaten') fibres. Regenerating fibres were present in moderate numbers, with cytoplasmic basophilia and vesicular nuclei. Widespread inflammatory changes were present with infiltration of muscle fibre bundles by mononuclear cells, mainly lymphocytes. The diagnosis of dermatomyositis was thus confirmed. Prednisolone, initially 40 mg/day, was reinstituted, with azathioprine 100 mg/day as a steroid sparing agent. On this regimen the patient improved dramatically and the dose of prednisolone was rapidly reduced. Despite this there were further osteoporotic complications with vertebral wedge collapse. One year later, following a fall, she sustained a fractured neck of femur necessitating a left total hip replacement.

The patient has now been followed up for three years and remains well but frail. To date there is no evidence of visceral malignancy.

Discussion

Dermatomyositis is an uncommon connective tissue disease, which may be idiopathic, or occur in association with another connective tissue disease, or as a non-metastatic manifestation of visceral malignancy.¹ The association between pulmonary fibrosis and dermatomyositis is well recognised, often with Raynaud's phenomenon, arthritis, and autoantibodies to the cellular enzyme histidyl-tRNA synthetase as the Jo-1 syndrome.²

Autoimmune haemolytic anaemia and thrombocytopenia are commonly found in association with connective tissue diseases—characteristically, systemic lupus erythematosus and scleroderma.^{3–5} The haematological abnormalities may be the presenting feature of the disease. Isolated immune thrombocytopenia has been described in only one other case of dermatomyositis as far as we know.⁶ That

Department of
Rheumatology,
Nether Edge Hospital,
Sheffield
E M Hay
J Winfield

Department of
Haematology,
Royal Hallamshire
Hospital, Sheffield
M Makris
D A Winfield

Correspondence to:
Dr E M Hay,
Rheumatism Research Centre,
Manchester Royal Infirmary,
Oxford Road, Manchester.

Accepted for publication
6 November 1989

patient had a strongly positive antinuclear antibody test, however, which may suggest an overlap syndrome with systemic lupus erythematosus. Autoimmune haematological complications are uncommon in dermatomyositis and to our knowledge the association with Evans' syndrome has not been previously reported. In our patient the haemolytic anaemia and thrombocytopenia presented at least four years after the onset of pulmonary fibrosis, but predated the dermatomyositis by eight months.

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