Childhood eosinophilic fasciitis presenting as inflammatory polyarthritis and associated with selective IgA deficiency

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Abstract
A 13 year old school boy presented with seronegative inflammatory polyarthritis after a flu-like illness. Four months later clinical features of eosinophilic fasciitis became apparent. After histological diagnosis treatment was started with prednisone 40 mg daily, with a good response. Routine investigations showed persistent selective IgA deficiency.

Eosinophilic fasciitis is a rare condition in children, and a recent review found only 18 reported cases to date.1 It is an important diagnosis to make, however, as in contrast with scleroderma, 60% of patients respond satisfactorily to prednisone or hydroxychloroquine and the prognosis is usually benign unless associated with haematological disorders.2

Case report
A 13 year old schoolboy had flu-like illness in September 1989, and generalised lymphadenopathy was noted on examination. He made a complete recovery after two weeks.

Two weeks later he complained of generalised fatigue and gradually developing stiffness, swelling, and tenderness in his hands, wrists, elbows, shoulders, knees, and ankles. Normally a keen sportsman and swimmer, he had great difficulty in walking short distances.

The only medical history was mild asthma, which was controlled by beclomethasone and salbutamol inhalers. He was not receiving any other drugs and, in particular, was not taking any products containing tryptophan. There was no relevant family history. Results of a systematic inquiry were unremarkable and showed no symptoms of Raynaud’s phenomenon or recurrent throat or gut infections.

In November 1989 he was examined by one of us (JPP), who found swelling and synovitis in his metacarpophalangeal joints, wrists, elbows, shoulders, knees, and ankles. There were no other positive findings, and skin thickening, rashes, and muscle weakness and tenderness were specifically noted to be absent.

Laboratory data showed an erythrocyte sedimentation rate of 64 mm/h and the presence of C reactive protein. The white blood cell was 13·1 x 10⁹/l and the differential showed marked eosinophilia of 3·5 x 10⁹/l (normal range 0·04–0·40 x 10⁹/l), granulocyte count 5·2 x 10⁹/l, monocyte count 1·3 x 10⁹/l, and lymphocyte count 3·1 x 10⁹/l. Haemoglobin was 121 g/l and the platelet count was 482 x 10⁹/l. Serum protein immunoelectrophoresis showed a low IgA concentration of 0·1 g/l (normal 0·73–4·22 g/l), a raised IgG concentration of 26·5 g/l (normal 6·78–17·12 g/l), and a slightly raised IgM concentration of 2·1 g/l (normal 0·54–1·9 g/l). Serum protein electrophoresis showed raised γ globulin, α₂ globulin, and total globulin concentrations, the last being 44 g/l (normal 18–35 g/l), but normal α₁ and β globulin fractions. Urea and electrolytes, muscle enzymes, results of liver function tests, and random blood glucose were normal. Antinuclear antibody and screens for rheumatoid factor, brucellosis, leptospirosis, and infectious mononucleosis were negative. The low IgA concentration was confirmed in a second laboratory at 0·2 g/l (normal 2·0–3·0 g/l).

A provisional diagnosis of juvenile chronic arthritis was made, and naproxen 250 mg twice a day was prescribed. This provided partial relief of his polyarthritis. In January 1990 early flexion contractures had developed in his elbows and knees. The range of movements in his fingers and wrists had become markedly reduced. A degree of skin firmness was noted in his lower legs, but there was no suggestion of skin thickening or tightening in his arms. In view of the early flexion contractures chloroquine 200 mg daily was started.

In February 1990 he had developed obvious skin thickening in his hands, forearms, shins, and feet. A full thickness skin biopsy showed a normal epidermis and dermis, a markedly thickened fascia with inflammatory infiltrate including eosinophils. The perimysium was also infiltrated with inflammatory cells (fig 1).

When the diagnosis of childhood eosinophilic fasciitis was confirmed treatment was started with prednisone 40 mg daily. The response was good with resolution of his joint pains, stiffness, and generalised fatigue. The peripheral eosinophilia disappeared and his erythrocyte sedimentation rate became normal.

Discussion
A history of excessive physical exertion often predates the onset of eosinophilic fasciitis. Although such a history was absent in our patient, there is a possibility that the flu-like illness a month before the onset of symptoms...
In the complexes, leading to eosinophilic disease. Girls described childhood eosinophilic fasciitis, and disease infiltrates, and the perimysium. Recent presentation of childhood eosinophilic fasciitis, focusing on circulating immune complexes, knowledge of this rare condition, eosinophilic fasciitis, has become more widespread, and it is likely we will learn more about this condition in the future.