RAPID REPORT

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 $13{\cdot}1\times10^9$/l and the differential showed marked eosinophilia of $3{\cdot}5\times10^9$/l (normal range $0{\cdot}04$–$0{\cdot}40\times10^9$/l), granulocyte count $5{\cdot}2\times10^9$/l, monocyte count $1{\cdot}3\times10^9$/l, and lymphocyte count $3{\cdot}1\times10^9$/l. Haemoglobin was 121 g/l and the platelet count was 482×10^9/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, α_2 globulin, and total globulin concentrations, the last being 44 g/l (normal 18–35 g/l), but normal α_1 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 tightening 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

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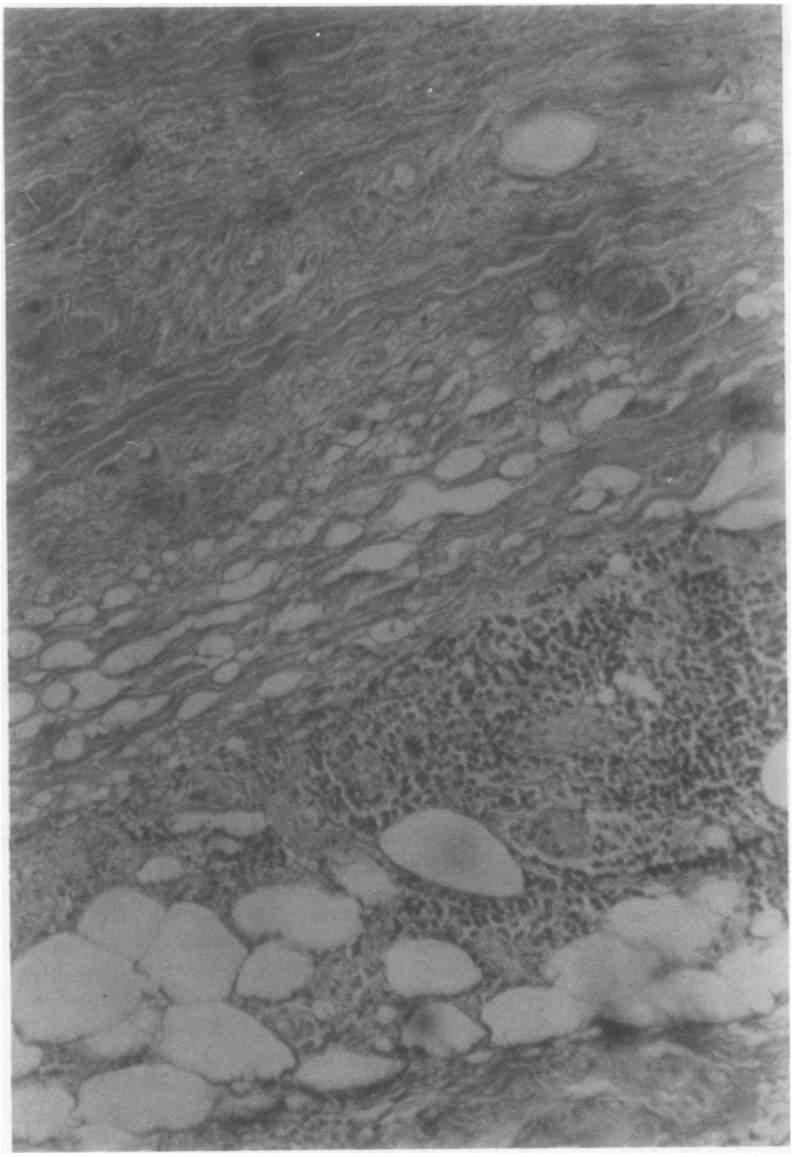

Focal inflammatory infiltrate in the perimysium.

might have precipitated circulating immune complexes,[3] leading to eosinophilic fasciitis.

The recent review on eosinophilic fasciitis in children[1] described four differences between childhood eosinophilic fasciitis and adult disease. Firstly, they found a preponderance of girls with childhood eosinophilic fasciitis, and, as far as we know, our patient is only the fifth case of male childhood eosinophilic fasciitis reported so far. Secondly, they showed increased muscle disease in childhood eosinophilic fasciitis, and focal inflammatory infiltrates were prominent in the perimysium in our patient. Thirdly, they found that arthritis occurred in 25% of children with eosinophilic fasciitis compared with 44% of adults with the disease, and the presentation as inflammatory polyarthritis has only been described once before.[4] Finally, they suggested that with its lack of haematological involvement, such as thrombocytopenia and aplastic anaemia, childhood eosinophilic fasciitis would seem to have a better prognosis than the adult disease.

In keeping with most cases of eosinophilic fasciitis, our patient had raised concentrations of γ globulin, though there has been one report of a case of eosinophilic fasciitis with hypogammaglobulinaemia.[5]

Kent and his colleagues described a 29 year old man with eosinophilic fasciitis who had mild IgA deficiency and a 23 year old woman with eosinophilic fasciitis who had complete IgA deficiency.[6] Our patient had marked IgA deficiency on repeated testing. He had no history of recurrent upper respiratory tract infections or gastrointestinal disorders, which may characterise some patients with selective IgA deficiency.

A variety of diseases, including autoimmune diseases, have been found to be associated with IgA deficiency.[7] The association with juvenile chronic arthritis is well described, especially in those with pauciarticular onset.[8–10] The significance of selective IgA deficiency in these disorders has yet to be determined.

Finally, with the recent epidemic of tryptophan induced eosinophilia-myalgia syndrome and eosinophilic fasciitis,[11–13] 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.

1 Grisanti M W, Moore T L, Osborn T G, Haber P L. Eosinophilic fasciitis in children. *Semin Arthritis Rheum* 1989; **19**: 151–7.

2 Lakhanpal S, Ginsburg W W, Michet C J, Doyle J A, Moore S B. Eosinophilic fasciitis: clinical spectrum and therapeutic response in 52 cases. *Semin Arthritis Rheum* 1988; **17**: 221–31.

3 Seibold J R, Rodnan G P, Medsger T A, Winkelstein A. Circulating immune complexes in eosinophilic fasciitis. *Arthritis Rheum* 1982; **25**: 1180–5.

4 Olson N Y, Lindsley C B, Kepes J J. Eosinophilic fasciitis presenting as inflammatory polyarthritis. *Pediatrics* 1986; **78**: 512–4.

5 Ormerod A D, Grieve T H K, Rennie J A N, Edward N. Eosinophilic fasciitis—a case of hypogammaglobulinaemia. *Clin Exp Dermatol* 1984; **9**: 416–48.

6 Kent L T, Cramer S F, Moskowitz R W. Eosinophilic fasciitis—clinical, laboratory and microscopic considerations. *Arthritis Rheum* 1981; **24**: 677–83.

7 Amman A J, Hong R. Selective IgA deficiency: presentation of 30 cases and a review of the literature. *Medicine (Baltimore)* 1971; **50**: 223–36.

8 Cassidy J T, Petty R E, Sullivan D B. Occurrence of selective IgA deficiency in children with juvenile rheumatoid arthritis. *Arthritis Rheum* 1977; **20** (suppl): 181–3.

9 Barkley D O, Hohermuth H J, Howard A, Webster A D B, Ansell B A. IgA deficiency in juvenile chronic polyarthritis. *J Rheumatol* 1979; **6**: 219–24.

10 Pelkonen P, Savilahti E, Makela A L. Persistent and transient IgA deficiency in juvenile rheumatoid arthritis. *Scand J Rheumatol* 1983; **12**: 273–9.

11 Hertzman P A, Blevins W L, Mayer J, Greenfield B, Ting M, Gleich G J. Association of the eosinophilia myalgia syndrome with the ingestion of tryptophan. *N Engl J Med* 1990; **322**: 869–73.

12 Varga J, Heiman-Patterston T D, Emery D L, *et al*. Clinical spectrum of the systemic manifestations of the eosinophilia myalgia syndrome. *Semin Arthritis Rheum* 1990; **19**: 313–28.

13 Shulman L E. The eosinophilia-myalgia syndrome associated with ingestion of L-tryptophan. *Arthritis Rheum* 1990; **33**: 913–7.