Eye (1989) **3,** 484–488

Letters to the Editor

Sir,

Eosinophilic fasciitis is an uncommon and only recently recognised connective tissue disorder characterised by painful swelling and induration of the skin and soft tissues. It most commonly affects the limbs and occasionally the trunk. It is associated with peripheral blood eosinophilia, hypergammaglobulinaemia and a raised ESR, while autoantibodies are typically absent.[1] To the best of our knowledge no ocular involvement has previously been described. We describe a patient who presented with severe conjunctival chemosis.

Case Report

A 51 year old caucasian female presented in March 1984 with a dramatic bilateral conjunctival chemosis of no obvious cause and which failed to respond to topical steroids or adrenaline. There were no other ocular abnormalities. She gave a past history of bronchiectasis, but had been otherwise quite well. Skin tests to common allergens were negative. A conjunctival biopsy was performed which showed elastotic degeneration of the collagen of the substansia propria and rather conspicuous lymphangiectasia (Fig. 1). A few lymphocytes and plasma cells were present and mast cells were noted. There was no evidence of amyloid or lymphoma. The chemosis persisted until June 1985 when she developed persistent aches and pains, followed by the development of generalised oedema. Examination revealed a right sided pleural effusion and ascites. A full blood count was normal, but a differential white cell count showed an absolute eosinophilia (1.75×10^9/l). The ESR was 45 mm/hr. Renal function, liver function tests and immunoglobulins were all normal. Antinuclear and rheumatoid factors were negative. Computed tomography scan of the abdomen confirmed ascites but failed to reveal any lymphadenopathy or organomegaly. Bronchoscopy was normal; a pleural aspirate showed no malignant cells but numerous eosinophils. Histology of lung and pleural tissues obtained at open lung biopsy revealed active chronic inflammation compatible with rheumatoid disease. A provisional diagnosis of polyserositis due to an underlying autoimmune disorder was made and she was given Prednisolone 30 mg daily, to which she made a dramatic response, with rapid resolution of her oedema and conjunctival chemosis.

However, in May 1986 with reducing doses of Prednisolone, she developed tightness and oedema of the thighs and arms. The new clinical findings were diffuse thickening, induration and swelling of the skin of the upper arms and inner thighs (Fig. 2). This was quickly followed by the development of a pancytopaenia and a bone marrow biopsy revealed severe aplasia. A skin biopsy was taken from the upper arm. This revealed no abnormality in the epidermis or dermis. However, the fibrous septa of the subcutaneous fat and the fascial planes were thickened with homogenisation of the collagen bundles. Some contained an inflammatory cell infiltrate consisting predominantly of lymphocytes and plasma cells (Figs. 3 and 4). In view of her clinical

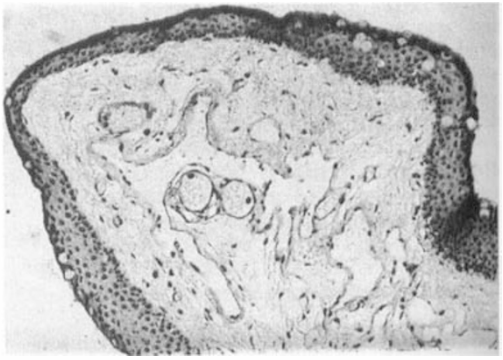

Fig. 1. *Conjunctival biopsy showing elastotic degeneration of collagen and lymphangiectasia.*

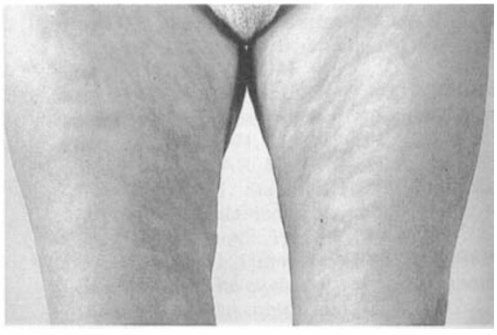

Fig. 2. *Indurated thickened skin of upper thighs.*

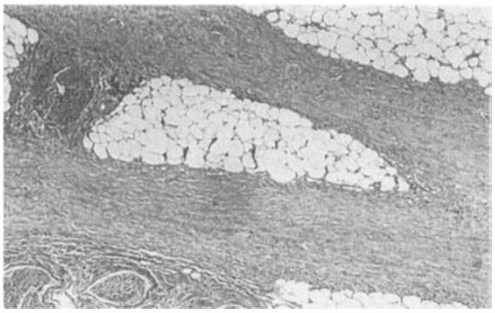

Fig. 3. *Skin biopsy showing thickening of fascial planes and interlobular septae of subcutaneous fat with homogenisation of collagen bundles. H & E×125.*

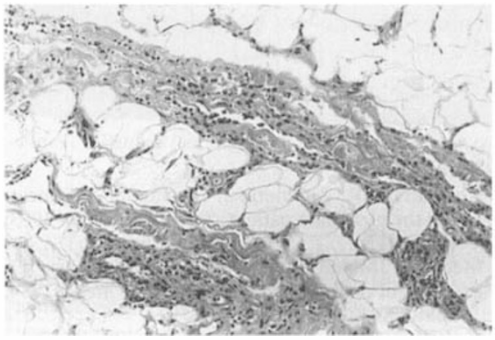

Fig. 4. *Skin biopsy showing inflammatory cells (predominantly lymphocytes and plasma cells) within the interlobular septae of subcutaneous fat. H & E×250.*

appearance, eosinophilia, raised ESR and the skin biopsy, a diagnosis of eosinophilic fasciitis was made.

The aplastic anaemia failed to respond to increased doses of steroid, antilymphocyte globulin, oxymetholone or plasmapheresis. She died in July 1986 of fungal pneumonia and post-mortem examination revealed disseminated infection with Zygomycetes.

Discussion

Many cases of eosinophilic fasciitis with multisystem presentation have been described.[2,3] However, no case has previously been described with ocular involvement. The diagnosis of eosinophilic fasciitis was somewhat delayed in this patient, as a result of the unusual presentation. Clear clinical, biochemical and pathological evidence substantiate the diagnosis. Autoimmune phenomena may account for the development of aplastic anaemia which has previously been reported in eosinophilic fasciitis.[4] The severe conjunctival chemosis with which this patient presented was the first manifestation of the generalised oedema which occurred some 14 months later. It occurred in the absence of any other ocular pathology or hypoproteinaemia and was resistant to usual treatments. It resolved rapidly and completely after administration of systemic corticosteroids.

B. Leatherbarrow and D. C. Saunders.
Manchester Royal Eye Hospital,
Oxford Road,
Manchester M13 9WH

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Sir,

Glaucoma: Ignorance and Apathy

I have interviewed 146 consecutive patients with chronic open angle glaucoma attending a general ophthalmology clinic in Norwich to assess their understanding of the disease and its treatment, and whether they would like to know more. A standardised questionnaire was used for each patient. New patients were excluded. The key question (a) "What is glaucoma" was awarded four marks for understanding of pressure, visual field loss, optic nerve compromise, and chronicity although considerable paraphrasing and flexibility of question and response was allowed. For example "Its something pressing inside my eyes which will eventually restrict my sight" would score four marks.

One patient declined to take part. The remaining 145 patients were 31 to 88 years old (mean 73). Results were analysed using the Chi-squared test.

There was a remarkably poor understand-