EOSINOPHILIC FASCIITIS – PROGRESSION TO LINEAR SCLERODERMA*

A Case Report

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Eosinophilic fasciitis is a rare disease in children. Although changes similar to linear scleroderma have been reported, the outcome is usually good. In this report, a 10-year-old boy who developed eosinophilic fasciitis without a good response to steroids is presented. He progressed to linear scleroderma within months. Our case reinforces the hypothesis that eosinophilic fasciitis may be an early manifestation or a variant of localized scleroderma similar to the other cases in the literature. *Key words: eosinophilic fasciitis, childhood scleroderma.*

Eosinophilic fasciitis (EF) is a connective tissue disease characterized by rapid development of thickening and induration of the skin in the extremities with pain, swelling and stiffness, but there is no internal organ involvement. A peripheral and cutaneous eosinophilia is present[1,2]. It is a rare disease in children. The outcome is usually good, although changes similar to linear scleroderma have been reported[3,4]. In this article, we present a 10-year-old boy who was diagnosed with eosinophilic fasciitis and who progressed to linear scleroderma within months.

Case Report

This 10-year-old boy was otherwise healthy when swelling of his left hand occurred after physical stress. Within a month, there was swelling of the left leg below the knee, and he was reluctant to use his left arm or leg due to pain.

Physical examination revealed diffuse swelling without pitting in his left forearm and distal leg. The skin was indurated and had mild erythema (Fig. 1a, b). The subcutaneous tissue felt thickened. His head, neck, and trunk were normal. These

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was full passive range of motion of his joints, but with pain in the left. The joint appeared normal and no definite muscle weakness was apparent. No facial swelling or heliotrope skin rash was present. His white blood cell count was 8,500/mm^3 with 10 percent eosinophils. The serum IgG level was 1,126 mg/dl (normal: 748-20,001 mg/dl). Antinuclear antibody titer (ANA) was positive. His erythrocyte sedimentation rate (ESR), blood hemoglobin and hematocrit, complement levels, serum creatine kinase, T3 and T4, serum chemistry profile, urine and stool analysis, chest and limb roentgenograms, barium swallow examination, and abdominal ultrasonography were normal. Skin biopsy of the indurated left forearm showed intense eosinophilic, leukocytic, and lymphocytic fasciitis with edema (Fig. 2).

A diagnosis of EF was made on the basis of the clinical and histological findings, and prednisone 2 mg/kg was prescribed, but his response to the steroid was not good. The lesion was active and involved the joints. Penicillamine 2 mg/kg/d was initiated in combination with prednisone for the first three months, followed by penicillamine alone. The swelling of the extremities decreased. The skin in these areas became tight and hard over the joints and range of movement was limited. These skin changes were identical to those of localized scleroderma (Fig. 1c). Physical therapy was initiated to preserve joint range of motion. The disease was in remission with penicillamine therapy.

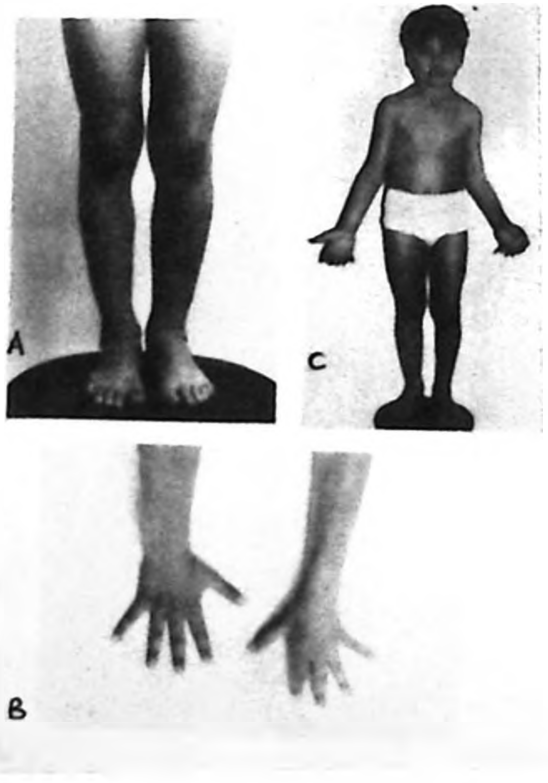

Fig. 1: Left distal leg (a) and forearm (b) of the patient depicting diffuse swelling without pitting at the beginning of the disease. Note the indurated and mild erythematous skin. After remission of the lesions, note the sclerotic areas of skin (c) in a linear, bandlike distribution crossing joint lines and leading to joint contractures on left.

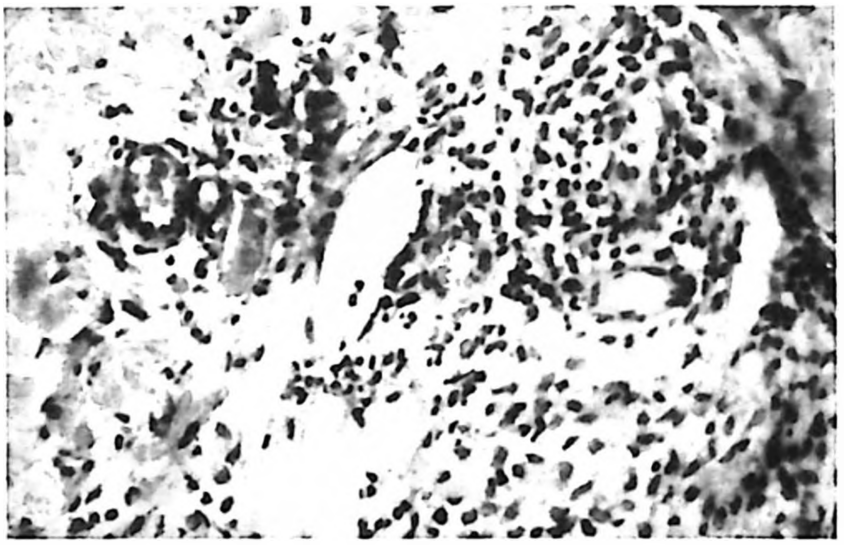

Fig. 2: Subcutaneous tissue in biopsy obtained from patient about a month after onset of symptoms. An intensive fasciitis with edema is evident. The inflammatory infiltrate was predominantly composed of eosinophils, lymphocytes and leukocytes. Patient was under no therapy at the time of biopsy (HE x 400).

Discussion

Eosinophilic fasciitis is a connective tissue disease characterized by painful induration and thickening of the skin and soft tissues, predominantly of the extremities[5]. Biopsy of involved tissue reveals diffuse inflammation of the fascia, often with eosinophilia.

Eosinophilic fasciitis (EF) in adults was originally described as a corticosteroid responsive disease which often completely resolves[3]. More recent reports of adult and pediatric cases describe initial resolution of soft tissue induration and correction of laboratory abnormalities with prednisone therapy; however, gradual progression to thickened skin occurs[6-8]. Some hemotological complications, such as aplastic anemia, thrombocytopenic purpura and others, which occur in adults, have not been reported in children[9]. These differences suggest that the pediatric form of EF may be a distinct clinical entity.

Pediatric EF predominantly affects girls and frequently involves the hands; associated arthritis is found in only 25 percent of case[1]. The outcome of adult eosinophilic fasciitis is generally favorable, and the majority of cases respond to steroid therapy. However, complications due to residual fibrosis occur in a substantial number of patients. These include flexion contractures secondary to fascial involvement, localized morphea which develops in up to one-third of cases, and carpal tunnel syndrome which is seen in about 20 percent[3,4]. However, the outcome of pediatric EF has not been well studied. Recently, Farrington et al.[2] evaluated long-term outcome in 21 pediatric patients with biopsy-proven EF. It was shown that two-thirds of pediatric EF cases eventually progress to a form of residual cutaneous fibrosis, while one-third enjoyed

complete resolution of disease. Progression to cutaneous fibrosis may occur over one or more years, indicating that a substantial period of follow-up is required to fully assess the outcome in pediatric EF[2]. In our patient, this progression occurred within months. According to results of Farrington's series[2], two possible risk factors that are associated with the likelihood of progression to cutaneous fibrosis were identified: age under seven years and more extensive initial disease at the time of diagnosis. Although our patient was over seven, he had extensive initial disease. Farrington et al.[2] detected no association between progression to cutaneous fibrosis and the sex of the patient, duration of symptoms prior to therapy, type of therapy, history of prior physical stress, or laboratory variables at diagnosis. Transition to scleroderma has been reported but is considered a rare event[1,2,10,11]. In Farrington's series[2], one-fourth of pedilatric patients with EF progressed to some form of scleroderma-like cutaneous fibrosis. Ten of 17 reported pediatric patients with EF also developed residual skin lesions consistent with cutaneous fibrosis.

Laboratory findings in EF show peripheral eosinophilia, hypergammaglobulinemia, elevated ESR and variable presence of rheumatoid factor, ANA, anti-DNA antibody and hypocomplementemia[3]. No clear correlation between clinical features and a particular ANA titer or pattern exists. Usually these serologic abnormalities are present during periods of disease activity and disappear during remission[12,13]. Our patient had peripheral eosinophilia and ANA positivity during the period of disease activity. Peripheral eosinophilia disappeared in remission, but he still had ANA positivity.

In summary, our patient had the characteristic clinical picture of EF but no good response to prednisone. The other disturbing feature was the presence and gradual worsening of the skin changes: there is now a puckered, brown, hidebound sclerodermatous appearance. These dermatological developments reinforce the notion that EF may be an early manifestation or a variant of localized scleroderma[8,14,15]. A younger age and more extensive disease should alert the clinician to the probable eventual progression of the disease to residual skin fibrosis.

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