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Case Report
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Eosinophilic Fasciitis: A Case Report and Review of Diseases Progressing with Sclerosis

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Abstract

Observation: Eosinophilic fasciitis is a rare connective tissue disorder with unknown etiology and pathogenesis, which is characterized by peripheral eosinophilia and sclerosis. Clinically, it differs from scleroderma by lack of involvement at distal areas, absence of Raynaud's phenomenon and presence of histopathological fascial involvement. It is still controversial whether it is a variant of systemic sclerosis. Systemic steroids and immunosuppressive agents are used in the treatment. Here, we presented a 79-years old man with sclerosis not involving distal parts of extremity and eosinophilia in complete blood count who was diagnosed as eosinophilic fasciitis.

Introduction

Eosinophilic fasciitis is a rarely seen inflammatory disorder with unclear etiology. In eosinophilic fasciitis, there is induration in skin and connective tissue at extremities in particular, and it manifests like scleroderma [**1**]. Here, we present a case diagnosed as eosinophilic fasciitis with limited extension, and we will discuss diseases progressing with sclerosis in particular.

Case Report

A 79 year-old man presented to our outpatient clinic with stiffening at skin of upper and lower extremities. He cited that the complaints began with itching and bruises one year ago and that there was also stiffening at skin of trunk. The patient reported no intensive exercises or use of any drug containing L-tryptophan before onset of complaints or no accompanying muscle weakness. It was found out that the patient had no dysphagia or coldness or discoloration in hands. In the history, it was found out that he had bladder mass. The patient had no abnormal finding in family history and systemic examination was normal. In the dermatological examination, it was seen that there were sclerotic changes extending from to wrist in both upper extremities and extending from knee to ankle in both lower extremities (**Figures 1 and 2**). There was limitation in extension of both upper and lower extremities. Eosinophil percent was found to be 12% in complete blood count. Magne-

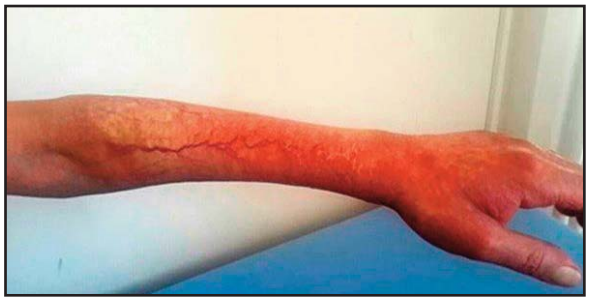

Figure 1. Diffuse sclerosis on the arm except from distal region

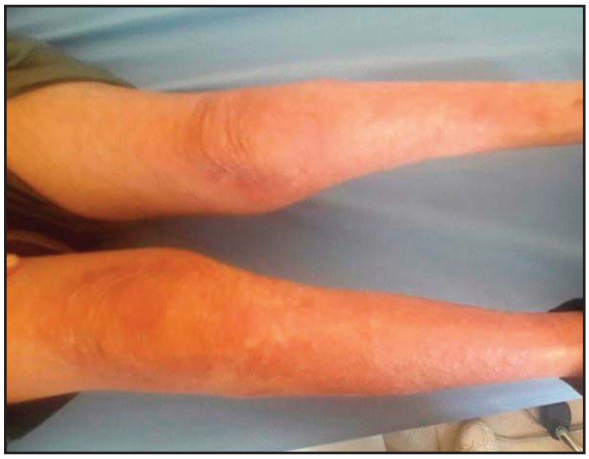

Figure 2 . Diffuse sclerosis and peau d'orange appearance in lower extremities

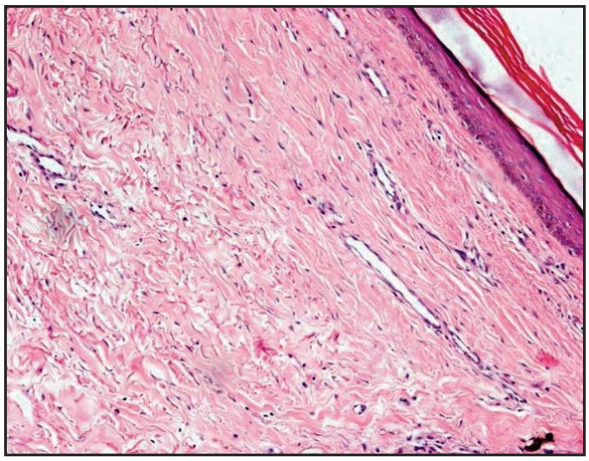

Figure 3 . Atrophic epidermis, loss of skin attachments, hyalinization in dermal connective tissue and thickening. Hematoxylin & Eosin (HE) x40

tic resonance imaging (MRI) was impossible due to prosthesis at lower extremity, while findings favoring sclerosis was observed on superficial tissue sonography. No pathological finding was detected on electromyography. The patient didn't allow biopsy involving muscle and fascia but punch biopsy performed on upper extremity revealed atrophic epidermis, loss of skin attachments, hyalinization in dermal connective tissue and thickening (**Figure 3**). By these findings, the patient was diagnosed as eosinophilic fasciitis. Deltacortril (10 mg/day) and salazopyrine (1500 mg/day) were prescribed to the patient. On the control visit after 2 months, partial recovery in sclerotic areas, especially at proximal sites, and marked improvement in extremity movements were observed.

Discussion

Eosinophilic fasciitis begins with symmetrical erythema and edema at extremities and trunk and progresses to sclerosis thereafter. Previously, it was thought that it is a form of scleroderma but it then was described as a distinct entity. Association of eosinophilic fasciitis with hematological malignancies such as polycythemia vera or multiple myeloma was shown in some cases [**2**]. The disorder is seen at fourth and fifth decades without gender predominance [**3**]. It is suggested there is a male predominance in some publications [**4**]. Our case was a 79 year-old man who had no systemic disease.

The etiology is unknown. Most cases are considered as idiopathic. It has been proposed that eosinophilic fasciitis develops after intensive exercise, Borrelia infection and hemodialysis in 30% of the patients [**5**]. Immune complexes have been identified in circulation but no individual pattern has been observed in direct immunofluorescent studies. Presence of immunoreactants, increased cytokine and elevated transforming growth factor-1 (TGF-1) were detected in fascia. It was reported that there was increased production of ınterleukin-5 (IL-5) and circulating T cell clones [**6**].

Although lesions are generally limited to extremities, they may involve trunk or any part of human body. Skin changes include induration and pitting edema initially while peau d'orange appearance, sclerosis and flexion contractures can be observed by advancing disease. Involvement of fascia leads separation of muscle groups by a demarcation line (grow sign) and veins appears to be sunk (sunken veins). It is detected in 40% of arthritis cases. Cranial and peripheral neuropathy may develop (Carpal tunnel syndrome, multiplex mononeuritis). Myalgia and fatigue are frequent complaints [**3**]. In our case, there was peau d'oranj appearance, especially in lower extremities, but no grow sign was observed.

In laboratory studies, peripheral eosinophilia is observed during acute phase. In a study by *Bobrowska-Snarska* et al., peripheral eosinophilia was observed in three of five cases [**7**]. There was elevated erythrocyte sedimentation rate in 29% and hypergammaglobulinemia in 35% of the patients. Creatine kinase is generally normal even in those with myalgia. Serum anti-nuclear antibodies are negative. Thrombocytopenia and anemia can be

observed [**3**]. Only eosinophilia was detected in our patient.

Massive thickening in fascia and profound subcutaneous fascia as well as patchy lymphohistiocytic and plasma cell infiltration are seen in histopathological examination [**6**].

In differential diagnosis, scleromyxedema is associated with diffuse induration mimicking eosinophilic fasciitis, while wax-like linear papules favor scleromyxedema. In chronic graft versus host disease (GVHD), there are morpheaform plaques that are located at trunk preferentially but also be generalized as well as history bone marrow transplantation. Sclerodermoid skin changes are commonly seen skin findings in patients with breast metastatic carcinoma after breast carcinoma (carcinoma en cuirasse) and in paraneoplastic syndromes such as primary systemic amyloidosis and carcinoid syndrome. L-tryptophan intake and history of toxic oil use are helpful in diagnosis of eosinophilia-myalgia syndrome and toxic oil syndrome. Again, nephrogenic systemic fibrosis developing after exposure to gadolinium based contrast materials is characterized by renal dysfunction accompanied by symmetrical indurated plaques in extremity and trunk [**5**].

In recent years, computerized tomography (CT) scan and MR imaging become important in the diagnosis of eosinophilic fasciitis, precluding need for biopsy in some cases. Fascial thickening, inflammation and diffuse edema are observed on MR imaging [**8**]. It couldn't be possible to obtain MR imaging due to prosthesis in our patient.

Systemic corticosteroids are effective in some patients but duration and response to treatment are variable despite presence of controlled studies [**5**]. Beside steroid, dapson, D-penicillamine, azathiopyrin, cyclophosphamide, methotrexate, cyclosporine, infliximab, rituximab, phototherapy and extracorporeal photochemotherapy are used successfully in the treatment [**9**]. However, addition of conversion to immunosuppressive agents may be needed in steroid-resistant or steroid-depended patients. In a study by *Lebeaux* et al., immunosuppressive agent use was needed in 44% of the patients [**9**].

In conclusion, eosinophilic fasciitis can be missed in clinical, causing delayed diagnosis. Thus, understanding of clinical characteristics is essential as early treatment can slow disease progression.

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