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CASE REPORT

Recurrent Eosinophilic Fasciitis: The First Case Report

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Abstract: Eosinophilic fasciitis (EF) is a rare sclerodermic form disease characterized by upper and lower limb edema. Here, we present a rare recurrent EF. A 57-year-old man presented to dermatology outpatient with skin induration on his both limbs. A diagnose of EF was made 12 years ago. According to swelling induration, eosinophilia >0.5 × 10^9/l, hyperintense fascia on MRI T2-weighted images, and fascial thickening with accumulation of lymphocytes and macrophages with eosinophilic infiltration, he was diagnosed as EF. Patient was successfully treated with oral prednisone and methotrexate (MTX). To our knowledge, this is the first report of recurrent EF during the last twenty years. We report this case to analyze the reasons for recurrent EF and review-related literature to further provide experience for the diagnosis and treatment of recurrent EF.

Keywords: eosinophilic fasciitis, sclerodermiform disease, hyperintense fascia, eosinophilia

Introduction

Eosinophilic fasciitis (EF) is a rare connective tissue disease of immune-mediated etiology and delayed diagnosis, characterized by diffuse, scleroderma-like thickening of skin on the extremities, peripheral eosinophilia, and eosinophil infiltration in fascia.[1] Typical linear depressions along the course of superficial veins ("groove sign") and "peau d'orange" appearance of the affected skin was present. Typical abnormal laboratory findings include peripheral eosinophilia, hypergammaglobulinemia, and elevated erythrocyte sedimentation rate.[2] Therapeutic options include systemic corticosteroids, methotrexate (MTX) and other immunosuppressive drugs.[3,4] Here, we report a case of evaluation of a 12-year history of skin induration involving both limbs, diagnosed as EF. We report this case to raise diagnostic awareness and treatment of EF and recurrent EF.

Case Report

A 57-year-old man with a history of diabetes presented for evaluation of a 12-year history of skin induration involving both limbs, with a 58-kg weight. The skin was indurated and noncompressible, and rheumatoid arthritis was initially suspected. Presence of one major criterion (a muscle biopsy) plus two minor criteria (peripheral eosinophilia and swelling induration, localised (extremities)), established the diagnosis of eosinophilic fasciitis (EF). A regimen of oral prednisone at 35 mg per day was prescribed. Hormone level was reduced with prednisone acetate tablets 3 tablets a day by herself, but the treatment was stopped 4 months ago. Three months ago, the condition relapsed and preceded by edema of the right forearms and bilateral legs, and stiff patches appeared on the abdomen. More than 1 month ago, no obvious cause of fatigue was observed, but he suffered from numbness and discomfort of the right upper arm and hand. No other types of discomfort, such as hand tremor and joint pain, were reported. The patient had a history of diabetes and open trauma of the right shank. He had no history of Raynaud's phenomenon. Physical examination revealed slow pace, induration and edema in his abdomen but not in the face, feet or trunk (Figure 1a–c).

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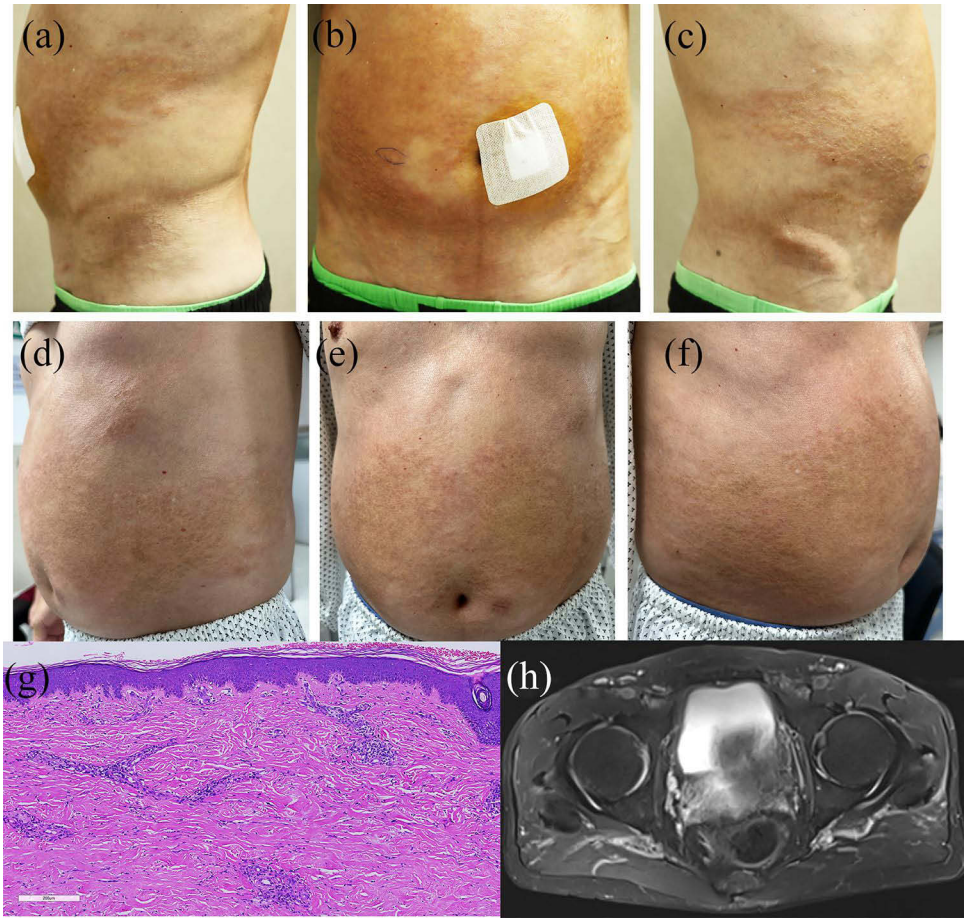

Figure 1 Manifestation of eosinophilic fasciitis: (**a-c**) prior to treatment, with the induration and edema pattern on his abdomen. (**d-f**) During treatment with systemic corticosteroids and methotrexate for 1 month. (**g**) The abdominal biopsy showed thickening of the collagen with inflammation composed of lymphocytes and a small number of eosinophils (hematoxylin eosin stain; original magnification: 100). (**h**) Magnetic resonance imaging of Hip fascia.

Laboratory tests found peripheral eosinophilia 630 cells per mm^3 (to convert to $\times 10^9$/L, multiply by.001) and an erythrocyte sedimentation rate of 55 mm/h. Liver and renal function, venereal disease, stool examination, myositis autoantibody, and electrocardiogram examination were within normal limits or negative. Magnetic resonance imaging (MRI) of the right hip demonstrated thickened fascia with enhanced signal intensity (Figure 1g). A biopsy of the abdomen revealed fasciitis with infiltration of lymphocytes, plasma cells, and eosinophils. Presence of one major criterion (swelling induration, and thickening of the skin and subcutaneous tissue that is symmetrical, localised (extremities)) plus three minor criteria (peripheral eosinophilia, muscle weakness, and hyperintense fascia on MRI T2-weighted images), the diagnosis of EF confirmed again (Figure 1h). Elastic fiber staining and CD34 in deep dermis were negative. Treatment with early combination of a regimen of prednisone at 30 mg/d (0.5 mg/kg) and 15 mg of MTX per week led to favorable outcomes (Figure 1d–f). After 2 months of follow-up, he continued to have clinical improvement. However, 2 months after the second diagnosis of EF, the patient was diagnosed with Moyamoya Disease and then lost to follow-up.

Discussion

Morphea can be classified into subtypes by the extent and depth of fibrosis: localized, generalized, linear, deep, mixed, and EF. EF is a rare scleroderma-like disorder characterized by upper and lower limb edema with poorly understood pathogenesis.[2,5] The cause of the condition is unclear, although an auto-immune mechanism is presumed. Possible reported trigger factors of the disease include intense exercise, trauma, and infections (such as SARS-CoV-2),[6] drug toxicity (antituberculous medications, phenytoin, simvastatin, atorvastatin, infliximab, pembrolizumab), immunotherapy-

association (immune checkpoint inhibitor),[7] radiation therapy, insect bites, and malignant neoplasia (the EF behaving as a paraneoplastic syndrome),[8] and an herbicide containing Florasulam (6.25 g/L) and 2,4-D 2-Ethylhexyl ester (300g/L).[9] Several possible etiologies of disease recurrence came into attention such as cancer,[10] activated inflammatory cells including eosinophils in the bronchial epithelium,[11] and c-ANCA positivity.[12] We also investigate the underlying disease factor of diabetes mellitus and open trauma to the recurrent EF in our case.

The disease mostly present in the fourth to fiftieth decades of life and slightly predominant in males.[9] Although the trunk may be involved, the hands, feet, and face are often spared. The most frequent locations of EF are the upper limbs (88%) followed by the lower limbs (70%). Raynaud phenomenon, and visceral involvement, is helpful in distinguishing EF from systemic sclerosis.[2] Approximately 10% of patients with EF have haematological disorders, and hypergammaglobulinaemia may be predictive.[13] Eosinophils occasionally present in biopsy, but tissue eosinophilia, is not essential to diagnosis, and peripheral eosinophilia is typical. And juvenile EF may manifest as swelling and progressive induration without apparent skin abnormalities. Unlike in adult populations, no underlying malignancies or associations with trauma are observed in Juvenile EF.[14]

Patients that have met the definitive diagnosis of EF in accordance with the previously proposed algorithm and qualified as two major and two minor clinical criteria were included.[15] See Table 1 Diagnostic criteria for the diagnosis of patients with eosinophilic fasciitis,[9] and Table 2 Severity classification of eosinophilic fasciitis.[15] The groove sign or vertical linear depressions were observed along superficial veins. This characteristic finding of EF is made prominent by limb elevation.[16] Our case had no prominent groove sign. Biopsy of the fascia is considered the gold standard for diagnosis, and MRI is considered an appropriate modality of EF diagnosis.[17] In light of the unexplained inflammatory syndrome, an 18F-FDG PET/CT scan is appropriate.[18]

Once the diagnosis of EF is established via major and minor clinical criteria, prompt treatment is essential to preserve mobility and function. There are no published clinical guidelines. Corticosteroids remain first-line therapies, and combination regimens, including MTX, hydroxychloroquine, cyclosporine, dapsone, MTX, PUVA, UVA1 +/− acitretin,

Table 1 Proposed Criteria for the Diagnosis of Patients with Eosinophilic Fasciitis[9]

Major criteria
1. Swelling induration, and thickening of the skin and subcutaneous tissue that is symmetrical or non-symmetrical, diffuse (extremities, trunk, and abdomen) or localised (extremities) 2. Fascial thickening with accumulation of lymphocytes and macrophages with or without eosinophilic infiltration (determined by full-thickness wedge biopsy of clinically affected skin)
Minor criteria
1. Eosinophilia > 0.5×10^9 /l 2. Hypergammaglobulinemia > 1.5 g/l 3. Muscle weakness and/or elevated aldolase levels 4. Groove sign and/or peau d'orange 5. Hyperintense fascia on MRI T2-weighted images
Exclusion criteria: diagnosis of systemic sclerosis.
Presence of both major criteria, or one major criterion plus two minor criteria, establishes the diagnosis of eosinophilic fasciitis

Table 2 Severity Classification of Eosinophilic Fasciitis[15]

Affected sites	Score
Joint contracture (upper limbs)	1 point
Joint contracture (lower limbs)	1 point
Limited movement (upper limbs)	1 point
Limited movement (lower limbs)	1 point
Expansion and worsening of skin rash (progression of symptoms)	1 point

Note: A total of 2 or more points is classified as severe.

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and TNF-α inhibitors, and other immunomodulatory agents, usually lead to favorable outcomes.[19] Features a/w refractory disease: Concomitant morphea-like skin lesions, truncal involvement, younger age of onset, and dermal fibrosis on histopathology. Biologic therapies may serve as effective adjunct treatments for patients with refractory EF, including anti-IL5 receptor monoclonal antibody benralizumab,[20] cyclophosphamide,[21] autologous hematopoiesis stem cell transplantation,[22] dupilumab,[23] and rituximab.[24]

In the first recurrent EF case during the last twenty years, recurrent features are limbs and abdomen involvement, and the specific etiological factor, such as diabetes mellitus and open trauma of the right shank, is still uncovered. Long-term studies of recurrent EF are needed. There are no clear indications to manage disease recurrence.[25] We report a patient of relapsing EF who responded positively to methylprednisolone and MTX.

EF patients be monitored for potential recurrence, such as cancer, diabetes mellitus and open trauma. Early diagnosis and aggressive treatment of EF is imperative to prognosis.

Ethics and Consent Statement

The written informed consent was obtained from the patient for the publication of the case details and images. No further institutional approval was required.

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Disclosure

The authors report no conflicts of interest in this work.

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