Case Report

A rarer association of eosinophilic fasciitis

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Abstract

Eosinophilic fasciitis (EF, also called Shulman syndrome) is a rare connective tissue disorder with poorly understood pathogenesis and unknown etiology. EF is characterized initially by limb or trunk erythema and edema and later by collagenous thickening of the subcutaneous fascia. Here, we present the case of a 16-year-old boy who presented with typical clinical features of EF with a history of typical trigger factor – preceding strenuous physical activity and had a rarer atypical association of hypercalcemia and raised angiotensin converting enzyme (ACE) levels.

Keywords: Eosinophilic fasciitis, hypercalcemia, sarcoidosis

Introduction

Eosinophilic fasciitis (EF) is a rare connective tissue disorder of unknown etiology and poorly understood pathogenesis.[1]

The etiology of EF is unknown. The following have been suggested as possible triggers or factors associated with EF:[2]

- Strenuous exercise
- Initiation of hemodialysis
- Infection with Borrelia burgdorferi
- Physical factors such as radiation therapy and burns
- Graft-versus-host disease
- Exposure to certain medications including statins, phenytoin, ramipril, and subcutaneous heparin
- Autoimmune diseases including thyroid disease, primary biliary cirrhosis, systemic lupus erythematosus, and Sjögren's syndrome
- Hematologic disorders

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Onset is typically acute, and findings include erythema, swelling, and induration of the extremities,[3,4] accompanied by peripheral blood eosinophilia.[5]

A close mimicker with thickened and hide bound quality skin is scleroderma-spectrum disorders; however, sclerodactyly, the hallmark of systemic sclerosis, and Raynaud's phenomenon are absent in EF.

Most of the patients with EF have a peripheral blood eosinophilia[6] and have an elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) as well as a polyclonal hypergammaglobulinemia.[7]

Most cases are confirmed with a full skin to muscle biopsy or by magnetic resonance imaging (MRI) if biopsy is contraindicated or nondiagnostic.

Biopsy: An elliptical full-thickness incisional biopsy of skin and subcutaneous tissues down to the muscle surface is taken. Early in the course of the disease, the deep fascia and lower subcutis are edematous and are infiltrated with lymphocytes, plasma cells, histiocytes, and eosinophils; these features are generally associated with peripheral eosinophilia. Eosinophil infiltrates are present in a majority of patients but may also be absent.[8]

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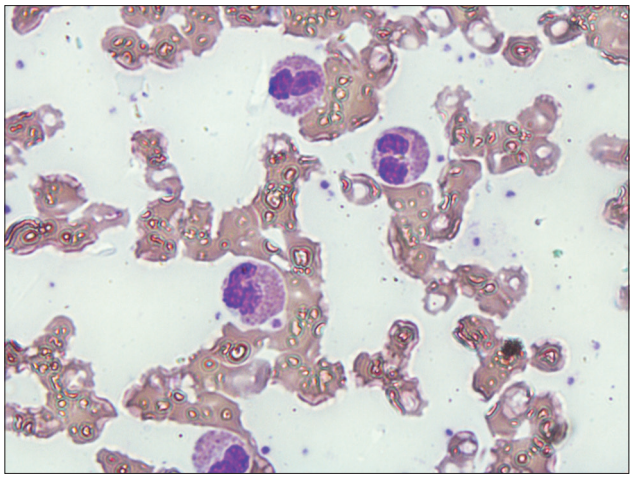

Figure 1: Eosinophils in the peripheral blood film

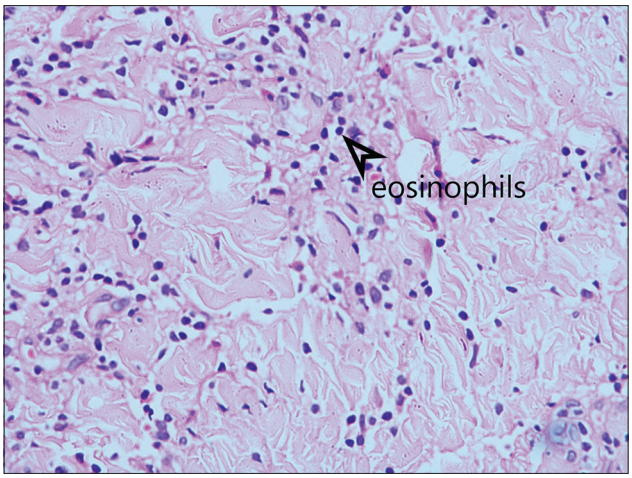

Figure 2: Eosinophils in the dermis

These structures become thickened and sclerotic as the illness progresses, with disappearance of inflammatory cell infiltrates.[9]

Magnetic resonance imaging: Findings on MRI are helpful to confirm fascial inflammation.

Management: Initial management is with systemic glucocorticoids 1 mg/kg/day.[10,11] If symptoms and signs of EF do not improve and eosinophilia persists, higher doses of glucocorticoids may be necessary. Second-line agents include methotrexate mycophenolate or hydroxychloroquine.[6]

Case Report

A 16-year-old boy who is a hypothyroid on treatment for around 6 years presented with tightening and swelling of both forearms of 3 months' duration with limitation of wrist movement and significant morning stiffness. This was preceded by a few days of intense workup in a gym to reduce weight. There was no history of fever preceding and no history of Raynaud's, photosensitivity, recurrent oral ulcers, and significant sicca. There was weight loss of around 17 kg during this period even though his appetite was normal. There was no history of difficulty in swallowing or alteration of bowel and bladder habits. On clinical examination, he had tightening of skin over the forearm with sparing of hands and face. He had pitting leg edema with no significant skin or oral lesions. His system examination was normal. His laboratory investigations have been listed in Table 1. His peripheral blood film [Figure 1] showed eosinophilia and the histopathological examination [Figure 2] showed inflammatory cell infiltrates consisting lymphocytes, variable number of eosinophils in the deep reticular dermis. The superficial fascia also had similar infiltrates and was found to be thickened and fibrosed.

Table 1: Laboratory investigations

Lab parameter	Patient value
Hemoglobin	11.6 g/dL
Total white blood cell count	11900/cumm
Differential Count	41 % eosinophils
Absolute eosinophil count	4900/cumm
Platelet count	3.7 lakhs/cumm
Erythrocyte Sedimentation Rate	24 mm/hr
C-reactive protein	39.9 mg/L
Creatine Phosphokinase	29 U/L
Lactate Dehydrogenase	72 U/L
Thyroid Stimulating Hormone	37.82 mIU/L
Serum Calcium	12.2 mg/dL
Free T4	1.1 ng/dL

The clinical diagnosis was eosinophilic fasciitis with hypercalcemia. Further detailed evaluation including a biopsy of full thickness skin with superficial muscle was done which confirmed the diagnosis of eosinophilic fasciitis. Serum protein electrophoresis showed polyclonal raise of gamma globulins with no M band. Bone marrow biopsy showed features of increased eosinophilic precursors with no abnormal cells. Hypercalcemia workup revealed 24-h urinary calcium of 672 mg/day, vitamin D total of 14.9 ng/mL with parathyroid hormone level of 7.3 pg/ml and Angiotensin Converting Enzyme level of 95.44 U/L. His chest X-ray was normal and ultrasound examination revealed hepatomegaly with mild splenomegaly.

Discussion

Our case on presentation had typical clinical features of eosinophilic fasciitis with history of preceding physical activity that is a trigger known for eosinophilic fasciitis. The atypical features were hypercalcemia and raised ACE levels. Hypercalcemia of idiopathic nature has been reported in only one case in the past.[12] Although our patient's ACE levels were elevated, there was no granuloma in the tissue obtained from the skin including superficial muscles and bone marrow biopsy. There are case reports of sarcoid-associated granulomatosis described in the past with clinical and histopathological features of sarcoid-like granulomatosis.[13,14] Our patient's hypercalcemia remitted with steroids and second-line agent methotrexate was started with good results. This case report is to highlight the

possible coexisting hypercalcemia that may have to be looked into in the setting of eosinophilic fasciitis and the elevation of ACE which could have been a nonspecific feature or an evolving overlapping CTD like sarcoidosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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