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Bilateral Eosinophilic Fasciitis in a 34-Year-Old Woman: A Case Report

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Abstract

Background: Eosinophilic fasciitis (EF), also known as Shulman syndrome, is a rare inflammatory connective tissue disorder characterized by fascial inflammation, peripheral eosinophilia, and progressive limb induration. Owing to clinical overlap with systemic sclerosis and inflammatory myopathies, diagnosis may be delayed. **Case Presentation:** We describe a 34-year-old Caucasian woman presenting with progressive skin tightening of the upper and lower extremities, burning forearm sensations, limited elbow extension, and spontaneous thigh bruising. Laboratory evaluation revealed marked eosinophilia (absolute eosinophil count 3703/μL) and elevated inflammatory markers. Autoimmune serologies were negative. Magnetic resonance imaging (MRI) of the right humerus demonstrated diffuse fascial edema and inflammation. Full-thickness biopsy of the left upper inner arm confirmed fascial thickening with lymphocytic infiltrates, consistent with eosinophilic fasciitis. **Management and Outcome:** Treatment with oral prednisone and methotrexate as a steroid-sparing agent resulted in progressive clinical improvement, normalization of inflammatory markers, restoration of skin flexibility, and recovery of joint mobility. Prednisone was successfully tapered and discontinued. The patient remains stable on a tapering methotrexate regimen. **Conclusion:** Early recognition of eosinophilic fasciitis and prompt initiation of immunosuppressive therapy are essential to prevent long-term fibrosis and functional impairment.

Keywords

Eosinophilic Fasciitis, Shulman Syndrome, Eosinophilia, Methotrexate, Corticosteroids, Connective Tissue Disease

1. Introduction

Eosinophilic fasciitis (EF) was first described by Shulman in 1974 [1]. It is char-

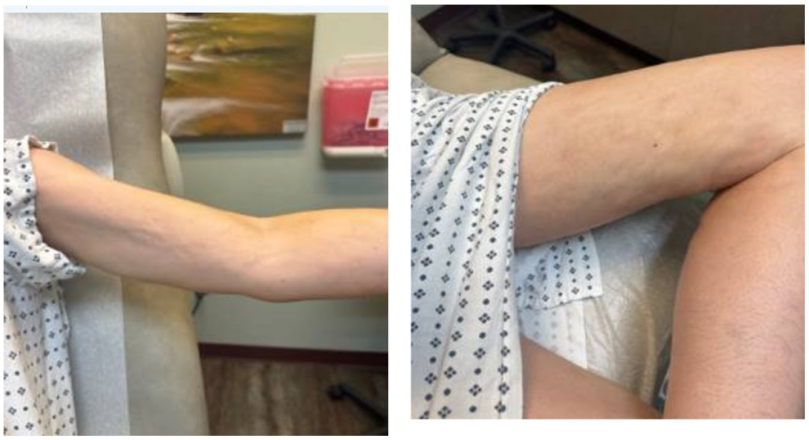
acterized by inflammation and fibrosis of the fascia, leading to progressive skin induration and restricted joint mobility. Typical features include symmetrical limb swelling and tightening, peripheral eosinophilia, and elevated inflammatory markers [2] [3].

Unlike systemic sclerosis, EF generally lacks Raynaud's phenomenon, digital ulceration, sclerodactyly, and significant internal organ involvement [4]. Early manifestations may overlap with other connective tissue diseases, contributing to diagnostic delay [4] [5]. Magnetic resonance imaging (MRI) and full-thickness biopsy extending to the fascia are frequently required to establish the diagnosis [3] [4]. Environmental triggers such as strenuous exercise, trauma, medications, and infections have been proposed, although the precise pathogenesis remains incompletely understood [3] [6].

We report a biopsy-confirmed case of EF in a 34-year-old woman with progressive limb induration and sensory symptoms, emphasizing diagnostic considerations and therapeutic response.

2. Case Presentation

A 34-year-old Caucasian woman was referred for evaluation of progressive skin tightening involving her extremities. Cutaneous changes initially appeared over the right biceps and subsequently involved both thighs. She reported difficulty fully extending her arms and raising them overhead due to increasing skin tightness.



(a) (b)

Figure 1. Symmetric cutaneous induration in eosinophilic fasciitis. (a) Upper extremity showing diffuse skin tightening and reduced pliability without sclerodactyly; (b) Lower extremity demonstrating subtle peau d'orange-like dimpling and woody induration of the thigh.

She additionally described burning sensations in her forearms, likened to "lactic acid buildup," and spontaneous bruising over her thighs associated with prominent superficial veins. She denied Raynaud's phenomenon, digital ulcers, dysphagia, fever, weight loss, or other systemic symptoms.

A detailed exposure history revealed no preceding strenuous exercise, trauma,

new medications, herbal supplements, recent vaccinations, travel, parasitic exposure, or infectious illness. She had no history of asthma, atopy, or allergic disease. She denied inflammatory joint symptoms, sicca symptoms, alopecia, or other autoimmune features.

Family history was notable for her father having similar diffuse skin changes reportedly diagnosed as deep morphea.

Physical Examination

Examination demonstrated symmetric induration and decreased skin pliability over the upper arms and thighs (**Figure 1(a)** and **Figure 1(b)**). Elbow extension was limited secondary to skin tightness. Prominent superficial veins were visible. There was no sclerodactyly, digital ulceration, or telangiectasia.

Mild cervical lymphadenopathy was present; the nodes were small, mobile, and non-tender. No progressive enlargement or systemic symptoms developed during follow-up.

3. Investigations

Laboratory Findings

- Absolute eosinophil count: 3703/μL (reference <500/μL);
- C-reactive protein: 1.57 mg/dL (reference 0.0 - 0.5 mg/dL);
- Erythrocyte sedimentation rate: 39 mm/hr (reference 1 - 30 mm/hr).

Creatine kinase and aldolase levels were normal. Autoimmune serologies including antinuclear antibody, extractable nuclear antigen panel, myositis antibodies, rheumatoid factor, anti-cyclic citrullinated peptide antibodies, and antiphospholipid antibodies were negative. Complement levels were within normal limits. Hepatitis B and C screening was negative.

Secondary causes of eosinophilia—including drug reactions, allergic disorders, parasitic infection, and hematologic malignancy—were excluded. In the absence of systemic organ involvement or persistent unexplained eosinophilia, hypereosinophilic syndrome was considered unlikely.

4. Imaging

MRI of the right humerus with and without contrast demonstrated diffuse edema throughout the musculature and along the fascial planes extending from the shoulder to the distal humerus. No fluid collections, soft tissue masses, or osseous abnormalities were identified. Prominent right axillary lymph nodes were interpreted as reactive. These findings were consistent with inflammatory fasciitis [4].

5. Histopathology

Full-thickness biopsy of the left upper inner arm revealed widening of subcutaneous septae by fibrous tissue with patchy interstitial lymphocytic infiltrates. Alt-

hough significant tissue eosinophilia was absent, the presence of fascial thickening and inflammation supported the diagnosis of eosinophilic fasciitis [3] [4].

6. Differential Diagnosis

- Systemic sclerosis: Unlikely given absence of Raynaud's phenomenon, negative serologies, and no evidence of internal organ involvement.
- Morphea: Typically confined to the dermis and subcutaneous tissue without primary fascial involvement.
- Inflammatory myopathies: Normal muscle enzymes and absence of objective weakness.
- Hypereosinophilic syndrome: Unlikely due to absence of systemic organ involvement and resolution of eosinophilia with immunosuppressive therapy.

7. Treatment

Oral prednisone 60 mg daily was initiated with gradual taper over eight weeks. Methotrexate 15 mg weekly with folic acid supplementation was added as a steroid-sparing agent. Serial monitoring included complete blood counts, liver function tests, and metabolic panels. Therapy was well tolerated.

8. Outcome and Follow-Up

Within 12 weeks, the patient experienced marked improvement in skin flexibility, resolution of burning sensations, and normalization of elbow range of motion. Inflammatory markers and eosinophil counts returned to normal. At six months, full functional recovery was achieved. Prednisone was discontinued, and methotrexate was tapered to 10 mg weekly with plans for eventual cessation.

9. Discussion

Eosinophilic fasciitis is a rare connective tissue disorder characterized by inflammation and fibrosis of the fascia, resulting in progressive limb induration and restricted mobility [1] [2]. Diagnosis relies on a combination of clinical presentation, laboratory findings, imaging, and histopathology [2]-[4]. Characteristic features include symmetrical limb involvement, peripheral eosinophilia, MRI evidence of fascial inflammation, and confirmatory full-thickness biopsy.

MRI has emerged as a valuable adjunct in the diagnostic evaluation of EF, facilitating identification of fascial inflammation, guiding biopsy location, and monitoring response to therapy [4].

Systemic corticosteroids remain the first-line treatment for EF [2] [6]. However, many patients require additional immunosuppressive therapy, most commonly methotrexate, to achieve sustained remission and permit corticosteroid tapering [2] [4] [6]. The favorable clinical response observed in this patient aligns with current therapeutic approaches.

Although EF is generally sporadic, occasional familial clustering has been re-

ported [3] [6]. The history of similar cutaneous findings in this patient's father raises the possibility of shared genetic or immune susceptibility.

Notably, this patient reported prominent dysesthetic symptoms described as a burning sensation. While skin induration and stiffness are well recognized, neuropathic symptoms may reflect inflammation involving fascial planes and adjacent peripheral nerves.

Early recognition and treatment are essential to prevent irreversible fibrosis, joint contractures, and functional impairment [2] [6].

10. Clinical Learning Points

- Consider eosinophilic fasciitis in patients presenting with symmetrical limb induration and peripheral eosinophilia, particularly when autoimmune serologies are negative [1] [2].
- MRI is a useful noninvasive tool for detecting fascial inflammation and guiding biopsy [4].
- Full-thickness biopsy extending to the fascia remains the diagnostic gold standard [3].
- Early initiation of corticosteroids and steroid-sparing immunosuppressive therapy improves outcomes and reduces long-term disability [2] [6].

11. Conclusion

Eosinophilic fasciitis should be included in the differential diagnosis of patients presenting with progressive symmetrical limb induration and eosinophilia. Timely diagnosis through MRI and confirmatory biopsy, followed by appropriate immunosuppressive therapy, can result in excellent clinical outcomes.

Patient Consent

Written informed consent was obtained for publication.

Conflicts of Interest

The authors declare no competing interests.

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