

# A rare case of unilateral eosinophilic fasciitis associated with ipsilateral extragenital lichen sclerosis

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## ABSTRACT

Eosinophilic fasciitis, also known as Shulman's syndrome, is a fibrosing scleroderma-like syndrome, which is a distinct entity. A 55-year-old man, presented with progressive skin darkening, thickening, and tightening over the left lower limb since 6 months. Dermatological examination revealed a hyperpigmented indurated area on the left thigh, extending to the anterior aspect of the left leg. A well-defined hypopigmented indurated plaque was present over the left iliac region. Histopathology and imaging studies confirmed the diagnosis of eosinophilic fasciitis and lichen sclerosis. The indurated lesion on the left lower limb responded dramatically well to oral corticosteroids. This is a rare case of unilateral eosinophilic fasciitis associated with ipsilateral extragenital lichen sclerosis.

**Key words:** Corticosteroids, eosinophilic fasciitis, lichen sclerosis, unilateral

## INTRODUCTION

Eosinophilic fasciitis (EF), synonymous with Schulman's syndrome, is a sclerosing disorder of the deep fascia with a poorly understood etiopathogenesis. Lichen sclerosis (LS), previously known as lichen sclerosis et atrophicus, is also a sclerosing inflammatory disorder of the dermis that affects the anogenital area in 85%–98% cases, with extragenital involvement in 15%–20% of patients.<sup>[1]</sup> Herein we report a rare case of unilateral EF with associated extragenital LS.

## CASE REPORT

A 55-year-old man, a bus conductor by occupation, presented with progressive skin darkening, thickening, and tightening over his left lower limb since six months. There was difficulty in extending the left knee with pruritus over the darkened skin. The patient consumed a strict vegetarian diet. Dermatological examination revealed a poorly demarcated area of brownish hyperpigmentation and induration over the anterolateral aspect of the left thigh, crossing the knee joint and extending to the anterior aspect of the left leg, sparing the foot [Figure 1a–d].

The overlying skin was puckered and Groove sign was positive. In addition, a well-defined 13 × 4 mm, hypopigmented, and indurated plaque was present over the left iliac fossa [Figure 1e]. Dermoscopy of the left iliac fossa lesion revealed an ivory white hue with follicular plugging and comedo-like openings [Figure 2a]. Laboratory evaluation revealed a differential eosinophil count of 31% with absolute eosinophil count of 2080/μL, an elevated erythrocyte sedimentation rate (ESR) of 26 mm/h, and a positive C-reactive protein. Serum IgE levels were mildly elevated (516 IU/L). Rheumatoid factor, antinuclear, and antithyroid peroxidase antibodies were negative. Peripheral blood smear showed no atypical cells. Serum calcium, lactate dehydrogenase levels, and liver and renal function tests were normal. Hematological

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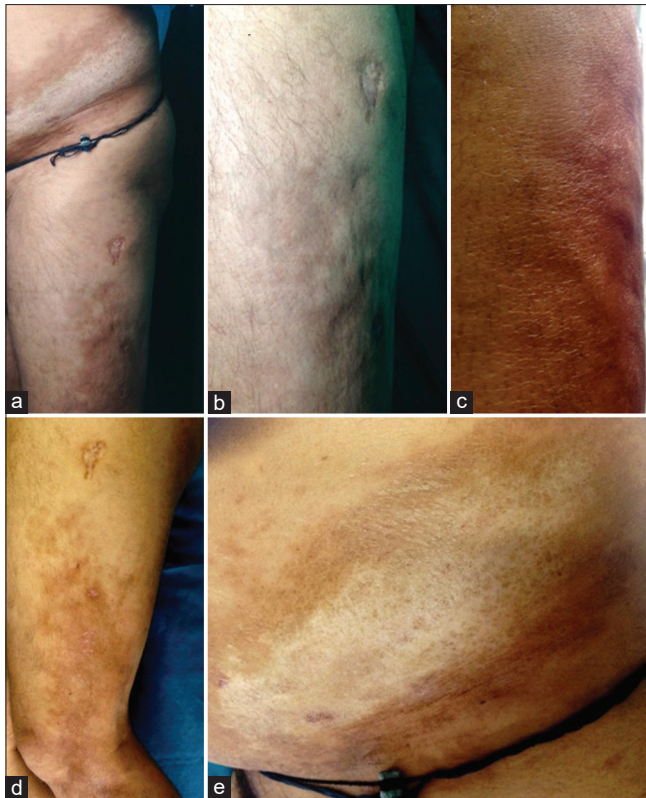
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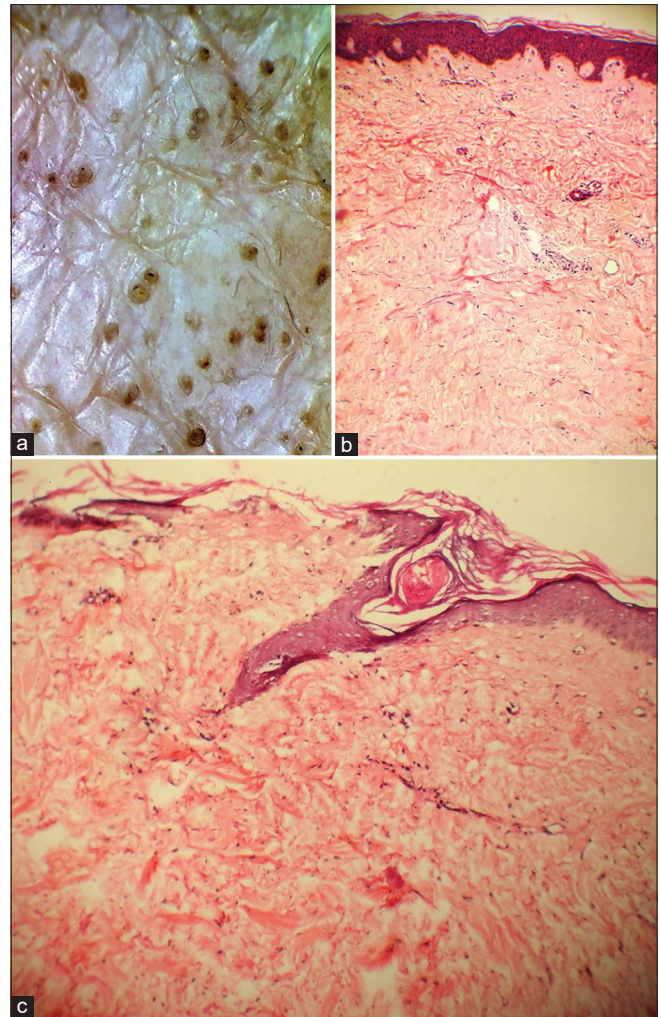
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**Figure 1:** Morphology of the clinical lesions: (a) Ivory white plaque on the left iliac region with indurated region on the left thigh, (b) close-up of the indurated area, (c) Groove sign, (d) indurated area extending to the leg, and (e) close-up of the lichen sclerosus lesion

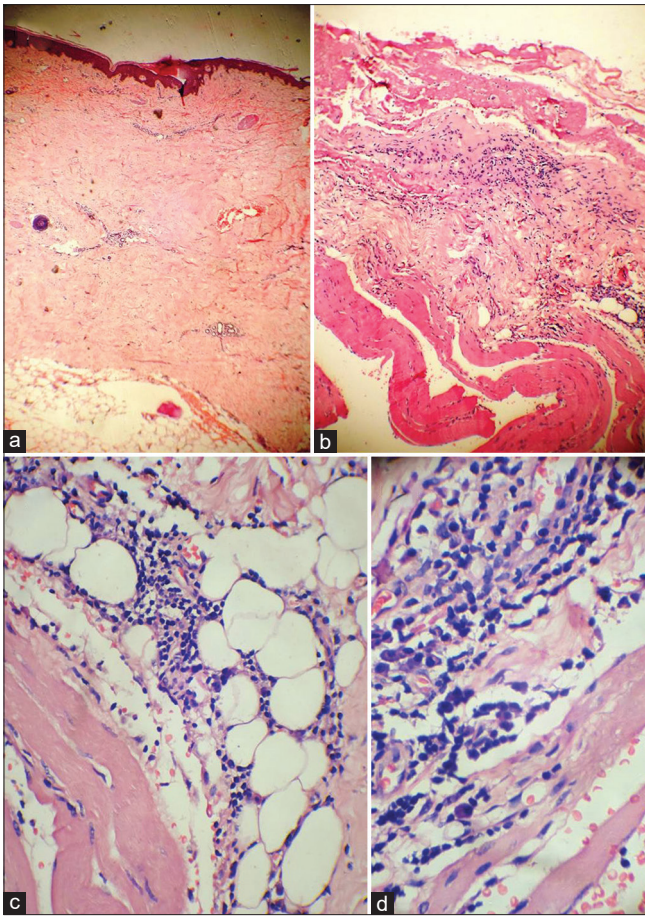
consultation was sought for the peripheral eosinophilia and possible malignant potential, which revealed no abnormality. Ultrasound study of the left lower limb revealed diffuse fascial thickening with subcutaneous edema. Deep biopsy done from the indurated lesion on the thigh revealed fascial thickening with homogenous collagen bundles present in the deep dermis and also replacing a large portion of the adipose tissue. A lymphoplasmacytic infiltrate with few eosinophils were observed in dermis, subcutaneous tissue, fascia, and muscle, consistent with the diagnosis of EF [Figure 3]. Magnetic resonance imaging showed fascial thickening without any myositis, with enhancement and increased signal intensity on fluid-sensitive sequences, corroborating EF. Biopsy from the plaque on the left iliac fossa revealed features consistent with LS [Figure 2b–c]. IgG and IgM titers to *Borrelia burgdorferi* were negative. The patient was treated with oral prednisone 60 mg daily and 1% topical tacrolimus ointment for the indurated lesion and clobetasol propionate ointment for the LS lesion. Within six weeks, there was significant improvement in edema, induration, and pliability of the EF lesion and decrease in ESR and peripheral eosinophilia. Prednisone was gradually tapered over three months. The patient is presently under periodic follow up. The LS lesion remained nonresponsive to therapy.



**Figure 2:** Lichen sclerosus lesion revealing (a) dermoscopic features of an ivory white hue and follicular plugging. (b) Histopathology revealing epidermal atrophy with the “sandwich” sign (upper dermal mild infiltrate with mid-dermal homogenization of collagen and lower dermal normal collagen) (H and E,  $\times 10$ ). (c) High power confirming the above features and follicular plugging (H and E,  $\times 40$ )

## DISCUSSION

Coined by Rodnan *et al.*, EF is a sclerosing disorder of the deep fascia. Shulman documented the first case in 1974, following which many cases have been reported worldwide. According to the classification by Petersen *et al.* in 1995, EF was a part of the scleroderma spectrum that includes systemic sclerosis, morphea, LS, atrophoderma of Pasini and Pierini, among others. However, as per the revised Consensus classification in 2004, both EF and LS have been removed from the classification. Multiple etiologic triggers such as drugs (statins, phenytoin), dietary supplements (L-tryptophan), infectious agents (*B. burgdorferi*, *Mycoplasma arginii*), vigorous physical exertion and trauma have been implicated, among others.<sup>[1,2]</sup> Our patient was a strict vegetarian who consumed dairy products and a liter of milk every day. Dairy products are a rich source of the amino acid tryptophan, that has been linked to EF.



**Figure 3:** Histopathology of the indurated area revealing features of eosinophilic fasciitis: (a) Homogenization of collagen throughout the dermis with thickened fascia (H and E,  $\times 10$ ). (b) Infiltration of lower dermis, subcutaneous fat and muscle (H and E,  $\times 10$ ). (c) Infiltration between the fat septa (H and E,  $\times 40$ ). (d) High power showing a plasmahistiocytic infiltrate with few eosinophils in the thickened fascia and muscle (H and E,  $\times 40$ )

EF is characterized by symmetrical swelling and tenderness of the distal extremities, progressing to woody induration, with or without restriction of movement. Signs like *peau d'orange* or cobblestone-like texture of the skin with puckering and the groove sign, evident by depressed grooves delineating dermal vessels and tendons, are classically described. Our patient had unilateral involvement of the left lower limb and groove sign was positive. Peripheral eosinophilia as high as 30% occurs in approximately 70% of cases,<sup>[3]</sup> as was also documented in our patient. Tissue inhibitor of metalloproteinase-1 (TIMP-1) is an emerging marker of disease activity.<sup>[4]</sup>

Krissin *et al.* reported ultrasonography as a measure of subcutaneous compressibility to distinguish EF from progressive scleroderma.<sup>[5]</sup> The subcutaneous layer of the indurated plaque was found to be edematous and compressible in our patient. Muscle-thickness biopsy remains the gold standard for diagnosis of EF. Muscle thickness biopsy in our patient revealed characteristic fascial thickening with a few

eosinophils in the dermis, subcutaneous tissue, fascia and muscle, consistent with EF.

Good therapeutic results have been observed with corticosteroids, as seen in our patient too.<sup>[6]</sup> Despite numerous case reports of EF, a unilateral presentation of EF, as in this case, has rarely been reported previously in literature.<sup>[7]</sup>

With regard to malignant potential, EF has been associated with malignancies, predominantly hematological,<sup>[8]</sup> and lymphoproliferative.<sup>[9]</sup> This patient showed a normal hemogram and peripheral smear, except for peripheral eosinophilia. Additionally, lactate dehydrogenase levels and other investigations were normal in our patient.

Extragenital LS on the other hand has very rarely been linked to malignancy, unlike its genital counterpart that may progress to squamous cell carcinoma (SCC), the risk ranging from 0.3% to 4.9%.<sup>[10]</sup> Our patient did not have genital lesions of LS. LS has also been linked with autoimmune disorders with prevalence rates peaking at 30%, the commonest being thyroiditis.<sup>[11]</sup> In our patient, all relevant markers were negative for autoantibodies. Even though no associated malignancy or autoimmune disease was found, our patient is on close monitoring and follow-up.

Our patient had a solitary plaque of extragenital LS associated with EF. Such a coexistence has been reported only once in a case series by Schaffer *et al.* on chronic GVHD, wherein, out of the six patients that developed LS, two developed EF. It further expands the clinical spectrum of sclerodermoid chronic GVHD, eliciting LS as its most superficial manifestation, and EF as the deepest.<sup>[12]</sup> *B. burgdorferi* is another common etiological factor for both the conditions. Both IgG and IgM antibodies to Borrelial antigen were negative in our patient.

Both EF and LS are recognized as part of the spectrum of scleroderma-like disorders; this fact could explain the simultaneous occurrence of both lesions in our patient.

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### Conflicts of interest

There are no conflicts of interest.

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