Eosinophilic fasciitis presenting as a unilateral, solitary plaque

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Abstract

Eosinophilic fasciitis is a rare connective tissue disorder characterized by inflammation of the fascia that leads to painful, indurated skin. Because of its variable clinical presentation and overlap with conditions, such as morphea, the diagnosis of eosinophilic fasciitis can be challenging and relies on clinical presentation, histopathologic and laboratory analysis, and response to therapy. Herein, we present an unusual, solitary, isolated plaque with pathologic features and response to therapy most consistent with eosinophilic fasciitis.

Keywords: eosinophilic fasciitis, morphea, prednisone

Introduction

Eosinophilic fasciitis is a rare disorder of unknown etiology that leads to progressive and sometimes disabling induration of the skin and connective tissue. Eosinophilic fasciitis was first described in 1974 [1] and since then, over 300 cases have been reported across the world [2]. Although the pathophysiology is still unknown, specific triggers have been reported, such as vigorous activity, medications, or infection [3]. Although eosinophilic fasciitis commonly presents symmetrically with distinct morphologic features, we describe an unusual solitary plaque with pathologic features consistent with eosinophilic fasciitis.

Case Synopsis

A 16-year-old previously healthy female athlete presented with acute worsening of an 8-month old painful plaque on her right thigh after competing in a cross-country competition. She had been treated by her primary care physician with minocycline 100mg daily and topical clobetasol 0.05% ointment twice daily for six months prior to presenting in our clinic. During her visit, she described increased pain and induration of her plaque. She denied taking any additional medications, history of medical conditions, or recent illness. She endorsed a family history of granulomatosis with polyangiitis in her grandfather.

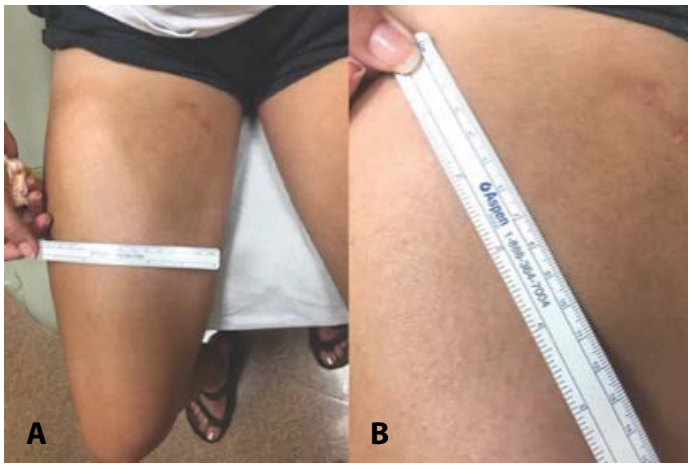

Figure 1. **A)** Solitary indurated plaque on the upper right thigh. **B)** Close-up of induration.

Upon presentation, physical examination revealed a 13×10cm firm, indurated plaque with no overlying epidermal changes on the right upper thigh (**Figure 1**). No other cutaneous, mucosal, or joint findings were noted. Given the localized unilateral appearance of the lesion on the proximal lower extremity, a working diagnosis of morphea was favored. Biopsy revealed thickening of the fibrous septae of the subcutis with an inflammatory infiltrate consisting of lymphocytes, plasma cells, histiocytes,

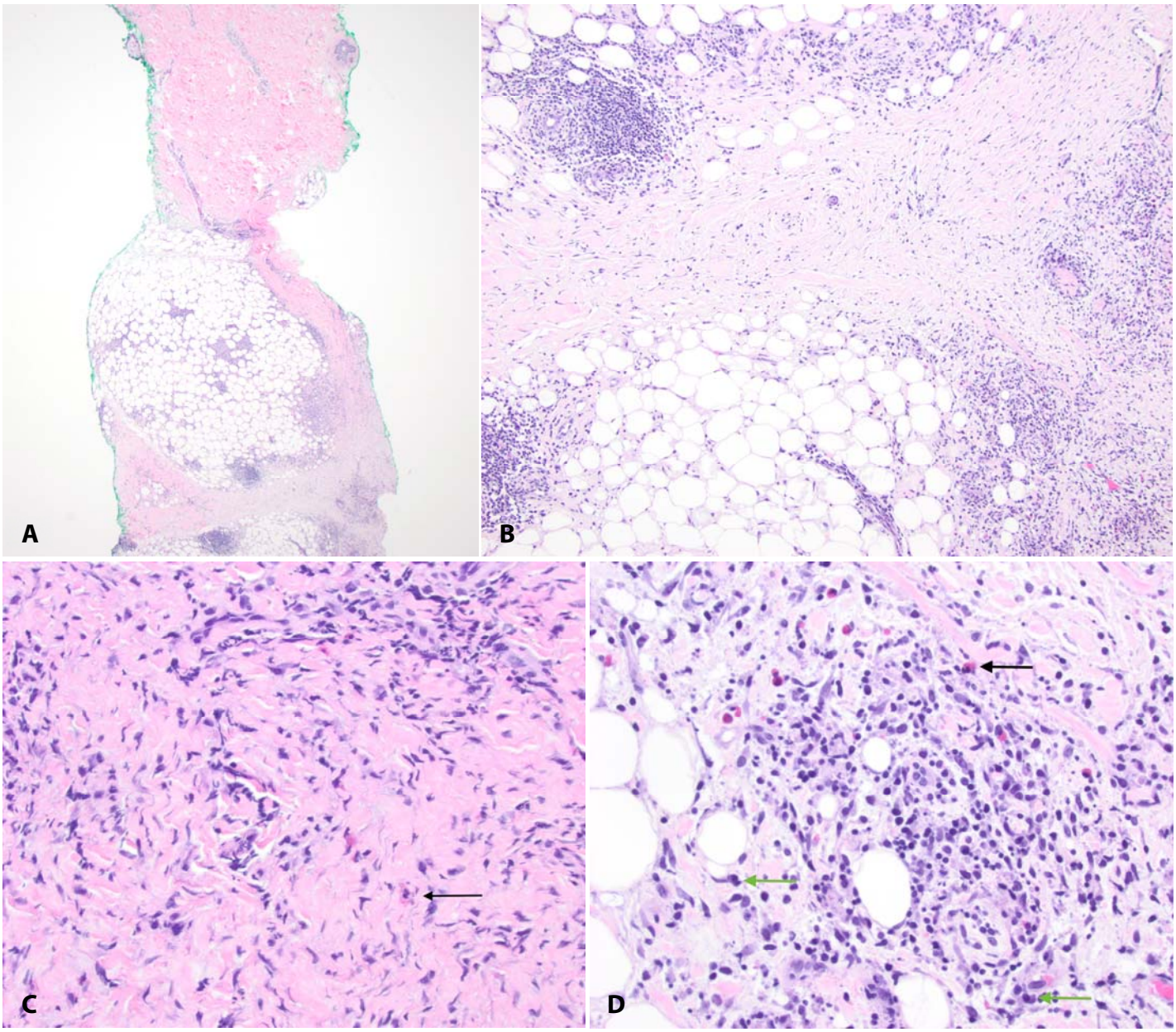

Figure 2. **A-C)** Biopsy revealed prominent septal thickening and fibrosis in the subcutis accompanied by inflammatory infiltrates, including **C, D)** lymphocytes, plasma cells (green arrows), histiocytes, and prominent eosinophils (black arrows). **A)** 20×; **B)** 100×; **C, D)** 400×.

and prominent eosinophils consistent with eosinophilic fasciitis (**Figure 2**). The epidermis and dermis were spared. Focal lymphoid follicles and fat necrosis were also evident.

Given the classic histological findings, the diagnosis of eosinophilic fasciitis was made. Minocycline was discontinued and the patient was started on a one-month oral prednisone taper starting at 40mg daily while continuing topical clobetasol 0.05% ointment twice daily. At three-week follow-up, she noted dramatic improvement in symptoms with no further pain and decreased induration. However, one month after completion of her prednisone taper, pain in the area began to return. An additional one-month prednisone taper was completed with resolution of symptoms without further recurrence. Five months after initial presentation, the patient remains pain-free and continues to run in cross-country competitions.

Case Discussion

Eosinophilic fasciitis, initially referred to as Shulman syndrome, has often been regarded as part of the

morphea spectrum [4]. The pleomorphic clinical appearance of eosinophilic fasciitis and its overlapping features with other sclerotic conditions can make clinical diagnosis challenging and can often cause delay in diagnosis. This is evident as an overwhelming 79% of eosinophilic fasciitis patients are initially misdiagnosed [5]. Further complicating clinical judgement, about one-third of patients report concurrent existence of both eosinophilic fasciitis and morphea [5-7]. Despite this overlap, there are several distinguishing features between eosinophilic fasciitis and morphea (**Table 1**).

Classic findings in early eosinophilic fasciitis appear as edema, followed by 'peau d'orange' dimpling with hyperpigmentation, and subsequent painful induration and thickening of the skin. Additionally, patients with eosinophilic fasciitis may display the 'groove sign,' denoting depressions on the skin overlying veins that are accentuated with limb elevation. Eosinophilic fasciitis usually presents symmetrically favoring the distal extremities but may additionally appear on the trunk, back, buttocks, face, or neck [6]. In contrast, morphea classically presents as a smooth yellow-white sclerotic plaque, typically unilateral and on the trunk or proximal extremities [8]. Thus, morphea was initially suspected in our patient given the exuberant clinical appearance of a smooth indurated plaque with proximal and unilateral limb localization.

Although clinical appearances can be misleading, histopathologic studies can provide diagnostic value to help distinguish eosinophilic fasciitis from morphea. The gold standard for diagnosis of eosinophilic fasciitis requires skin-to-muscle biopsy; however, in this case, subcutaneous tissue was able to provide sufficient evidence for diagnosis. Typically, histopathology of eosinophilic fasciitis reveals a thickened fascia with inflammatory infiltrate composed of lymphocytes, plasma cells, histiocytes, and eosinophils [9]. Eosinophil infiltrates have been reported in as high as 80% of patients but is not required for diagnosis [6, 10, 11]. In contrast, histologic evaluation of morphea usually reveals a perivascular infiltrate with lymphocytes and plasma cells with thickened collagen bundles, primarily focused in the papillary and reticular dermis [4].

Table 1. Clinical features of eosinophilic fasciitis and plaque morphea.

Features	Eosinophilic Fasciitis	Plaque Morphea
Onset with physical exertion	Common	Uncommon
Clinical appearance	Edema, followed by 'peau d'orange' dimpling with hyperpigmentation, progressing to a painful induration and thickening of the skin	Violaceous/erythematous lesion, progressing into a smooth yellow-white sclerotic plaque surrounded by a violaceous rim ('lilac ring')
'Groove sign' overlying veins	Present	Absent
Location	Most commonly found on extremities	Most commonly found on trunk
Localization	Symmetric, distal	Unilateral, proximal
Peripheral Eosinophilia	Common	Uncommon
Skin Histology	Usually involvement of fascia, eosinophils present; dermal and epidermal sparing	Dermal thickening and fibrosis with epidermal atrophy; rarely eosinophils
Extracutaneous manifestations	Joint contractures (50-67%), inflammatory arthritis (40%), carpal tunnel syndrome (17-23%) [3, 5, 6]	Arthralgias (19%), myalgias (14%), joint swelling (10%), limb contractures (2%), [13]
Treatment	Initial therapy usually requires systemic steroids. Additional therapies include oral methotrexate, mycophenolate mofetil, and phototherapy	Initial therapy is systemic steroid-sparing, including topical tacrolimus, topical/intralesional corticosteroids, phototherapy, and oral methotrexate

Specifically, in our patient, septal fibrosis and mixed inflammatory infiltrates, including abundant eosinophils in the subcutis with dermal and epidermal sparing, favored the diagnosis of eosinophilic fasciitis over morphea. Although not diagnostic, associated laboratory abnormalities of peripheral eosinophilia and hypergammaglobulinemia may also provide additional information to support the diagnosis of eosinophilic fasciitis [12].

The mainstay of treatment for eosinophilic fasciitis includes oral corticosteroid monotherapy. Delineating these two conditions is imperative as morphea may require more long-term therapy with steroid-sparing agents, such as methotrexate, whereas systemic steroid therapy may be required for more severe or generalized cases (**Table 1**), [4]. Early recognition and proper treatment of eosinophilic fasciitis is critical in preventing complications from disease progression leading to joint contractures, which may affect over half of patients with eosinophilic fasciitis [3, 5, 6]. Hematologic malignancies and solid malignant tumors have been associated with eosinophilic fasciitis [3, 6, 7], lending further support to the necessity of early diagnosis of this disease.

Although eosinophilic fasciitis is a rare disease, its similarity to cutaneous manifestations of other diseases has raised the question that the prevalence of this disease may be higher than previously reported owing to misdiagnosis [3]. Further exploration of these two conditions is necessary to distinguish the clinical classification and etiology of these pathologies.

Conclusion

In summary, we present a patient with eosinophilic fasciitis confirmed by histology with an unusual clinical presentation as a unilateral solitary plaque. Clinicians should be aware of the clinical variability of eosinophilic fasciitis mimicking morphea, as well as eruptions of other sclerotic etiologies. Thus, it is important for clinicians to have an accurate clinical concept of both eosinophilic fasciitis and morphea in order to provide appropriate treatment for patients. This may lead to decreased misdiagnosis and improved patient outcomes.

Potential conflicts of interest

The authors declare no conflicts of interests

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