Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Internal Medicine

Beyond Systemic Sclerosis: Eosinophilic Fasciitis in a 28-Year-Old Woman

Meriem Mouharir[1*], Zakaria Chahbi[1], Said Kaddouri[1], Hassan Qacif[1], Mohamed Zyani[1]

[1]Department of Internal Medicine, Avicenne Military Hospital, Marrakech, Morocco, Faculty of Medicine and Pharmacy of Marrakech - Cadi Ayyad University. Marrakech. Morocco

DOI: https://doi.org/10.36347/sjmcr.2025.v13i12.027 | **Received:** 13.10.2025 | **Accepted:** 21.12.2025 | **Published:** 23.12.2025

***Corresponding author:** Meriem Mouharir
Department of Internal Medicine, Avicenne Military Hospital, Marrakech, Morocco, Faculty of Medicine and Pharmacy of Marrakech - Cadi Ayyad University. Marrakech. Morocco

Abstract **Case Report**

Eosinophilic fasciitis, also known as Shulman's disease, is a rare scleroderma-like fibrosing disorder characterized by inflammation and thickening of the fascia, often associated with peripheral eosinophilia. Because of its rarity and heterogeneous clinical presentation, EF is frequently misdiagnosed, particularly as systemic sclerosis or other connective tissue diseases. We report the case of a 28-year-old woman with no significant past medical history who presented with a two-week history of painful swelling and progressive induration of the upper and lower limbs, associated with morning stiffness and functional impairment. There was no Raynaud's phenomenon, no digital ulcers and no visceral involvement. Laboratory investigations revealed marked peripheral eosinophilia at 1,100/mm³, autoimmune serology, including antinuclear and extractable nuclear antigen antibodies, was negative. Magnetic resonance imaging of the affected limbs demonstrated diffuse thickening and enhancement of the superficial and deep fasciae. A full-thickness skin-to-muscle biopsy confirmed the diagnosis by showing dense inflammatory infiltrates rich in eosinophils within the fascia, with associated collagen thickening and fibrosis. The patient was treated with three consecutive daily pulses of intravenous methylprednisolone, then switched to oral prednisone at 1 mg/kg/day with gradual tapering. Clinical response was rapid, with significant improvement in pain, regression of edema and induration, and normalization of eosinophil count and inflammatory markers. This case highlights the importance of considering eosinophilic fasciitis in patients presenting with limb edema and skin induration without Raynaud's phenomenon, especially in the presence of unexplained eosinophilia. Early recognition and prompt initiation of systemic corticosteroid therapy are crucial to prevent irreversible fibrosis and functional sequelae.

Keywords: Eosinophilic fasciitis, Shulman's disease, eosinophilia, limb induration, corticosteroids, case report.

INTRODUCTION

Eosinophilic fasciitis (EF), also referred to as Shulman's disease, is a rare fibrosing disorder first described in 1974 in patients with painful swelling and induration of the extremities, associated with peripheral eosinophilia, elevated inflammatory markers and hypergammaglobulinemia [1,2]. EF is characterized by inflammation and thickening of the fascia and subcutaneous tissues, leading to progressive skin induration and joint contractures [1,3]. Clinically, it may mimic systemic sclerosis or morphoea, but typically lacks Raynaud's phenomenon, digital ulcers, telangiectasia and visceral involvement [3].

Because of its rarity and the absence of specific serological markers, diagnosis is often delayed and relies on a combination of clinical features, imaging particularly magnetic resonance imaging and histological confirmation by deep skin-to-fascia biopsy [2,4]. Early recognition is essential, as timely initiation of systemic corticosteroid therapy can halt progression and improve functional outcomes, whereas delayed treatment may result in irreversible fibrosis and disability [3,5]. We report a case of EF in a 28-year-old woman with progressive limb swelling and induration, illustrating the diagnostic challenges and emphasizing the importance of integrating clinical, radiological and pathological data.

CASE PRESENTATION

A 28-year-old woman with no significant medical history presented to our internal medicine department with a two-week history of progressive swelling and pain in both forearms and legs, followed by skin tightening and a limited range of motion of the wrists and ankles. The symptoms had begun insidiously,

without any obvious trigger, and progressively worsened, leading to functional impairment and difficulty walking and performing daily activities. There was no history of Raynaud's phenomenon, photosensitivity, small joint arthralgia, dry eyes or dry mouth, myalgia, fever, recent infection, new medication, or weight loss.

On physical examination, the patient was afebrile and hemodynamically stable. Inspection of the limbs revealed symmetric, non-pitting edema and "woody" induration of the skin over the forearms and legs, more pronounced on the left side. The overlying skin appeared thickened and bound down to the underlying tissues, with a "peau d'orange" aspect in some areas. Palpation disclosed tenderness and firmness along the course of the fascia, with reduced joint mobility at the wrists, elbows, ankles and knees. The hands and feet were spared, without sclerodactyly, digital ulcers or nailfold capillary abnormalities. No lymphadenopathy, organomegaly or signs of visceral involvement were noted.

Laboratory investigations showed anemia with a hemoglobin level of 10 g/dL and a white blood cell count of 11,570/mm³ with eosinophilia at 1,100/mm³. The erythrocyte sedimentation rate was elevated at 60 mm/h and C-reactive protein at 94 mg/L. Serum protein electrophoresis revealed polyclonal hypergammaglobulinemia. Creatine kinase was normal at 188 U/L. Renal and liver function tests were within normal limits. Serological tests for antinuclear antibodies, extractable nuclear antigens, rheumatoid factor, anti-cyclic citrullinated peptide, antineutrophil cytoplasmic antibodies and anti-ds DNA were negative. Screening for hepatitis B and C viruses, HIV and parasitic infections was unremarkable.

Given the suspicion of a scleroderma-like disorder, contrast-enhanced MRI of the leg was performed. It demonstrated diffuse thickening and hyperintense signal of the superficial and deep fasciae on T2-weighted and short tau inversion recovery (STIR) sequences, with marked enhancement after gadolinium administration, consistent with active fasciitis. The underlying muscle and bone structures were preserved.

A full-thickness incisional biopsy including skin, subcutaneous tissue, fascia and superficial muscle was obtained from the leg. Histopathological examination revealed thickened fascia infiltrated by dense inflammatory cells, predominantly lymphocytes and eosinophils, with areas of collagen deposition and early fibrosis extending into the adjacent septa. The dermis was relatively spared, with no significant vascular changes suggestive of systemic sclerosis. These findings confirmed the diagnosis of eosinophilic fasciitis.

The patient was initially treated with three consecutive daily pulses of intravenous methylprednisolone, then switched to oral prednisone at 1 mg/kg/day, along with calcium and vitamin D supplementation and gastric protection. No additional immunosuppressive agent was introduced. Over the following weeks, she experienced a marked improvement in pain and limb mobility, with progressive softening of the skin and reduction in edema. Peripheral eosinophil counts and inflammatory markers normalized within three months. At six-month follow-up, she remained in clinical remission on low-dose prednisone, without evidence of visceral involvement or evolution towards systemic disease.

DISCUSSION

EF or Shulman's disease, is an uncommon sclerodermiform disorder characterized by inflammation and thickening of the fascia [3,6]. It typically presents with subacute onset of painful limb swelling followed by skin induration and joint stiffness. Because of its rarity and the absence of specific serological markers, EF is often misdiagnosed as systemic sclerosis, inflammatory myopathy, or localized scleroderma, leading to diagnostic delay and risk of permanent functional impairment [2,3,7,8].

The pathogenesis of EF is not fully elucidated, but immune dysregulation with eosinophil activation and release of profibrotic mediators is thought to play a major role. Triggers such as intense physical exertion, trauma, infections, medications and autoimmune or hematologic disorders have been reported in association with EF [1–3].

Several clinical features help distinguish EF from systemic sclerosis. EF usually involves the limbs with non-pitting edema and "woody" induration of the skin, often sparing the hands, feet and face. The "peau d'orange" appearance and "groove sign" along superficial veins have been described as characteristic but not pathognomonic [2,6,9,10]. Raynaud's phenomenon, digital ulcers, telangiectasias and visceral involvement are generally absent. Biologically, peripheral eosinophilia, elevated inflammatory markers and polyclonal hypergammaglobulinemia are frequent but not constant findings and may fluctuate during the course of the disease. In our patient, the association of limb induration, eosinophilia and hypergammaglobulinemia was typical of EF [5,10].

Imaging and histology play a central role in confirming the diagnosis. MRI is particularly useful as a noninvasive tool, typically showing diffuse thickening and enhancement of the superficial and deep fasciae with relative preservation of muscle and bone [4,5,7]. In this patient, contrast-enhanced MRI of the leg demonstrated a characteristic pattern of active fasciitis, which helped to narrow the differential diagnosis and guide biopsy. Deep skin-to-fascia biopsy remains the gold standard, showing inflammatory infiltrates rich in lymphocytes

and eosinophils within the fascia, with varying degrees of collagen deposition and fibrosis [2,3]. The histological findings in our case were typical and confirmed the diagnosis of eosinophilic fasciitis.

From a therapeutic standpoint, systemic corticosteroids remain the cornerstone of treatment for eosinophilic fasciitis [2,9]. Most authors recommend initiating high-dose prednisone at 0.5–1 mg/kg/day, with slow tapering guided by clinical response and normalization of inflammatory markers and eosinophil counts. In more severe presentations, or in patients with marked functional impairment, short courses of intravenous methylprednisolone pulses may be used at the beginning of therapy, as in our patient, to obtain a rapid anti-inflammatory effect. Early introduction of corticosteroids during the inflammatory phase is crucial to prevent progression to irreversible fascial fibrosis and joint contractures [2].

In cases of incomplete response, frequent relapses, or corticosteroid dependence, various immunosuppressive and immunomodulatory agents have been used as steroid-sparing therapies, including methotrexate, azathioprine, mycophenolate mofetil and cyclosporine [2,3,9]. These drugs may be introduced when long-term high-dose corticosteroids are required or when adverse effects become limiting. In more refractory cases, other options such as hydroxychloroquine, intravenous immunoglobulins or biologic agents (for example rituximab) have been reported in small series [2,3,8,10].

This case emphasizes the importance of considering eosinophilic fasciitis in young patients presenting with limb edema, skin tightening and functional limitation, particularly in the presence of unexplained eosinophilia and inflammatory syndrome. Awareness of this rare entity, combined with appropriate use of MRI and deep fascial biopsy, is essential to avoid misdiagnosis and to initiate timely corticosteroid therapy, thereby reducing the risk of irreversible fibrosis and long-term disability.

Conclusion

Eosinophilic fasciitis (Shulman's disease) is a rare but important cause of limb edema and skin induration that can mimic systemic sclerosis and other connective tissue diseases. The combination of limb induration, peripheral eosinophilia and elevated inflammatory markers should raise suspicion for this entity. Clinicians should consider EF in the differential diagnosis of scleroderma-like presentations, particularly in the absence of Raynaud's phenomenon and visceral involvement.

Conflict of Interest: The authors declare no conflicts of interest.

Funding: No external funding was received.

Consent: Written informed consent was obtained from the patient for the publication of this case.

References

1. Feldman SR, Silver RM, Maize JC: A histopathologic comparison of Shulman's syndrome (diffuse fasciitis with eosinophilia) and the fasciitis associated with the eosinophilia-myalgia syndrome. Journal of the American Academy of Dermatology. 1992, 26 :95–100. 10.1016/0190-9622(92)70014-7
2. Sène D : Fasciite à éosinophiles (maladie de Shulman) : mise au point diagnostique et thérapeutique. La Revue de Médecine Interne. 2015, 36 :738–45. 10.1016/j.revmed.2015.08.002
3. Knapp S, Bolko L, Servettaz A, Didier K : Fasciite à éosinophiles : actualités physiopathologiques et nouvelles voies thérapeutiques. La Revue de Médecine Interne. 2024, 45 :488–97. 10.1016/j.revmed.2024.03.006
4. Marie I, Sauvetre G : La tomographie par émission de positons au fluorodéoxyglucose dans la fasciite à éosinophiles. Revue du Rhumatisme. 2015, 82 :56. 10.1016/j.rhum.2014.07.011
5. Ihssan HH, Sara E, Kaoutar M, *et al.*,, : Unraveling Shulman's syndrome : A rare case of eosinophilic fasciitis in a pediatric patient with fascial abnormalities on MRI. Radiology Case Reports. 2025, 20:1760–5. 10.1016/j.radcr.2024.10.123
6. De Boysson H, Chèze S, Chapon F, Le Mauff B, Auzary C, Geffray L : Fasciite à éosinophiles avec hémoglobinurie paroxystique nocturne. Revue du Rhumatisme. 2013, 80 :82–4. 10.1016/j.rhum.2012.09.005
7. Ernest V, Sautereau N, Masson E, *et al.*,, : Hyperéosinophilie isolée précédant le diagnostic d'une fasciite à éosinophiles (maladie de Shulman). La Revue de Médecine Interne. 2017, 38:840–3. 10.1016/j.revmed.2017.07.010
8. Naoui A, Bouslama K, Abdallah M, *et al.*,,: Fasciite avec éosinophilie (syndrome de Shulman) : à propos de 11 patients. La Revue de Médecine Interne. 2010, 31:535–9. 10.1016/j.revmed.2010.03.344
9. Anadure RK, Shankar S, Mohimen A, Pemmaraju A, Kalita J: An unusual case of eosinophilia, myalgia and skin contractures: Shulman's disease revisited. Medical Journal Armed Forces India. 2022, 78:S273–6. 10.1016/j.mjafi.2019.12.006
10. Veyssier-Belot C, Zuech P, Lumbroso-Le Rouic L, Récanati G, Dendale R: Fasciite à éosinophiles révélant l'extension métastatique d'un mélanome de la choroïde : un syndrome paranéoplasique ? La Revue de Médecine Interne. 2008, 29:1013–6. 10.1016/j.revmed.2008.05.019