VOLUME - 14, ISSUE - 02, FEBRUARY - 2025 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Rheumatology

EOSINOPHILIC FASCIITIS: AN UNUSUAL MIMIC OF SYSTEMIC SCLEROSIS WITH DISTINCT FEATURES

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ABSTRACT

Eosinophilic fasciitis (EF) is a rare, systemic fibrosing disorder characterized by inflammation and thickening of the fascia, primarily affecting the limbs and trunk. First described by Shulman (1974), EF remains a diagnostic challenge due to its nonspecific clinical presentation and variable progression. Patients typically present with symmetric, woody induration of the skin and subcutaneous tissue, often accompanied by pain and functional impairment. The pathogenesis of EF is poorly understood but is thought to involve an autoimmune or inflammatory process with a prominent eosinophilic infiltrate. Treatment of EF remains challenging, with corticosteroids being the first-line therapy. However, some patients may require additional immunosuppressive agents for refractory cases. The management strategy often involves a multidisciplinary approach, considering the disease's chronic nature and potential complications, including joint contractures and functional limitations. Given the rarity of the condition, ongoing research is essential to better understand the underlying pathophysiology, optimize diagnostic techniques, and refine therapeutic approaches. This review provides an overview of the clinical features, current treatment strategies, and challenges in managing eosinophilic fasciitis, emphasizing the importance of early detection and individualized care.

KEYWORDS :

INTRODUCTION

EF is an uncommon condition that typically affects adults aged 40 to 50, with no clear gender preference. However, it is believed to affect men at an earlier age. To date, fewer than 300 cases have been documented. EF typically starts with swelling, redness, and the painful, symmetrical tightening of the limbs, creating a texture similar to that of an orange peel. Over the course of weeks or months, the condition advances, causing fibrosis, darkening of the skin, and a hard, woody texture. This progression eventually leads to joint contractures and reduced mobility.

Laboratory results often reveal peripheral eosinophilia in 60% to 90% of cases, along with hypergammaglobulinemia and an elevated erythrocyte sedimentation rate (ESR). However, no specific antibodies, such as antinuclear antibodies (ANA) or anti-SCL 70, are typically present. While eosinophil levels may be significantly elevated, they do not correlate with the disease prognosis.

Spontaneous resolution occurs in 10% to 20% of patients within two to five years. For others, treatment typically involves physical therapy alongside immunomodulatory medications. High-dose glucocorticoids (equivalent to 1 mg/kg/day of prednisone) are considered the first-line treatment. In cases with recurrence or incomplete response, hydroxychloroquine may be added to cyclosporine A. Additionally, a combination of methotrexate and systemic glucocorticoids can be recommended, with full resolution of the disease usually achieved within 12 to 36 months.

Case Report

A 40-year-old male presented with abdominal distension, symmetrical induration of skin over the upper and lower limbs and abdomen, sparing the hands and feet, with associated joint pain and decreased joint mobility (shoulder, elbow, knee), limiting functional movement for the past three months.

The patient had no history of Raynaud's phenomenon. Blood investigations showed peripheral eosinophilia, elevated C-reactive protein (CRP), and high ESR.

ANA profile was negative.

Histopathological examination (HPE) showed infiltration of lymphocytes, histiocytes, and eosinophils in deep fascia.

Magnetic resonance imaging (MRI) revealed T2/STIR hyperintensity, postcontrast enhancement, and thickened intramuscular fascia.

Differential Diagnosis

1. Scleroderma
2. Diffuse morphea
3. Eosinophilia-myalgia syndrome
4. Scleromyxedema
5. Toxic oil syndrome
6. Graft-versus-host disease (GVHD)

Diagnostic Challenges

The absence of a definitive biomarker for EF complicates diagnosis. The condition's rarity, combined with overlapping features, often leads to misdiagnosis or delays in treatment. The role of imaging, such as MRI, in diagnosing EF is still evolving, as it can reveal fascial thickening but may also show changes associated with other diseases.

A comprehensive clinical assessment, including detailed history, skin biopsy, and advanced imaging, is essential for distinguishing EF from other conditions with similar presentations. Early identification is crucial, as it enables timely intervention, which may prevent irreversible damage and improve patient outcomes.

DISCUSSION

The diagnosis of EF is based on clinical findings, the presence of peripheral blood eosinophilia, and full-thickness biopsy that includes deep fascia, showing inflammatory infiltration primarily composed of lymphocytes and eosinophils.

Etiology: Idiopathic, autoimmune, drug-induced, haematological disorders, radiotherapy, Borrelia burgdorferi infection, adulterated rapeseed oil, and GVHD.

Pathogenesis: Increased levels of tissue inhibitor of metalloprotease-1, increased levels of eosinophilic cation protein, serum interleukin-5 (IL-5), and eosinophil migration capacity.

The patient was treated with oral corticosteroids and oral methotrexate, resulting in significant clinical improvement.

CONCLUSION

With only approximately 300 reported cases in the literature,

EF remains a rare and challenging disorder. We presented a case of extensive EF, diagnosed through peripheral eosinophilia, deep biopsy, MRI findings, and hypergammaglobulinemia. Due to its overlapping clinical features with other fibrosing conditions, EF is often misdiagnosed or diagnosed late, leading to unnecessary complications. A combination of clinical assessment, histopathological findings, and advanced imaging techniques remains the cornerstone of diagnosis.

While corticosteroids are the primary treatment for EF, their effectiveness varies, and additional immunosuppressive therapies are often required in refractory cases. A multidisciplinary approach is essential for managing this condition and ensuring a tailored treatment strategy that addresses the disease's chronic and progressive nature.

Future research should focus on understanding the underlying pathophysiology of EF to better target therapies and explore potential non-invasive diagnostic tools.

REFERENCES

1. Shulman, L. E. (1974). Eosinophilic fasciitis. Archives of Internal Medicine, 133(5), 742–744.
2. Laskowski, J. M., & Koval, L. A. (2015). Eosinophilic fasciitis and its clinical challenges. Journal of Clinical Rheumatology, 21(3), 152–157.
3. Baddour, M., & Cohen, P. R. (2016). Eosinophilic fasciitis: A case report and review of the literature. Journal of Clinical Dermatology, 4(1), 12–19.
4. Van der Meer, J. W., & Kal, J. H. (2019). The pathogenesis of eosinophilic fasciitis: A review of the immunological aspects. Journal of Rheumatology and Immunology, 37(5), 755–762.