ARP Rheumatology 2024;3:332-333
https://doi.org/10.63032/FBUI7356

IMAGES IN RHEUMATOLOGY

Eosinophilic fasciitis, a rare cause of skin thickening: a case report

Soares C[1], Roriz D[2], Pontes-Ferreira M[1], Cunha A[1], Almeida S[1], Santos-Faria D[1]

Eosinophilic fasciitis (EF) is a rare disease characterized by diffuse fasciitis with eosinophilia. The etiology is unknown, although many features suggest the involvement of autoimmune mechanisms. Some triggers of the disease onset have been documented, such as intense physical exercise, hematological, infectious, or autoimmune diseases[1].

Symptoms typically include the onset, sudden or gradual, of a full-circumference swelling and symmetrical plate-like sclerotic lesions on the four limbs, mainly forearms and lower legs. General symptoms, arthralgia and myalgia may be present. Laboratory findings include peripheral eosinophilia, elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) and, in some cases, polyclonal hypergammaglobulinemia. Immunology is usually negative. Magnetic Resonance Imaging (MRI) is a valuable imaging tool for diagnosis, revealing symmetric fascial thickening, signal hyperintensity in fluid-sensitive sequences and enhancement after gadolinium injection in the affected areas. Biopsy should include fascia to identify the pathological findings[1,2,3].

[1] Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal
[2] Radiology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal

Submitted: 11/11/2024
Accepted: 12/12/2024

Correspondence to: Catarina Soares
E-mail: catarina.dantas.soares@gmail.com

The initial treatment for EF involves oral prednisolone, with a dosage ranging from 0.5 to 1 mg/kg per day, gradually tapered based on clinical response. In patients resistant to glucocorticoids or to spare its use, alternative immunosuppressants such as methotrexate have been suggested as treatment options. The duration of treatment for non-severe cases may extend for two years, but relapses may occur afterward[2,3].

A 64-year-old woman, diabetic, presented with complaints of edema and sensation of skin thickening in the lower limbs, with progressive involvement of the trunk and forearms with a 6-month evolution. The unique constitutional symptom was asthenia. She denied other skin changes or organ symptoms. There was no Raynaud's phenomenon, history of trauma, or initiation of new medication. On physical examination, thickened skin with the Groove sign (Figure 1) was observed on the forearms, as well as a *peau d'orange* aspect on the trunk and lower limbs (Figure 2). There was no involvement of the hands or face, and no signs of digital ulcers, telangiectasias, or other skin changes. Muscle strength and the remaining physical examination were unremarkable.

Laboratory examination showed normochromic normocytic anemia, eosinophilia, and elevated acute phase markers (ESR 36; CRP 5.27 mg/dL). Immunology revealed ANA 1/160 with negative scleroderma specific antibodies. Muscle enzymes, lactate dehydrogenase and thyroid function were normal. Figure 3 shows the results of the full-body MRI, revealing diffuse fascial hyperintensity.

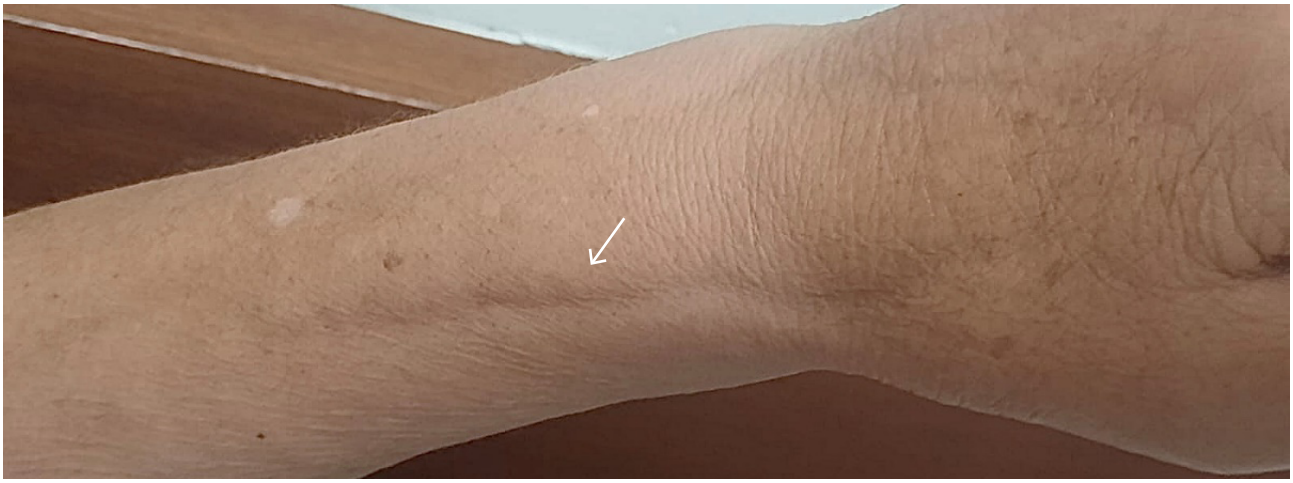

Figure 1. Goove sign observed in the left forearm.

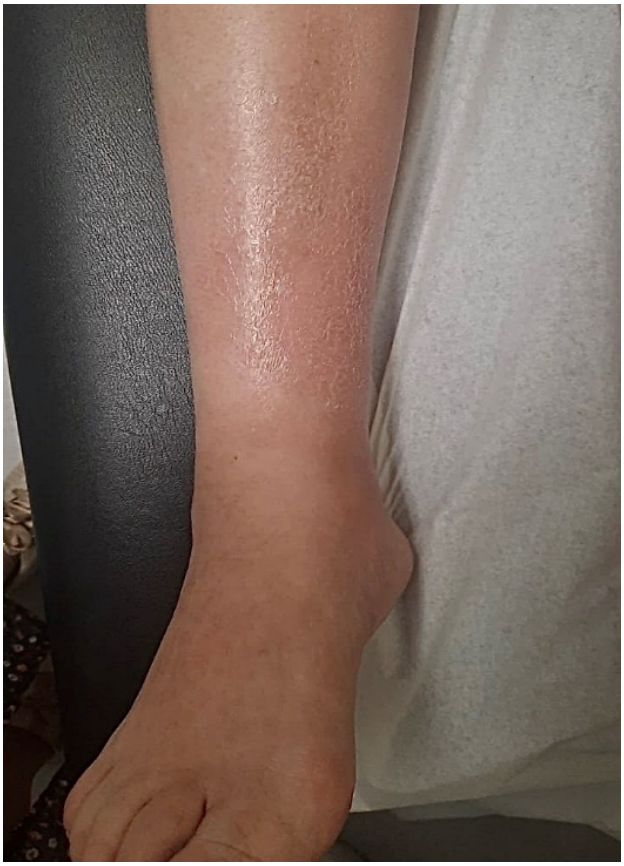

Figure 2. The "peau d'orange" aspect observed in the patient's right leg.

The diagnosis of EF was made, and the patient was started on prednisolone at a dose of 1mg/kg/day. Methotrexate was later introduced, and prednisolone were gradually tapered down until its suspension. The patient achieved clinical and analytical remission.

This case addresses a condition that presents a differential diagnosis with systemic sclerosis (SS). Clinical distinctions between EF and SS include the absence of Raynaud's phenomenon, normal periungual capillaroscopy, and rarely involvement of hands and face in EF. SS may affect multiple organs, unlike the primarily cutaneous involvement of EF. Specific antibodies for SS contribute to the differential diagnosis[1]. The distinction of these entities is very important, as they have different treatment approaches.

REFERENCES

1. Mazilu D, Boltaşiu Tătaru LA, Mardale DA, et al. Eosinophilic Fasciitis: Current and Remaining Challenges. Int J Mol Sci. 2023;24(3):1982. https://doi.org/10.3390/ijms24031982
2. Ihn H. Eosinophilic fasciitis: From pathophysiology to treatment. Allergol Int. 2019; 68(4): 437-439. https://doi.org/10.1016/j.alit.2019.03.001
3. Jinnin M, Yamamoto T, Asano Y, et al. Diagnostic criteria, severity classification and guidelines of eosinophilic fasciitis. J Dermatol. 2018;45(8):881-890. https://doi.org/10.1111/1346-8138.14160

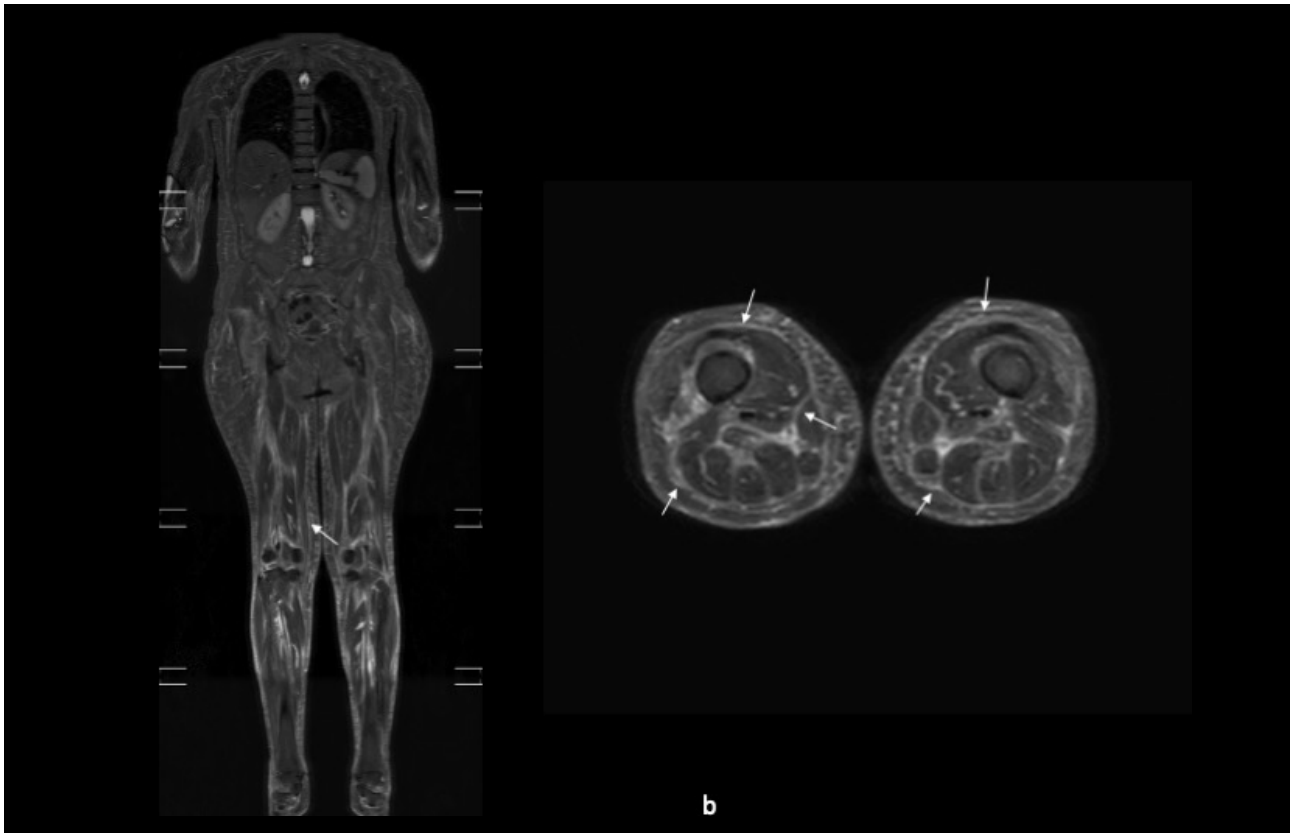

Figure 3. a) Coronal STIR image show symmetrical diffuse fascial thickening involving the compartments of the lower limbs, arms and forearms; b) An axial STIR image of bilateral legs with diffuse fascial thickening and increased signal intensity with muscle sparing.