Posttraumatic Eosinophilic Fasciitis: Diagnostic Usefulness of Magnetic Resonance Imaging

Fascitis eosinofílica asociada a traumatismo: utilidad diagnóstica de la resonancia magnética

To the Editor:

Eosinophilic fasciitis is a rare scleroderma-like disorder that mainly affects the muscle fascia. It usually occurs in adults between the ages of 20 years and 70 years.\(^1\)

We present the case of a 54-year-old patient with no past medical history of interest, who came to the clinic because of a rapidly progressive induration of the limbs, abdomen and lower back 1 month after an accidental fall (Fig. 1A). The condition had started as edema and erythema of the lower limbs, spreading to other areas, with the subsequent appearance of induration. The diagnoses considered were eosinophilic fasciitis, scleroderma, scleredema, and scleromyxedema. The patient did not present Raynaud phenomenon, dysphagia, or sclerodactyly.

Complete blood count showed a raised white cell count with marked eosinophilia (3.10\(^{10^3}\)/\(\mu\)L, 22%). Other laboratory tests, including the erythrocyte sedimentation rate, antinuclear antibodies and rheumatoid factor, were normal or negative.

Histopathology of a deep biopsy of the forearm showed interlobular septal thickening and an inflammatory infiltrate with numerous eosinophils affecting the septa and adjacent subcutaneous tissue, which was moderately edematous (Fig. 2). There were no changes of note in the epidermis and dermis; the muscle fascia could not be assessed. Magnetic resonance imaging (MRI) of the lower limbs showed thickening of the muscle fascia with signal hyperintensity on short TI inversion recovery sequences, indicating inflammation (Fig. 3).

The patient was diagnosed with eosinophilic fasciitis on the basis of the peripheral eosinophilia, histological findings, and the fasciitis observed on MRI. Treatment was initiated with oral prednisone at a dose of 1 mg/kg/d and hydroxychloroquine, with the subsequent addition of methotrexate to reduce the necessary dose of corticosteroids. The clinical course was favorable, with blood parameters returning to normal at 2 months and disappearance of the induration after 12 months of treatment (Fig. 1B).

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Figure 1 Photograph of the forearm. A, Pinching the skin is difficult due to induration. B, There is a decrease in induration after 12 months of treatment.

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References

Eosinophilic fasciitis was first described by Shulman in 1974 in patients with diffuse fasciitis and peripheral eosinophilia; it is clinically characterized by induration of the subcutaneous tissues, predominately affecting the limbs, preceded by an initial stage involving erythema and edema. Peripheral eosinophilia is a typical finding and is present in up to 80% of cases. In general, there is no visceral involvement, Raynaud phenomenon, or sclerodactyly. Its appearance has been associated with various triggers such as extreme physical exercise, insect bites, *Borrelia* infection, and the use of certain drugs such as statins. Toxic oil syndrome caused by adulterated rapeseed oil has also been associated with the disorder. To date, only 5 cases have been reported following an uncomplicated trauma. In our case, the close temporal association with the accidental fall and the absence of other predisposing factors suggests that this was the trigger. A similar case has been described in the literature, in which eosinophilic fasciitis developed 1 week after a fall. It has been suggested that in these patients the trauma could reveal antigenic properties of the fascia and subcutis, thus initiating the process. If eosinophilic fasciitis is suspected, a full-thickness biopsy is needed to determine whether the muscle fascia is affected. Histology shows fibrous thickening of the fascia with a diffuse lymphoplasmocytic inflammatory infiltrate and varying numbers of eosinophils. These changes often extend locally into the interlobular septa of the adjacent adipose tissue and into the muscle layers.

The role of MRI has become of increasing importance in the diagnosis of eosinophilic fasciitis and in its follow-up. Characteristic findings include thickening and increased signal intensity of the muscle fascia on T2-weighted and short T1 inversion recovery sequences, with enhancement following administration of contrast material on T1-weighted sequences; these changes correspond to the degree of microscopic inflammation. However, in some patients the biopsy is suboptimal or cannot be performed, and thus the findings from MRI are used to identify involvement of the fascia and reach a diagnosis. Our case provides a novel example of eosinophilic fasciitis that was diagnosed by MRI after biopsy proved inconclusive.

Therefore, although biopsy remains the diagnostic procedure of choice, MRI can be very useful to confirm inflammation of the muscle fascia in cases in which this cannot be assessed on the biopsy or when the findings are inconclusive. In addition, by confirming that the process mainly affects the fascia, MRI helps to distinguish eosinophilic fasciitis from other disorders such as polymyositis or scleroderma. Attention has also been drawn to its usefulness when selecting the optimal area from which to take a biopsy, and to identify improvement in fascial inflammation after starting treatment.

In conclusion, the case presented demonstrates the usefulness of MRI in establishing a diagnosis of eosinophilic fasciitis in patients when histopathological findings are inconclusive. In addition, a history of an accidental fall in the absence of other precipitating factors supports the role of trauma as a possible trigger of the disease.

References


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Malignant Fibrous Histiocytoma in a Scar From Excision of a Melanocytic Nevus

Histiocitoma fibroso maligno sobre cicatriz tras extirpación de un nevus melanocítico

To the Editor:

Malignant fibrous histiocytoma, considered the most common soft tissue sarcoma in adults, can be located in any organ, since it arises from mesenchymal tissue. In the skin this tumor presents as subcutaneous nodules whose texture varies from elastic to firm. The pathogenesis of these tumors is unclear.¹

Malignant fibrous histiocytoma in a scar has been reported only occasionally and most cases have developed in postburn scars (8 in the literature).²–⁴

The diagnosis of malignant fibrous histiocytoma is one of exclusion, reserved for cases of undifferentiated pleomorphic sarcomas not otherwise specified. Immunohistochemistry is essential; neoplastic cells must be positive for vimentin and negative for cytokeratin and protein S-100,² while CD68 expression neither supports nor excludes diagnosis.

The prognosis for patients with this tumor is generally considered to be poor.

Wide surgical excision after diagnosis is associated with the best survival statistics. Radiation therapy combined with surgery is also appropriate, particularly when there is evidence of high-grade malignancy and surgical margins are positive.¹

Atypical fibrous xanthoma is difficult to distinguish from a superficial malignant fibrous histiocytoma and these 2 diagnoses are currently included within the same spectrum. For some authors, the involvement of subcutaneous tissue, fascia, and muscle, along with intense vascular invasion or necrosis, is an indication of malignant fibrous histiocytoma.⁵

We present the case of a 78-year-old woman who was allergic to penicillin, its derivatives, and sulfonamides and whose medical history included type 2 diabetes mellitus, osteoarthritis, hypertension, and glaucoma.

In 2003 a compound cellular nevus was excised from the right ear lobe, and subsequent follow-up revealed a keloid scar in the area of excision.

In 2007 the patient came to the emergency department with a rapidly growing tumor in the scar (Fig. 1). A skin biopsy revealed a tumor with a fascicular pattern, cells with elongated nuclei and marked atypia, including the presence of large nuclei and giant cells; mitotic activity was high. Immunohistochemistry was positive for CD68, vimentin, and cytokeratin, and negative for AE1/AE3, epithelial membrane antigen, actin, desmin, CD34, C-kit; Ki-67 (Figs. 2 and 3) expression was high. These data led to a diagnosis of malignant fibrous histiocytoma.

Surgical excision confirmed infiltration of muscle and the parotid gland. After analyzing the results, an interdisciplinary committee decided on radical excision. Follow-up data on clinical course is unavailable.

Physicians have been familiar with the malignant degeneration of previously damaged tissue for some time. Squamous cell carcinoma is the most common histological type of malignant degeneration arising in scar tissue; basal cell carcinoma is the next most common. Other types include melanoma, liposarcoma, osteosarcoma, adenocarcinoma, and fibrosarcoma.

The literature contains few reports of malignant fibrous histiocytoma in scars, and the pathogenesis is unclear.