Diagnostic utility of Magnetic Ressonance Imaging in Eosinophilic Fasciitis

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A 30-year-old caucasian woman was referred to our Rheumatology Unit, with a 6-month history of non-pitting edema and myalgia of the arms and legs that progressively evolved into skin thickening, sparing however the skin of the hands and feet. The skin took on an exuberant woody appearance and some areas of the arms and legs revealed an "orange peel" texture (Figure 1), with no other inflammatory signs.

There was no history of arthralgia, dyspnea, cough, dysphagia or constitutional symptoms and Raynaud phenomenon was absent.

Regarding the medical history, the patient was being treated with fluoxetine 20mg/per day for depressive syndrome and denied recent start of new drugs.

Other than the above described skin changes the patient presented a mild tetraparesis (grade IV/V), predominantly proximal, with no other relevant findings on the neurological examination. Physical examination was otherwise normal.

Laboratory investigation revealed peripheral eosinophilia (900x10^9/µL), increased erythrocyte sedimentation rate (99 mm/h) and C-reactive protein (3.65mg/dL) and a polyclonal hypergammaglobulinemia. The screening for anti-nuclear antibodies was negative and creatine-kinase and aldolase were both within normal values.

In the context of myalgia complaints, an electromyography of upper and lower limbs was performed, that showed no signs of muscle fiber injury. The patient was also submitted to a Magnetic Resonance Imaging (MRI) of the lower limbs which revealed diffuse thickening and increased T2 signal of superficial and deep fascia involving all muscle groups, with no abnormalities found in muscle fibers, compatible with the diagnosis of eosinophilic fasciitis (EF) (Figure 2).

The fascia and muscle biopsy of the upper arm confirmed the diagnosis of EF, showing significant thickening of the superficial fascia accompanied by an inflammatory infiltrate formed by lympho-mononuclear cells with a significant number of eosinophils (Figures

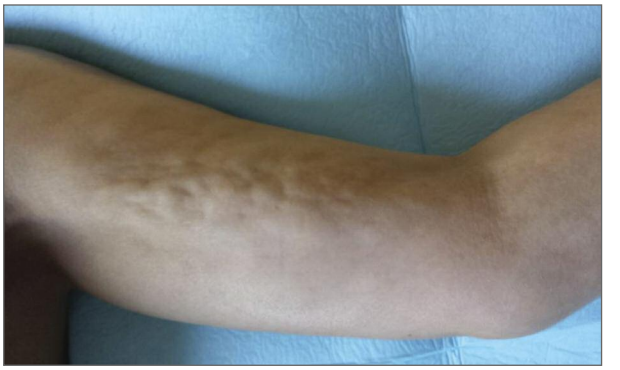

FIGURE 1. Orange peel appearance of the skin of the upper arm

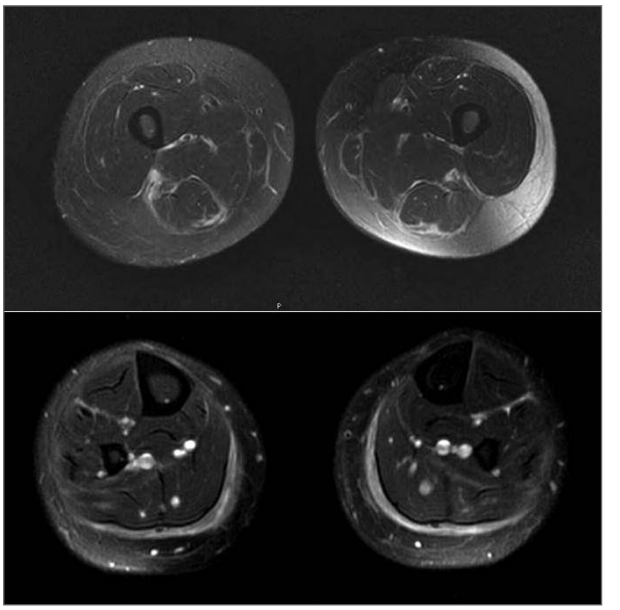

FIGURE 2. T2 weighted MRI images of both thigs and legs showing diffuse thickening and increased T2 signal of superficial and deep fascia, involving all muscle groups

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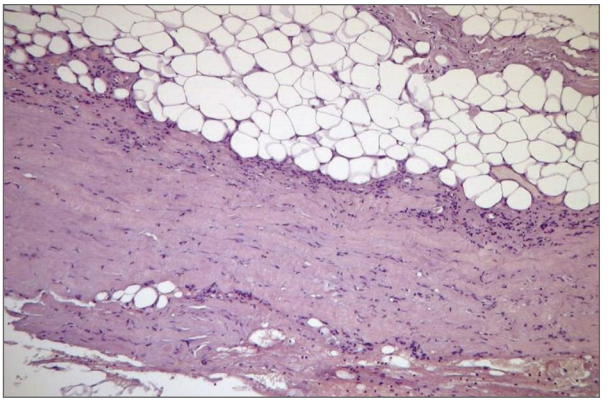

FIGURE 3. Fascia and muscle biopsy showing significant thickening of the superficial fascia accompanied by an inflammatory infiltrate

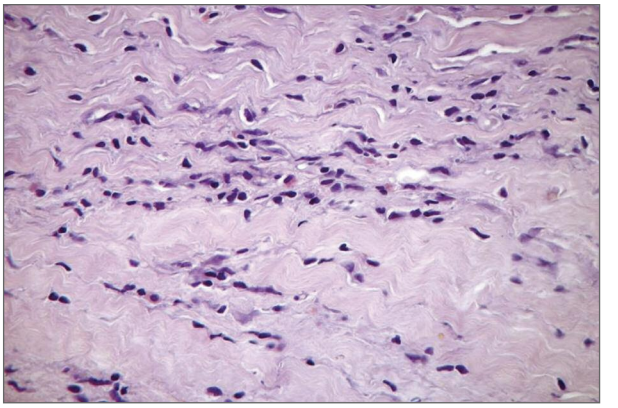

FIGURE 4. Fascia and muscle biopsy showing inflammatory infiltrate with predominance of eosinophils

3 and 4).

Although classically, the diagnosis of EF is confirmed by skin-to-muscle biopsy, the interest of MRI in diagnosing EF has increased in recent years. The thickening of the deep fascia, involving all muscle groups, with hyperintense signal on T2, is the most characteristic finding.[1] Abnormalities are typically limited to the fascia, sparing muscles and hypodermic fat, and thus allowing differential diagnosis with myositis and other inflammatory myopathies and potential lethal clinical situations as necrotizing fasciitis. MRI also helps detect the best place for biopsy, which could be important in mildest presentations of the disease[2].

Although there is no general consensus, some authors have proposed MRI findings as a minor diagnostic criteria for EF, being an important additional criteria, especially when eosinophilia is absent on biopsy[3]. In this context, a small number of case-reports highlighted the diagnostic value of MRI in establishing a diagnosis of EF without the need for biopsy[4].

Some studies also suggest the usefulness of MRI on the follow-up of these patients, as its findings tend to correlate with the disease evolution, but no strict criteria have been defined to assess the response to treatment[2].

Despite the promising features, prospective studies are needed do define the sensitivity and specificity of this imaging technique and prove its value in the diagnosing and prognosis of EF[4-5].

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