Case 15718

Eurorad

Eosinophilic fasciitis

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Section: Musculoskeletal system
Area of Interest: Musculoskeletal soft tissue
Procedure: Imaging sequences
Procedure: Perception image
Procedure: Image compression
Imaging Technique: MR
Special Focus: Inflammation Case Type: Clinical Cases
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Patient: 35 years, male

Clinical History:

A 35-year-old male patient with no relevant medical history presents pain and an increased diffuse volume of the four limbs of 1 month of evolution. The laboratory analysis shows an elevated C-reactive protein (CRP), with eosinophilia and hypergammaglobulinaemia.
Imaging Findings:

The MRI shows a thickening and a generalised oedema of the deep fascial planes of all the muscular compartments of the thighs (Fig. 1 and Fig. 2). After the gadolinium administration, a diffuse and an increased contrast uptake was observed in all deep fascial planes of the legs (Fig. 3a and Fig. 3b). The images show (Fig. 2) a discrete oedema of the subcutaneous cellular tissue adjacent to the deep muscular fascia, with no signal alterations of the muscles or the bones structures.
In our patient the diagnosis of the disease was reached, adding the clinical history, the alterations in the laboratory tests, the MRI images and a skin biopsy.
Discussion:

Eosinophilic fasciitis (Shulman's syndrome) is a rare connective tissue disorder. The aetiology, the incidence and the pathophysiology are unclear [1, 2].

The clinical presentation is characterised by the cutaneous manifestations that initially include extremities oedema, usually in a symmetrical way and sometimes with a painful presentation, that gradually gives way to skin induration sclerodermiform-like (orange peel skin) [1, 2].

Eosinophilia is a frequent finding; however, it’s not an essential analytical alteration for the diagnosis; there is not a relationship between the degree of alteration and the severity of the clinical presentation. Normally the eosinophilia tends to decrease once the disease has been established. In addition, similar to other systemic inflammatory conditions, it is common to find elevation in the erythrocyte sedimentation rate, CRP and immunoglobulins [1, 2].

The diagnostic study has to be completed with a full-thickness skin biopsy (from the skin to the muscle tissue). The anatomopathological findings include the thickening and the infiltration of the deep fascia by lymphocytes and a variable percentage of eosinophils, the latter may even be absent, independent of blood eosinophil levels [1, 2].

MRI has been regarded as a fundamental part of the study of this pathology, given that it provides information that

helps establish the diagnosis; helps the physicians select a proper site to perform a biopsy according to the degree of alteration observed; and it helps monitor the course of the disease and the response to the treatment. It has been described that the findings observed in the MRI images have a good correlation with the clinical evolution of the disease [3].

The protocol for the MRI study of this disease should include T1-weighted images and sequences with fat suppression of both extremities, since the alterations are usually bilateral, with the purpose to provide evidence of the thickening and the pattern of fascial oedema. It is not necessary to carry out routine MRI with contrast, although it emphasises the conspicuity of the findings [3, 4, 5].

Although there is no consensus regarding the treatment, it is considered that the first line drug intervention is the administration of corticosteroids at high doses (0.5–1mg/kg/day), followed by a progressive decrease of the doses, depending of the clinical course. In general, with a very high favourable clinical and imaging response; as in the case of our patient who presented a decrease in oedema, and normalisation of laboratory tests. In cases with steroid resistance, immunosuppressants are used as a second line treatment [1, 2].
Differential Diagnosis List: Eosinophilic fasciitis, Scleroderma / morphoea, Erysipelas, Oedema by ectasia, Necrotising fasciitis, Myositis

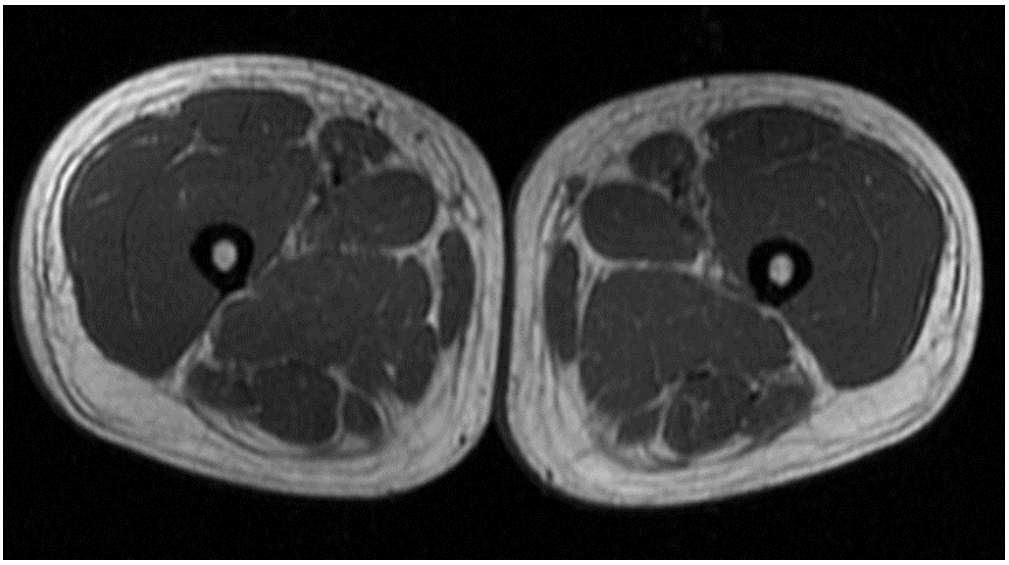
Final Diagnosis: Eosinophilic fasciitis

References:

Lebeaux D, Sène D. (2012) Eosinophilic fasciitis (Shulman disease). Best Pract Res Clin Rheumatol 26: 449-58 (PMID: 23040360)
Antic M, Lautenschlager S, Itin PH. (2006) Eosinophilic fasciitis 30years after-what do we really know? Report of 11 patients and review of the literature. Dermatology 213: 93-101 (PMID: 16902285)
Moulton S, Kransdorf M, Ginsburg W, Abril A, Persellin S. (2005) Eosinophilic Fasciitis: Spectrum of MRI Findings. Am J Roentgenol 184(3): 975-978. (PMID: 15728627)
Kirchgesner T et al. (2016) Eosinophilic fasciitis: Typical abnormalities, variants and differential diagnosis of fasciae abnormalities using MR imaging. Diagn Interv Imaging 96(4):341-8. (PMID: 25746223)
Malghem J et al. (2013) Necrotizing fasciitis: contribution and limitations of diagnostic imaging. Joint Bone Spine 80(2):146-54 (PMID: 23043899)

Figure 1

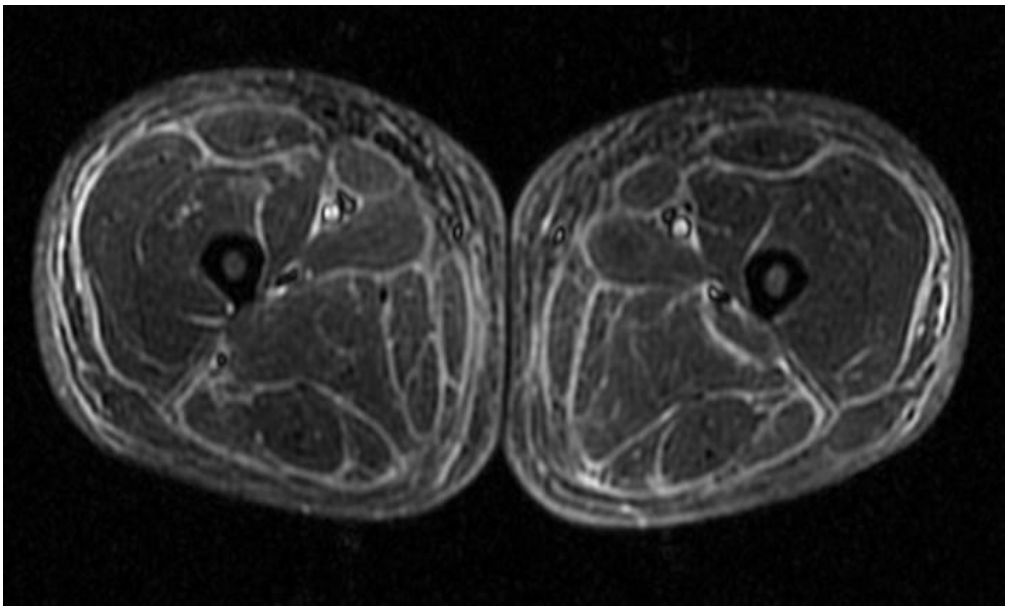
a



Description: Axial image of the thighs in T1-weighted sequence. There is bilateral diffuse thickening of the thighs fascia. **Origin:** ACIM, Hospital Universitario y Politécnico La Fe, Valencia, Spain.

Figure 2

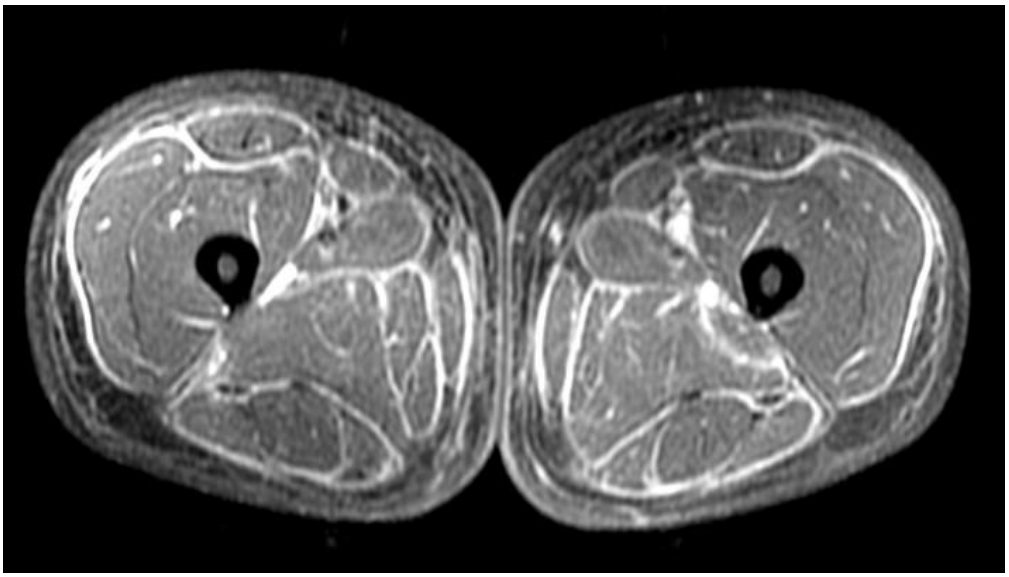
a



Description: Axial image of thighs in STIR sequence. There is a marked diffuse hyperintensity of the thighs fasciae. No signal alterations are observed in the adjacent musculature. **Origin:** ACIM, Hospital Universitario y Politécnico La Fe, Valencia, Spain.

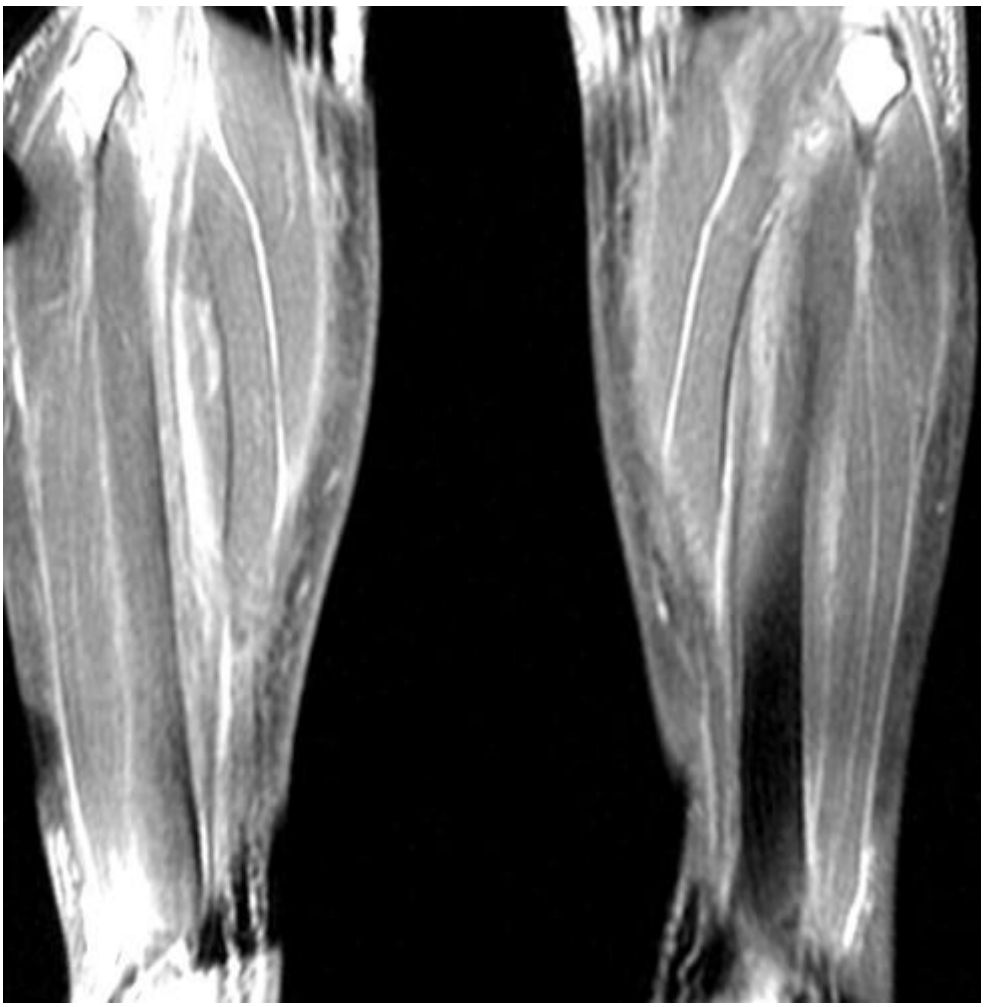
Figure 3

a



Description: Axial images of the thighs in T1-weighted sequence with fat suppression after administration of gadolinium. A thickening and a diffuse hyper-uptake contrast of the bilateral fascia of the extremities is observed. **Origin:** ACIM, Hospital Universitario y Politécnico La Fe, Valencia, Spain.

b



Description: Coronal images of the legs in T1-weighted sequence with fat suppression after administration of gadolinium. A thickening and a diffuse hyper-uptake contrast of the bilateral fascia of the extremities is observed. **Origin:** ACIM, Hospital Universitario y Politécnico La Fe, Valencia, Spain.