

CLINICAL CASE - TEST YOURSELF

Musculoskeletal Imaging

A 68-year-old female patient presenting with peripheral arthralgia

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PART A

A 68-year-old female patient presented to the Rheumatologic Outpatient Clinic with peripheral arthralgia of hands and feet for the last 3 months. Her past medical history included a thyroid cancer 25 years earlier.

Clinical examination revealed mild symmetrical polyarthrititis with skin thickening and sclerosis of the upper extremities, more prominent on the right

humerus (**Fig. 1**).

Laboratory work-up showed slightly elevated ESR and CRP and borderline blood eosinophilia. The complete blood count was normal and RF autoantibodies, anti-CCP and ANA were negative. She was referred for further evaluation with magnetic resonance (MR) imaging of the right upper limb (**Figs. 2-4**).



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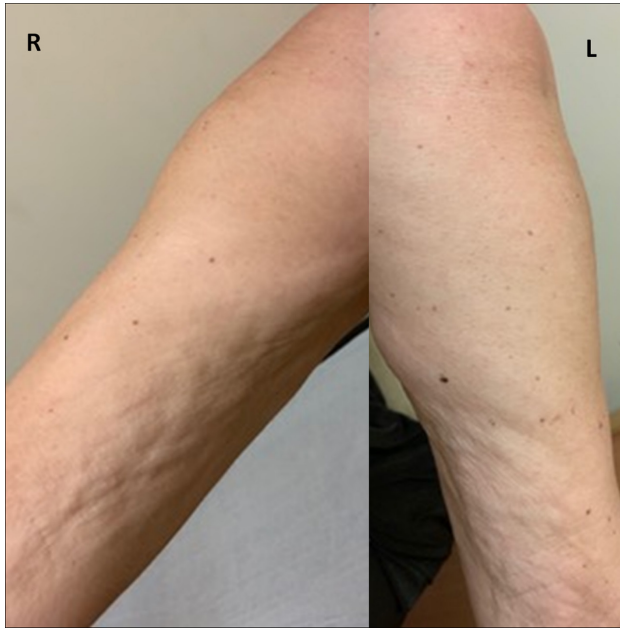


Fig. 1. Photograph of the patient's right and left arm at presentation.

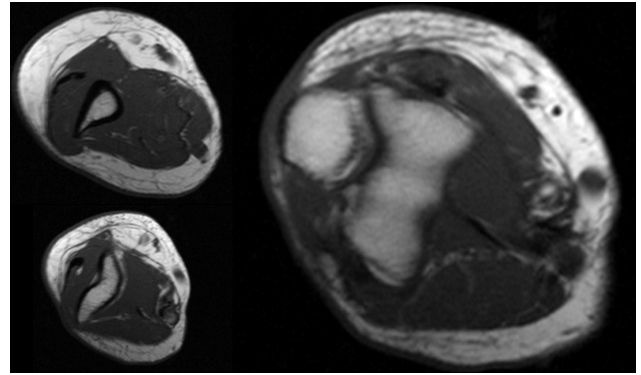


Fig. 2. Axial T1W MR images.

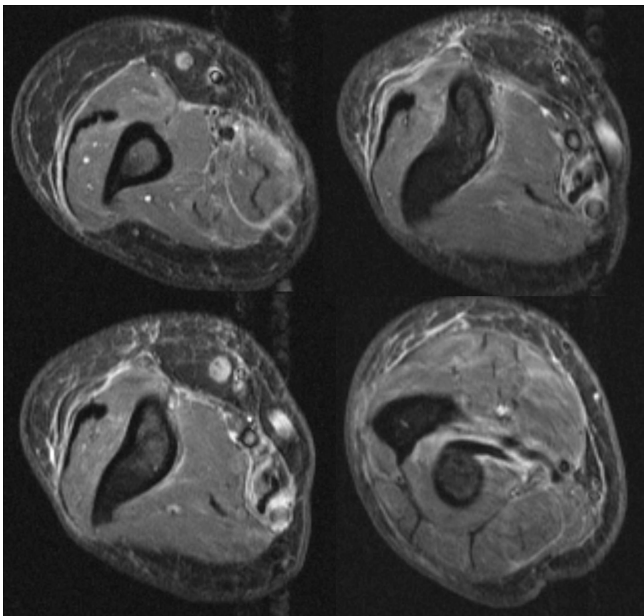


Fig. 3. Axial fat suppressed PDW MR images.

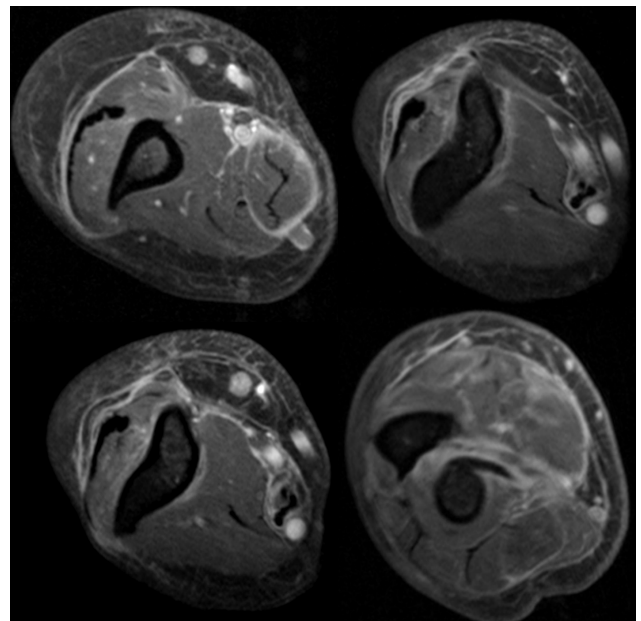


Fig. 4. Axial fat suppressed contrast-enhanced T1W MR images.

PART B

Diagnosis: Eosinophilic fasciitis

Eosinophilic fasciitis (EF), also known as “Shulman's syndrome”, is a rare connective tissue disorder, included in the scleroderma-mimics spectrum, first described in 1974 [1]. EF affects men and women equally, often aged between 40 and 50 years and its aetiology is still unclarified [2]. The disorder has been related to exercise, drugs, infection and exposure to chemicals as well as autoimmune mechanisms. Histologically there is thickening of the fascia and infiltration with lymphohistiocytes, plasma cells and occasionally eosinophils in the early stages [3, 4]. Hyalinisation of the collagen leads to sclerosis in later stages [5].

Clinically, EF in the early stages manifests as an abrupt development of a usually symmetrical swelling that gradually progresses into indurations and thickening of the skin, affecting the extremities and trunk and typically sparing the hands and feet [6]. Associated features include erythema, hyperpigmentation and sensitivity of the dermis and systemic changes such as inflammatory arthralgias and myalgias [2, 6]. Characteristic cutaneous manifestations of EF are a) the “groove sign”, representing furrows along the course of the superficial veins and b) the “peau d' orange” appearance of the skin. The above are considered hallmarks of the disease but are present only in about 50% of cases [3].

Without proper treatment, EF can lead to limited range of motion due to joint contractures or tendon retractions [6]. Involvement of the trunk, joint contractures and fibrosclerosis of the dermis, seen in advanced stages, morphea-like lesions and patients younger than 12-year-old, are considered poor prognostic factors [2, 3, 6]. A wide variety of diseases, such as localised scleroderma (morphea), autoimmune (systemic lupus erythematosus, rheumatoid arthritis, autoimmune thyroiditis and others) and haematological (aplastic anaemia, myelodysplastic syndrome, myeloproliferative syndromes and others) disorders, have been associated with EF, complicating up to 30% of the cases [1, 3].

The clinical differentials showing diffuse skin thickness include systemic sclerosis and its subtypes, epidemic fasciitis syndromes caused by toxic agents' lymphomas, scleroderma, nephrogenic systemic fibrosis and scleromyxoedema [5, 7]. The presence of acute on-

set's limb swelling has a totally different differential list consisting of deep-vein thrombosis, erysipelas, stasis oedema, dermatomyositis, necrotising fasciitis, pyomyositis and acute denervation [5, 8].

Laboratory analysis usually reveals an inflammatory response with elevated CRP and ESR, accompanied by hypergammaglobulinaemia and elevated aldolase levels. Peripheral eosinophilia is highly suggestive, being present in 60-90% of cases. However, the latter is temporary and unremarkable at later stages and its absence cannot exclude the diagnosis or determine the prognosis. Furthermore, it cannot be used as a follow-up index, in contrast to aldolase levels, which seem to relate with the activity of the disease [6].

The gold standard for the diagnosis of EF is a full thickness biopsy (skin to muscle) [2]. Recently, Pinal-Fernandez et al. suggested a combination of diagnostic criteria which can lead to replacing biopsy [4]. Subsequently, MR imaging findings of hyperintensity of the fascia on fluid sensitive sequences, in conjunction with the compatible clinical background, are highly suggestive of EF [2].

Imaging methods that are increasingly used in the diagnosis and monitoring of EF are ultrasound (US) and MR imaging [5, 6, 8]. Both methods can highlight the fascial involvement and guide skin-muscle biopsy. US in EF reveals thickening of the superficial and deep fascia, sometimes followed by soft tissue oedema or fluid surrounding the fascia [5, 8]. The compressibility of the subcutaneous tissue can be proved useful in the diagnosis of EF [7].

MR imaging findings in the acute phase include thickened fasciae (superficial, deep or both) on T1-weighted (W) images, increased signal intensity on fluid sensitive sequences and enhancement of the affected fasciae on T1W after gadolinium injection. The subdermal tissues may show fine linear oedema on fluid sensitive sequences, typically without enhancement [9]. MR imaging aids in differentiating EF from other muscle and subcutaneous disorders, showing little if any involvement of the muscles, mainly as increased signal intensity of the fibers adjacent to the fascia [9]. Differential diagnosis with necrotising fasciitis is based on MR imaging regarding the thicker, low



Fig. 1. Orange peel appearance of the skin of both upper arms (arrows).

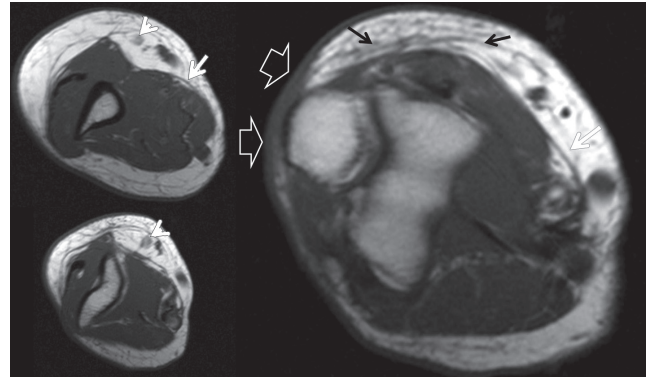


Fig. 2. Axial T1W MR images show slight thickening of the skin at the level of olecranon (open arrows), subcutaneous oedema (black arrows), thickening of the superficial fasciae (white arrows) and small lymph nodes (white arrowheads).

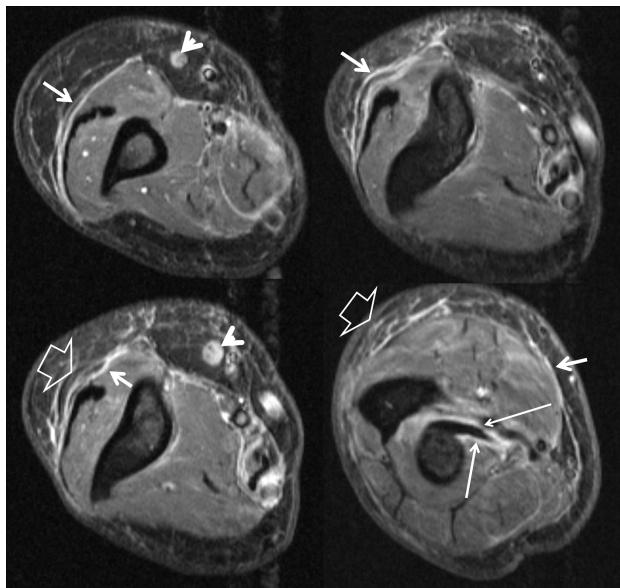


Fig. 3. Axial fat suppressed PDW MR images show signal hyperintensity in superficial fasciae (arrows), deep signal hyperintensity around the distal biceps tendon insertion (long thin arrows), subcutaneous soft tissue oedema (open arrows) and small lymph nodes (arrowheads).

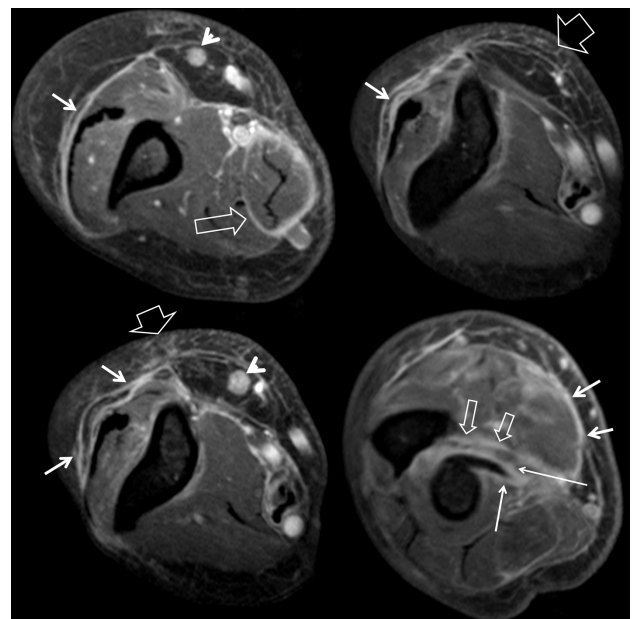


Fig. 4. Axial fat suppressed contrast-enhanced T1W MR images show enhancement of the superficial fasciae (thin short arrows), around the distal biceps tendon insertion (long thin arrows), deep fasciae (thin open arrows), subcutaneous soft tissue (thick open arrows) and the small lymph nodes (arrowheads).

signal on T2W images and non enhancing portions of the abnormal deep fascia and involvement of three or more compartments in one extremity [10]. After the initiation of treatment, MR imaging findings resolve in those who respond, whereas resistant cases show incomplete regression of imaging findings, which mirrors the disease's activity [2, 9].

Our patient presented with the typical "peau d' orange" skin appearance (Fig. 1), slightly elevated ESR and CRP, borderline peripheral eosinophilia (500 cells/ μ L), and typical MR imaging findings, consisting of subcutaneous oedema, skin thickening, superficial and deep fascial hyperintensity and abnormal enhancement (Figs. 2-4). Interestingly, there were also lymph nodes in the subcutaneous tissues, enhancement of the subcutaneous oedematous fatty tissue, and peritendinous hyperintensity and enhancement, not previously described in the literature. The MR imaging findings suggested the diagnosis of EF.

Treatment consists of corticosteroid administration, either oral alone or combined with intravenous pulse therapy, which are highly effective in EF. Methotrexate was regarded a second-line therapy, but is currently commonly used combined with systemic corticosteroids as the first treatment choice, with a higher effec-

tiveness. Physiotherapy could also be beneficial for patients with limited range of motion [6]. The possibility of a relapse or of an appearance of haematological disorders such as aplastic anaemia makes a close clinical monitoring of great importance [7].

Treatment's early initiation is related with better outcome and even complete resolution of clinical findings [6]. The amelioration of the long-standing EF's symptoms, such as skin induration and joint contractures, might require a longer recovery time [7]. Our patient received betamethasone i.m. on the first visit. Following establishment of diagnosis, treatment consisted of per os methylprednisone (20 mg with gradual tapering) and 15 mg methotrexate with rapid improvement of arthritis. Arthritis/arthralgia symptoms improved quickly after the i.m. steroid administration and after a month of receiving methotrexate and p.o. methylprednisone, the patient presented with moderate improvement of skin thickness.

In conclusion, MR imaging shows typical findings for establishing the diagnosis of EF, obviating thus the need for biopsy. **R**

Conflict of interest

The authors declared no conflicts of interest.



KEY WORDS

Eosinophilic fasciitis; Magnetic resonance imaging; Eosinophilia/diagnosis; Glucocorticoids/therapeutic use; Methotrexate/therapeutic use; Connective tissue diseases

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