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CASE REPORT

From diagnosis to remission: place of MRI in eosinophilic fasciitis

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Eosinophilic fasciitis is a rare disease, but it must be considered in patients with adult myalgia. Here, we report the case of 32-year-old man who presented with a 4-month history of bilateral myalgia of the lower limbs, which subsequently spread to the upper limbs. There was no specific medical history, no trauma, and no intense activity. Physical examination revealed induration of the skin with irreducible flexion of the fingers as observed in scleroderma. Laboratory examination showed peripheral eosinophilia associated with moderate elevation of both the erythrocyte sedimentation rate and C-reactive protein. No infectious, neoplastic, hemopathic, or immunological abnormality was detected. Magnetic resonance imaging (MRI) showed thickening of the muscular fascias of the thigh in high signal intensity on T2-weighted images, slightly enhanced after contrast agent injection on T1 images, without myositis. MRI-guided muscle biopsy demonstrated fascia infiltration characterized by mononuclear inflammatory cells and polynuclear eosinophils. A diagnosis of eosinophilic fasciitis was confirmed and the patient received prednisolone. One month later, he reported improvements in general health, pain, motion, joint mobility, and skin induration associated with normalization of both hypereosinophilia and biologic inflammation. After 12 months, clinical MRI and laboratory parameters were normal and the patient was considered to be in clinical remission.

Keywords Eosinophilic fasciitis · Follow-up · MRI

Introduction

Adult myalgia occurs in a wide range of disorders. One of them, eosinophilic fasciitis (EF), is a rare entity that predominantly affects adults aged about 40 years but has been reported in all age groups. EF is associated with myalgia, skin induration similar to scleroderma, and peripheral eosinophilia. Diagnosis is confirmed by muscle biopsy showing fascia infiltration of inflammatory cells. We report the case of a young (32 years old) patient in which MRI helped to guide the diagnosis and biopsy, enabled us to start treatment early with a better prognosis, and confirmed remission at 1 year.

Observation

A 32-year-old man presented with a 4-month history of diffuse myalgia with no traumatic factors, intense exercise, or sports activity. Pain was exacerbated by exercise, had started in the calves, and had spread to the thighs. At 5 months, pain had spread to the forearms. His general health had deteriorated, with weight loss of 4 kg and asthenia. Treatment with NSAIDs and analgesics was ineffective.

This patient was of Algerian origin, had no medical history, and reported no allergy or atopy. He also made no

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mention of recent travel, vaccination, drug consumption, or contact with any toxins prior to the onset of symptoms.

On examination, there was no peripheral arthritis. Joints were normal with the exception of the fingers of the left hand, in which a retractile flexed pattern was noted. Palpation of muscles was painless; however, passive stretching of the calf muscles and forearms did cause pain. The dermis had a slightly stiff appearance as in scleroderma, with no relevant skin lesions. Neurological examination was normal. There was no lymphadenopathy or organomegaly, and the remaining physical examination was unremarkable.

Laboratory tests showed marked hypereosinophilia (2,360/mm^3), a moderate inflammatory syndrome (erythrocyte sedimentation rate (ESR) 20 mm in the first hour, C-reactive protein (CRP) 46.5 mg/L). Blood electrolytes and liver function were normal, as were thyroid hormones and muscle enzymes. Serology was negative for viruses (hepatitis B and C, HIV, parvovirus B19, cytomegalovirus, herpes virus), bacteria (brucellosis, chlamydia trachomatis and pneumoniae, borreliosis), and parasites (taeniasis, toxocariasis, trichinosis, schistosomiasis, fascioliasis). Immunological tests were non-contributory (antinuclear antibodies, anti-Scl 70, anti-centromere, anti-JO1, anti-RNP, anti-cytoplasmic). However, circulating immune complexes reached 4.8 g/L (normal value <1.5 g/L). There was no evidence for a malignant hematologic eosinophilia.

Chest and abdomino-pelvic CT revealed no lesions suggestive of malignancy. There was no destructive bone lesion on radiography. Functional respiratory investigation suggested a restrictive ventilatory disorder. MRI of the lower limbs (Fig. 1b) revealed an inflammatory thickening of the muscular fascia of the thighs on T2 and T1 sequences with fat suppression and after gadolinium injection. The inflammatory lesions disseminated to the superficial fascia of the gastrocnemius muscles. MRI signal of muscle was normal, with no sign of myositis. The lesions were indicative of a diffuse fasciitis, and MRI was very useful in guiding the surgical skin–muscle biopsy at the anterior thigh.

Histology (Fig. 1a) demonstrated septal cellulitis (only the epidermis, dermis, and hypodermis were analyzable), with lymphoplasmacytic and eosinophilic cells.

Diagnosis was therefore an eosinophilic fasciitis. Corticosteroids 0.5 mg/kg per day were started in association with a preventive regimen to protect against glucocorticoid-induced osteoporosis. In 1 month, treatment resulted in complete regression of the eosinophilia and inflammatory syndrome. With ESR 4 mm and CRP below 5 mg/L, corticosteroids could be gradually decreased. The patient reported improvements in general health, pain, and motion. Improvements of joint mobility and skin induration were also observed in the forearms and lower limbs. After 12 months of treatment, clinical and biological remission was confirmed and MRI showed muscle fascia of normal appearance (Fig. 1c). The patient was therefore considered to be in complete remission.

Discussion

Diffuse eosinophilic fasciitis was first described in 1974 by Laurence E. Shulman. Since that time, over 200 cases have been reported in the literature. It has long been a subject of debate, with some authors describing it as a clinical form of scleroderma. The current trend is towards considering it to be an autonomous entity as a systemic inflammatory rheumatic disease [1]. It predominantly affects adults aged about 40 years but can occur at any age and 30 pediatric

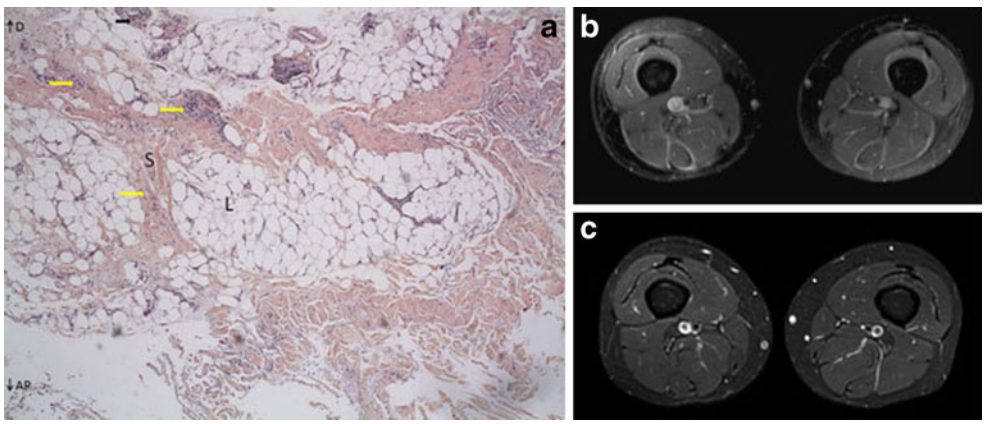

Fig. 1 **a** Biopsy of quadriceps fascia (H & E staining, magnification ×50). Deep dermis with fat lobules (*L*) and interlobular septas (*S*). Inflammatory infiltrate of septas (*yellow arrows*) and perivascular sites (*black arrow*), spread to deep aponevrosis (*AP*), composed with lymphocytes, mononuclear cells, and occasional eosinophilic cells. *D*, dermis. **b** Axial T2-weighted fat-suppressed magnetic resonance (MR) images, showing thickening and signal hyperintensity of muscular fascias, concerned both thighs. **c** Axial T2-weighted fat-suppressed MR images (water IDEAL sequences) after corticosteroid treatment, showing normal findings, revealed complete remission

cases are described in the literature [2]. The pathophysiology of this rare syndrome is not fully understood but is known to involve genetic, environmental, and systemic inflammatory (cytokine secretion) factors [1, 3]. Onset is often fast, sometimes following an intense physical effort (though not in our patient).

Symptoms usually begin with myalgia, quickly followed by swelling and induration of the subcutaneous structures. Symptoms principally affect the limbs, often symmetrically, and the distal parts of the forearms and/or legs are commonly involved. Sometimes the arms, back, hands, thighs, neck (anterior part), and trunk may be affected. Fever and deterioration of general health with weight loss are frequently encountered [3, 4]. Limitation of joint mobility due to infiltration of the skin is seen, particularly in the fingers where a scleroderma-like pattern has been described. True synovitis has occasionally been reported, as has tenosynovitis [4].

Visceral lesions are exceptional but may affect the esophagus (dystonia and GERD), liver (hepatomegaly), lung and respiratory system (restrictive ventilatory disorder), and nervous system (polyneuropathy) [3, 4]. Severe complications such as myelodysplasia and lymphoma have been described several months after a diagnosis of fasciitis and are considered as bad prognostic factors [5].

Hypereosinophilia (> 500/mm^3) is seen in more than 80% of cases, and the sedimentation rate is increased in 30%. Currently, diagnosis is based on the skin–muscle biopsy. Histologically, sclero-inflammatory damage is limited to the fascia in the early forms, with associated edema, perivascular infiltrate (lymphocytes, plasma cells, and eosinophils), and deposition of immunoglobulin G and complement [2–4]. In more advanced forms, sclerosis and fascia thickening reaching several centimeters have been described without eosinophilic infiltration at this stage [5]. In our observation, the presence of eosinophils on histology corresponds to the early inflammatory phase of the disease.

In patients with inflammatory myalgia, MRI can play a very useful role in: (1) highlighting fasciitis, (2) eliminating a diagnosis of myositis, and (3) guiding the skin–muscle biopsy. Classically, it is done for iconographic purposes or to monitor therapy. Reported in only 20 patients in the literature, MRI shows high signal intensity on T2-weighted sequences or STIR, of muscle fascia, with enhancement after gadolinium injection on T1-weighted sequences, while muscle pattern is normal [6–14]. These intense signals concern mainly the deep fascia of extensor muscles, in contrast to polymyositis and dermato-polymyositis, in which the fascia of the flexor muscles are more often affected. Enhancement of the fascia on the injected sequences is correlated with the degree of microscopic inflammation fasciitis [8, 13]. The original aspect of this observation was the contribution MRI made to the diagnosis by guiding biopsy. Furthermore, three new cases have been reported, illustrating the diagnostic value of MRI in establishing a diagnosis of eosinophilic fasciitis without the need for muscle biopsy [12, 15]. In our case, MRI confirmed the relationship between inflammation of the fascia in MRI and histological data.

The treatment of eosinophilic fasciitis is corticosteroid at a dose of 0.5 or 1 mg/kg/day in case of failure [1–5]. Prognosis is favorable with conventional steroids at an early stage, but treatment seems less effective in chronic disease [5]. In our patient, corticosteroids started at 0.5 mg/kg led, within a month, to decreased skin induration and the complete regression of eosinophilia and inflammation. At 12 months of treatment, the patient was considered to be in clinical and biological remission and MRI was normal with an "ad integrum" pattern of muscle fascia. Only sporadic cases published in radiological reviews have described MRI changes after treatment. A decrease in inflammation of the fascia is usually observed within a few months, but one case of "MRI remission" was reported after 30 months [13]. In our patient, remission was observed earlier by MRI.

In conclusion, MRI may be considered a very valuable tool in the diagnosis of EF and in assessing remission status in the absence of inflammation of fascia. It also helps clinicians adapt their therapeutic strategy in order to reduce corticosteroid dose. Only a prospective study can determine: (1) the sensitivity and specificity of MRI patterns in differentiating disorders such as scleroderma, graft versus host disease, and eosinophilia–myalgia syndrome and (2) the sensitivity of change of MRI when assessing early therapeutic response in these diseases.

Disclosures None

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