AN AMYOTROPHIC LATERAL SCLEROSIS MIMICKER

Eosinophilic fasciitis

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Eosinophilic fasciitis (EF) is a rare localized scleroderma-like disorder[1]. Until today both the etiology and pathomechanism remain elusive. In general, the skin changes are symmetric and include an initial edema and erythema of the extremities, followed by 'peau d'orange' and woody skin induration in a later phase. The skin induration can lead to joint contractures with limited mobility. Laboratory tests may show hypergammaglobulinemia, peripheral eosinophilia and an elevated erythrocyte sedimentation rate[2]. Clinical pictures of lower motor neuron (LMN) disease associated to EF have already been described[3,4]. The classical features of amyotrophic lateral sclerosis (ALS) are a combination of upper motor neuron (UMN) and LMN signs on examination and/or electromyography (EMG) without sensory, extraocular muscle or sphincter involvement[1].

We present an unusual report of a patient with EF and clinical and EMG characteristics of a lower neuron disease suggestive of motor neuron disease (MND), such as ALS. We also discuss the role of clinical signs in differential diagnosis of MND and some ALS mimickers.

CASE

A 35-year-old right-handed white man presented a 3-month history of fasciculations and loss of strength in right wrist. His medical record was unremarkble, except for a chronic complain of dry eyes, which did not interfere in his activities. He was an amateur sportist and used to play tennis regularly. Three months before his first evaluation he noticed that he could not extend his right wrist properly associated with an intermitent complaint of rigidity of his right arm. One month later, he started to present difficult for walking due to rigidity in his right leg.

His general clinical examination was unremarkable, excpet for bilateral swollen knees. On neurological examination, he did not present any cognitive abnormality. His strength was normal, except for discrete limitation for flexion and extension of his right wrist. Deep tendon reflexes were normal, but we observed spontaneous fasciculations in both arms and legs. Coordination and sensitivity were preserved as well as cranial nerve functions.

An EMG, including nerve conduction studies and needle EMG, was preformed. Motor nerve conduction studies yielded normal conduction velocities and distal motor latencies with low amplitude at the compound motor action potential. The results of sensory-nerve conduction studies were normal. Needle EMG study showed spontaneous activity (fibrilations and positive sharp waves), abundant fasciculations and changes in the configuration of motor unit potentials that were of increased interference patern indicative of a net motor unit loss bilaterally, in myotomos C5, C6, C7, L5 and S1. There were also signs of denervation in rest in bilateral paravertebral lumbosacral muscles. These findings were consistent with a disorder of the LMN, their axons, as seen in ALS.

A complete blood count was normal, except for mild eosynophilia (760 eosynophilies, 9% of total leucocytes). There was no evidence of parasitic or other recognized cause of eosinophilia. Creatine kinase was 269 UI/mL and aldolase, 3.7 UI/mL. Levels of serum sodium, potassium, calcium, magnesium, phosphorus, vitamin B12, folate, glucose, thyroid-stimulating hormon and creatine were normal, as were the results of liver function tests. FAN was positive 1:640, but other reumathologic tests, including, anti-Ro and anti-La were normal. He was tested for HIV, syphilis, hepatitis B and C, and the results were negative. Hypergammaglobulinemia was detected in serum analysis.

Magnetic resonance imaging (MRI) of the right knee disclosed edema of soft adjacent parts to the muscular bellies of the evaluated segment, of circunferencial aspect, and there was intense contrast enhacement. These characteristics were suggestive of inflammatory involvement of soft tissue (Fig 1A).

The patient was submitted to muscle and skin biopsies . These biopsies disclosed thickned fascia, extending from the subcutaneous tissue to muscle. This striking increase in collagenous conective tissue was accompained by an intense infiltra-

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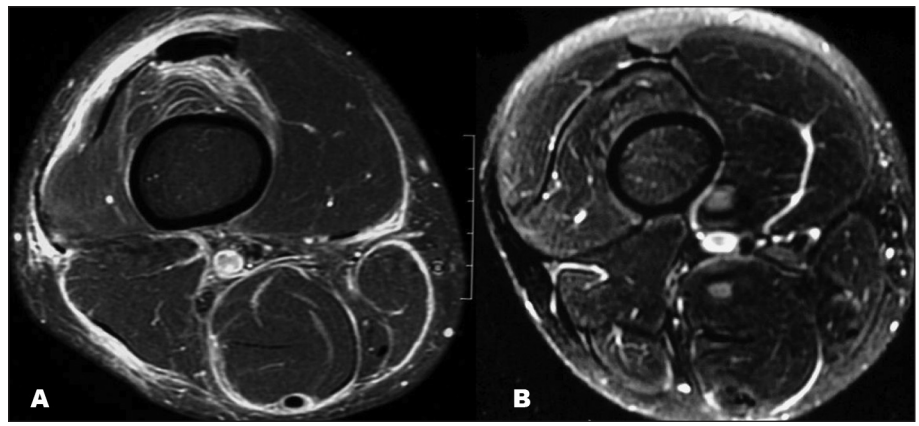

Fig 1. (A) MRI of the right knee showing edema of soft adjacent parts to the muscular bellies of the evaluated segment, of circunferencial aspect, and there was intense contrast enhacement. (B) Control MRI of his right knee was normal

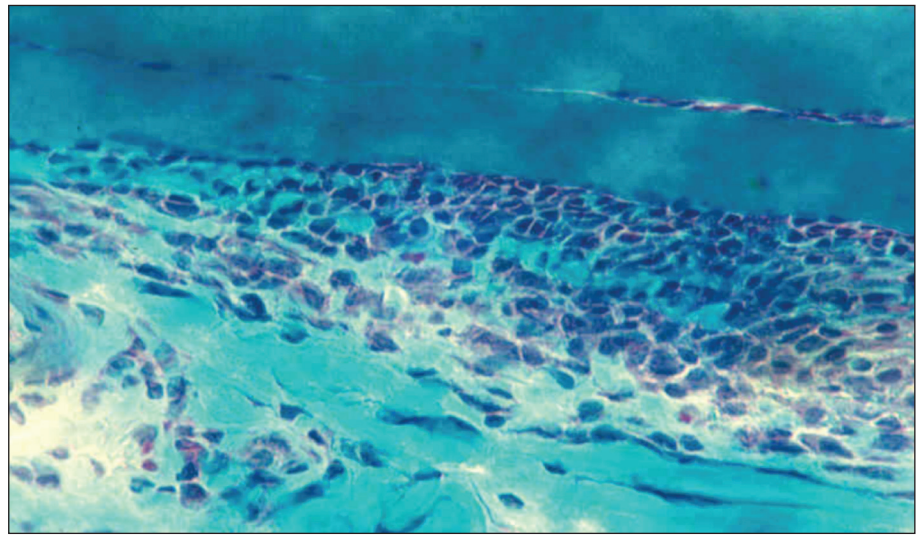

Fig 2. Muscle and fascia biopsies disclosing thickned fascia, extending from the subcutaneous tissue to muscle, accompained by an intense infiltration with eosinophilia.

tion of diffuse fasciitis with eosinophilia. Muscle fascile is imaged (Fig 2).

He was diagnosed with EF and treated with 60 mg prednisone for one month, presenting complete resolution of his complains. Control MRI of his right knee, six month after treatment, was normal (Fig 1B).

DISCUSSION

EF has been associated to neuropathies, but not with MND[3,4]. One of the clinical signs in our patient that pointed out the possibility of MND was fasciculation. It is observed that spontaneous fasciculations may be noted by 70% of normal healthy individulas[5]. Alhough, in retrospect, the first symptom in many patients with ALS may be cramps and fasciculations, it is exceptionally rare for patients later diagnosed with ALS to present on the time of diagnosis with fasciculation alone[6]. However, another report[7] observed that twenty-one of 312 (6.7%) patients who fulfilled the criteria of typical ALS, had fasciculation as the first and only symptom at the time of diagnosis. The mean time interval between developing fasciculation and other defictis was 7.2 months.

In our report we present a 35-year-old man with early-onset generalized fasciculations and an EMG compatible with MND. However, he also presented an unusual clinical symptom which is swollen knees. The presence of unusual symptoms in patients suspected to have ALS demands further investigation, especially if the patient presents the following 'red flags': age less than 40, history of early bowel/bladder involvement and possible exposure to toxic substances[8].

The findings of our subsidiary investigation disclosed

peripheral eosinophilia and histological evidence on the muscle/fascia of EF, mimicking a MND. Two mechanisms of neural damage induced by eosinophils have been proposed. One is the direct neurotoxic effect by eosinophil products, such as MBP, eosinophil-derived neurotoxin, and eosinophilic cationic protein. The other is the neural damage related to vasculitic involvement of the *vasa nervorum*[9]. The neurogenis findings on EMG study in our patient could be related to inflammatory involvement of muscle fiber mainly in nerve terminals. After administration of prednisone, fasciculations were completed resolved, showing that the clinical signs were directly caused by EF.

This case illustrates that different diseases may mimic ALS, and that even if the EMG confirms the combination of active denervation and reeinervation, which is the hallmark of ALS, we must search for an alternative diagnosis when the clinical signs are atypical.

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