



Letters to the Editor

Central Nervous System Involvement on Eosinophilic Fasciitis*

Afectación del sistema nervioso central y periférico en la fasciitis eosinofílica

Dear Editor,

Eosinophilic fasciitis (EF) is a rare sclerodermiform syndrome of unknown cause described by Shulman in 1974.¹ It features²: induration, peripheral eosinophilia, increased erythrocyte sedimentation rate (ESR) and hypergammaglobulinemia. Various systemic manifestations associated with EF have been described, such as renal, cardiac, respiratory, joint and peripheral nervous system manifestations. Below we present the case of a patient with EF of the central and peripheral nervous system.

The patient was a 71-year-old male with a medical history of hypertension and tuberculosis. He came to the hospital with induration of the arms, legs, chest and neck, which had started 5 months prior. He referred dysphagia to solids and loss of 10 kg in the past year. Physical examination revealed skin induration in the arms, legs, trunk and neck, respecting the hands and feet. He had a positive sulcus sign and orange skin. Neurological examination was normal. Laboratory tests showed peripheral eosinophilia ($8.1 \times 10^9/l$ leukocytes with 2% eosinophils) and an ESR of 40 mm in the first hour. Blood biochemistry was normal. Protein, tumor markers, chest X-ray, I capillaroscopy and immunological tests were normal. The upper GI series revealed esophageal motor incoordination with passage of contrast between the larynx and esophagus. An MRI showed changes consistent with eosinophilic fasciitis. A muscle biopsy was performed which revealed deep and reactive inflammatory changes in adipose tissue and fascia, all compatible with EF. Glucocorticoid therapy was initiated at doses of 1 mg/kg/day. At 2 months he showed a left facial central paralysis and hypoglossal nerve palsy. Cranial MRI was normal. A lumbar puncture was performed which was also normal. Subsequently, paralysis of the external popliteal nerve was observed. Physical examination showed persistent induration of the arms, legs and trunk. The neurological examination revealed a central left facial nerve palsy, left hypoglossal nerve palsy, paresis of the external cyatic popliteal nerve, but sensitivity and tendon reflexes were present and symmetrical as was the bilateral flexor plantar cutaneous reflex. The blood count was normal and ESR was 22 mm in the first hour. Blood biochemistry showed mild hypoproteinemia. Tumor markers and a CT scan were normal. An electromyogram was performed which revealed an asymmetric

non sensitive polyneuropathy. Treatment was begun with azathioprine 50 mg/day and glucocorticoids at a dose of 1 mg/kg/day. Six months later azathioprine was withdrawn. Today, 10 years after diagnosis, he remains in treatment with 2 mg of methylprednisolone, without neurological or skin complications.

Since the first description of the syndrome, over 250 cases of eosinophilic fasciitis have been published. The most frequent neurological manifestation of peripheral neuropathy is due to carpal tunnel syndrome (both unilateral and bilateral), which occurs in approximately 25% of cases.^{3,4} Lower limb neuropathy secondary to^{5,6} mononeuritis multiplex and multifocal peripheral neuropathy have also been described.⁷ In the case reported, involvement was seen simultaneously in the central and peripheral nervous system: central left facial paralysis, left hypoglossal nerve palsy and paresis of the cyatic popliteal nerve, with good response to corticosteroids and azathioprine. In a literature review using PubMed (descriptors: eosinophilic fasciitis & nervous system; 1974–2012) we have not found more cases of eosinophilic fasciitis associated with this type of neurological involvement. It may be recalled that eosinophilic fasciitis is a disease with many extracutaneous manifestations, including the nervous system.

References

- Shulman LE. Diffuse fasciitis with eosinophilia: a new syndrome? *Trans Assoc Am Phys.* 1975;88:70–86.
- Trallero Araguas E, Selva O'Callaghan A, Simeón Aznar C, Sanjurjo Golpe E, García-Patos V, Vilardell Tarrés M. Fasciitis eosinofílica: estudio de 10 pacientes. *Med Clin.* 2005;125:145–8.
- Lakhanpal S, Ginsburg WW, Michet CJ, Doyle JA, Moore SB. Eosinophilic fasciitis: clinical spectrum and therapeutic response in 52 cases. *Semin Arthritis Rheumat.* 1988;17:221–31.
- Jones Jr HR, Beetham Jr WP, Silverman ML, Margles SW. Eosinophilic fasciitis and the carpal tunnel syndrome. *J Neurol Neurosurg Psychiatry.* 1986;49:324–7.
- Moriguchi M, Terai C, Kuroki S, Tanaka E, Someya N, Tsunoda Y, et al. Eosinophilic fasciitis complicated with peripheral polyneuropathy. *Intern Med.* 1998;37:417–20.
- Pardal-Fernández JM, Sáez-Méndez L, Rodríguez-Vázquez M, Godes-Medrano B, Iñiguez-De Onzoño L. Multiple mononeuritis and eosinophilic fasciitis in a patient with idiopathic hyper-eosinophilic syndrome. *Rev Neurol.* 2012;54:100–4.
- Satsangi J, Donaghy M. Multifocal peripheral neuropathy in eosinophilic fasciitis. *J Neurol.* 1992;239:91–2.

Dolors Grados,* Melania Martínez-Morillo, Pilar Latorre, Alejandro Olivé

Sección de Reumatología, Servicio de Neurología, Hospital Germans Trias i Pujol, Barcelona, Spain

* Corresponding author.

E-mail address: dgrados23@hotmail.com (D. Grados).

* Please, cite this article as: Grados D, et al. Afectación del sistema nervioso central y periférico en la fasciitis eosinofílica. *Reumatol Clin.* 2013. <http://dx.doi.org/10.1016/j.reuma.2012.09.004>.