Central Nervous System Involvement on Eosinophilic Fascitis

Afectación del sistema nervioso central y periférico en la fascitis eosinofílica

Dear Editor,

Eosinophilic fascitis (EF) is a rare sclerodermiform syndrome of unknown cause described by Shulman in 1974. It features: 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 × 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, 1 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 fascitis. 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 dosages 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, and ophthalmoplegic, 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 hypoprotenemia. 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 fascitis 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. Lower limb neuropathy secondary to mononeuritis multiplex and multifocal peripheral neuropathy have also been described. 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 right popliteal nerve, with good response to corticosteroids and azathioprine. In a literature review using PubMed (descriptors: eosinophilic fascitis & nervous system; 1974–2012) we have not found more cases of eosinophilic fascitis associated with this type of neurological involvement. It may be recalled that eosinophilic fascitis is a disease with many extracutaneous manifestations, including the nervous system.

References


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