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EOSINOPHILIC FASCIITIS AS DIFFERENTIAL DIAGNOSIS OF SYSTEMIC SCLEROSIS

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BACKGROUND

Eosinophilic fasciitis (EF) is a rare disorder and its etiology is still obscure. It seems to occur equally in both sexes, in individuals between 40-50 years of age. Main involviment is cutaneous and, usually, it is sequential: symetricall and painful infiltration of extremities; oedema and peau d'orange appearance of the skin; cutaneous thickening and hypopigmentation. The groove sign is a characteristic feature. Visceral findings are rare, generally including renal and hematological affections. It is mandatory to differentieate EF from systemic sclerosis (SSc). Unlike EF, in SSc fascia is spared. On the other hand, in EF microstomia, Raynaud phenomenon, sclerodactyly and telangiectasia are absent. There are no globally accepted diagnostic criterea for ES, making it difficult to establish differencial diagnoses. Laboratory findings include high levels of Erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and IgE, eosinophilia (not obligatory) and hypergammaglobulinemia. Eventually, Rheumatoid factor (RF) and antinuclear antibodies (ANA) may be present, whereas Antineutrophil cytoplasmic antibodies (ANCA), anti-DNA and Extractable Nuclear Antigens (ENA) are negative. Magnetic Resonance Imaging (MRI) has a important role in ES, revealing characterist conditions (such as thickening of the fascia), guiding the biopsy and assisting in monitoring the disease. Fascial biopsy remains the gold standard for the diagnosis of EF.

CASE REPORT

A 50-year-old woman presented with difuse pain, swelling and thickning of the skin in the past five months. She also complained about weigh loss, fever, difficulty in mobilizing and pruritus. Physical examination revealed the groove sign, peau d'orange appearance of the skin and indurated edema. Raynaud phenomenon, sclerodactyly and telangiectasia were not found. Laboratory results showed hypereosinophilia (3276/mm³ - 36%), ESR and CRP were elevated, ANA was positive in low titer (1:80). No marker of systemic esclerosis was found (anticentromere and anti-Scl70 were negative). Capillaroscopy was normal. Imaging exams were inconclusive (there was no fascia abnormalities in MRI). A biopsy of the right leg was performd revealing moderate active chronic fasciitis. The diagnosis of eosinophilic fasciitis was established. Due to the agressive behavior of the disease, methylprednisolone pulse therapy was initiated, followed by ciclophosphamide and prednisone in high doses (0,5mg/kg), with no clinical benefit until this moment.

CONCLUSION

The differential diagnosis between EF and SSc is a challenge, often requiring invasive procedures, such as facial biopsy. It is primordial, however, to establish the correct diagnosis, once the treatment of these two entities Is different.