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ORBITAL INFLAMMATORY PSEUDOTUMOR ASSOCIATED WITH SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) IN THE ELDERLY: A CASE REPORT

Viviane Alves Costa^{1,*}, Iane Tamara Dondé¹, Gizelle Gouveia Rezende¹, Viviane Queiroz de Oliveira Maia¹, Danielly Dantas Pimentel¹, Luciana Akita¹, Gustavo Roberto Lourenço¹, Juliana de Jesus Boscolo¹, Taísa Morete da Silva¹, Roberto Acayaba de Toledo¹

1. Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto (SP), Brazil.

*Corresponding author: vivicosta_med@hotmail.com

BACKGROUND

Systemic lupus erythematosus (SLE) is a chronic, autoimmune, multisystem disease which may affect the eyes and/or visual system in up to one-third of patients. Any structure of the eye might be involved in SLE, with keratoconjunctivitis sicca, retinal vasculopathy and episcleritis the most common manifestation. Rare ocular presentations including orbital inflammatory pseudotumor, orbital myositis, panniculitis, acute orbital ischemia and infarction. The patient consents for publication images.

CASE REPORT

Female, 58 years old, mulatta, with a history of hypertension and smoking, was diagnosed with Sweet's syndrome in November 2019, when she started to have skin rashes and histopathological compatible biopsy, weight loss, fever, lymph node enlargement, myalgia, arthralgia in wrists and metacarpophalangeal joints. Hematological cancer was ruled out and sent to our service in June 2020 for collagen investigation, with 20 mg/day of prednisone. Laboratory tests revealed antinuclear antibody (ANA) 1:320 nuclear homogeneous pattern, low complement (C3 and C4), positive anti-DNA, anti-Ro and anti-La. All the others autoantibodies were negative. Considering Sweet's syndrome and the immunological findings, we concluded that it was SLE. After 2 months, the patient presented severe left retro-orbital pain, evolving with edema and eyelid hyperemia, conjunctival hyperemia, a lot of chemosis, pain on palpation and weakness of the external eye muscles (Fig. 1). Magnetic resonance imaging (MRI) revealed a diffuse edema of the left periocular soft tissue, with no signs of mass, without the involvement of the nasal, frontal and maxillary sinuses. Serum concentrations of IgG4 were normal. After excluding infectious causes, the hypothesis of orbital pseudotumor was raised. We managed dexamethasone eye drops and intravenous in immunosuppressive dose, and started using methotrexate to help with the reduction of steroid. The patient rapidly gave us a good response, improving periorbital edema and muscle weakness (Fig. 2). We outlined an interesting case of SLE in an elderly with unusual clinical manifestations, who started the medical conditions with Sweet's syndrome and orbital pseudotumor.

Figure 1. Important edema and difficulty to open left eye (admission).

Figure 2. After three days with dexamethasone.

CONCLUSION

Eye manifestations in SLE may be sight-threatening and might be an indicator of active systemic disease. Our case is remarkable due to ocular inflammation being the only active manifestation of SLE without any signs of active joint, skin, brain, or kidney involvement. There are many case reports as well as references of therapeutic agents being used in orbital inflammatory pseudotumor, including corticosteroids, radiotherapy, cyclosporine-A, methotrexate and infliximab.