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SJOGREN SYNDROME ASSOCIATED WITH ORBITAL PSEUDOTUMOR: A CASE REPORT

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BACKGROUND

Sjögren Syndrome (SS) is a systemic autoimmune disease whose pathophysiology concerns about a lymphocytic infiltrative process in lacrimal and salivary glands, destroying those structures, leading to clinical manifestations like xerostomia and xerophtalmia. Other clinical manifestations may be present, like arthralgia, myalgia, peripheral neuropathy, pulmonary, thryoid, kidney and hematological disorders.

Orbital Pseudotumor is an inflammatory benign process that affects the orbital region, commonly one-sided. There are three clinical different manifestations: acute, subacute and chronic. Symptoms are pain, proptosis and local phlogistic signs, but also can occur optic neuropathy and motility dysfunction. In rheumatology scenario, Orbital Pseudotumor usually is associated with granulomatosis with polyangiitis and rarely with other conditions.

CASE REPORT

25 years old female pacient previously diagnosed with hypothroydism and autoimmune hemolytic anemia, reports symptoms as xerostomia, xerophtalmia, fatigue and generalized pain with two years of evolution. The clinical condition evolved with right eye pain and proptosis and the patient was submited to a magnetic ressonance imaging, revealing findings compatible with orbital pseudotumor.

Retro-ocular biopsy results were negative for malignancy. Besides, Schirmer test was positive. Blood tests detected ANA 1/640 nuclear fine speckled pattern. Rheumatoid factor positive, Anti- Ro 89 U/mL, CRP 20,4 mg/dl, without anemia nor hemolysis signs. ANCA was negative. There were no sinus or pulmonary disease. Salivary gland biopsy revealed lymphocytic infiltrate – Focus score >1. Inicial therapy consisted in methylprednisolone pulse therapy, followed by azatioprine, prednisone and hydroxychloroquine. There was a decrease of the inflammatory process seen at magnetic ressonance imaging and improvement on the patient's clinical condition with the treatment.

CONCLUSION

SS and Orbital Pseudotumor are rare conditions and there is no published data reporting this association. Furthermore, the established treatment was effective to control disease activity. Another therapy option if new disease activity occurs could be rituximab, commonly used in serious SS manifestations.