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RETRO-ORBITAL AND TEMPORAL MASS SECONDARY TO IMMUNOGLOBULIN G4-RELATED DISEASE

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BACKGROUND

Immunoglobulin G4-related disease is a fibroinflammatory disorder of unknown etiology characterized by a mass-forming lesion that may involve one or more organs. The diagnostic criteria include clinical manifestations, elevated serum levels of immunoglobulin G4 (IgG4), and histopathologic findings of a dense lymphoplasmacytic infiltrate and infiltration of IgG4-positive plasma cells upon immunohistochemical examination.

CASE REPORT

In 2015, a 63-year-old female patient was hospitalized to investigate recent onset of isolated left ptosis. The remainder neurological examination was within normal limits. Magnetic resonance image (MRI) of skull and orbits revealed an expansive lesion measuring 2x1.6 cm in the orbital cavity between the orbit roof and upper rectus muscle, with intense impregnation by the gadolinium, and a lesion measuring 4x1 cm in the right superficial temporal region. A PET Scan indicated focal metabolic increases near the upper portion of the left orbit and in the mediastinal and hilar lymph nodes bilaterally. Lymph node biopsy by mediastinoscopy presented no evidence of lymphoma or sarcoidosis; bone marrow biopsy was normal. At this moment an orbital biopsy was performed, revealing a lymphocytic infiltrate with increased IgG4 deposit. Because of retro-orbital biopsy results, as well as an elevated serum IgG4 (1630, reference value 700-1600), the patient was discharged with a high suspicion of IgG4-related disease, being treated initially with prednisone and azatiprone, the last one suspended because of elevation of transaminases, maintaining only with corticosteroids until january 2018, when the treatment was stopped. In december 2018 she noticed a lesion in the right temporal area, which remarkably improved in january after using prednisone 40 mg per day. Then, it was gradually reduced the dose. The patient was re-admitted to the hospital in march 2019, using prednisone 5mg/day, and had no lesion growth or new noticeable lesions after starting the medication. A new brain and orbit MRI indicated expansive soft tissue lesion in the right temporal region (approximately 5.3 x 5.2 x 1.4 cm in diameter), possibly related to IgG4 disease. Since the risk of facial nerve involvement hindered an excisional biopsy, we decided to treat for IgG4-related disease with rituximab. The result of this treatment was excellent and the patient presented complete regression of the expansive lesion in the temporal territory.

CONCLUSION

The growing recognition of IgG4-related systemic disease highlights the importance of considering this diagnosis in patients with multivisceral lesions. It makes possible treatment directed to each case, including use of rituximab, with recent evidence supporting its use.