records, the major goals set at the beginning of the treatment were successfully achieved, providing the patient with adequate masticatory function and pleasant facial aesthetics.

IGG4-Related orbit mass: a diagnostic challenge

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Aim: The aim of the present study was to report a case of an orbital mass IgG4-related. IgG4-related disease (IgG4-RD) is a newly described systemic autoimmune disease, characterized by infiltration of IgG4+ plasma cells accompanied by tissue fibrosis of multiple organs throughout the body and swelling or tumor-like, nodular or proliferative lesions. It was initially described as affecting the pancreas, but commonly involves the head and neck region as well.
Case report: A 75-year-old female in good health came to our hospital on October 2019 complaining intermittent diplopia since August 2019. Clinical examination revealed the presence of a left orbital subcutaneous neoformation. Subsequent CT with contrast medium showed a neoformation of soft tissue the left eyelid which erodes the medial wall of the left orbit and the nasal process of the frontal bone. The chest radiography excluded pleuroparenchymal lesions. Blood exam highlighted an iron deficiency anemia framework. The clinical suspects were of a lymphoma, Mikulicz disease or Kuttner tumour. In local anesthesia an incisional biopsy of the mass was performed. The histopathological examination was not conclusive,: inflammatory process with the presence of histiocytes, lymphocytes, granulocytes and plasma cells, (CD68+, CKS 100-, SMA-) which cause necrosis of the muscle structure. Serum immunoglobulin levels, including IgG4, were required and a second biopsy was performed in general anesthesia. Serum level of IgG4 were higher >135mg/dL and pathological examination pointed out the presence of an intense intense lymphocytic infiltrate mainly plasma cell, with histiocytes and granulocytes. Immunohistochemistry highlighted the positivity of CD138 (negative: CD3, CD20) and ratio of IgG4/IgG positive cell> 50%. These results allowed a definitive diagnosis of ophthalmic IgG4-related disease as proposed by Umehara et al.(2017): Imaging studies show enlargement of the lacrimal gland, trigeminal nerve, or extraocular muscle as well as masses, enlargement, or hypertrophic lesion in various ophthalmic tissues. Histopathologic examination shows marked lymphocyte and plasmacyte infiltration, and sometimes fibrosis. A germinal centre is frequently observed. IgG4 þ plasmacytes are found and satisfy the following criteria: ratio of IgG4 þ cells to IgG þ cells of 40% or above, or more than 50 IgG4 þ cells per high-power field (400). Blood test shows elevated serum IgG4 (>135 mg/dl). Diagnosis is classified as 'definitive' when (1), (2) and (3) are satisfied; 'probable' when (1) and (2) are satisfied; and 'possible' when (1) and (3) are satisfied. Screening of the neck, chest, abdomen and pelvis with computed tomography did not reveal any other organ involvement. The patient underwent rheumatologic examination, and subsequent oral Prednisolone and Azathioprine treatment with resolution of the proptosis. There was also improvement of the eye pain. A CT was performed after 1 month and a reduction of the mass was highlighted.
Conclusion: In the head and neck region, IgG4 disease has been described to cause enlargement of the extraocular muscles, pituitary stalk, retrobulbar soft tissues, salivary glands, lacrimal glands, and cranial nerve branches. IgG4-RD commonly involves the maxillary and mandibular branches of the trigeminal nerve, with preferential involvement of the infraorbital nerves. The common differential diagnosis on radiology includes lymphoma, granulomatosis with polyangiitis, Sarcoidosis and Grave's orbitopathy. The degree of fibrosis in the affected organ is a major determinant of treatment response. Prognostically, orbital IgG4-RD usually shows dramatic response with steroids but relapses are common and hence long term low dose steroid or combination with immunosuppressant may be necessary.