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Case report

IgG4-related disease of the infratemporal fossa: A case report☆

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ABSTRACT

IgG4-related disease is a relatively new disease entity characterized by elevated serum IgG4 levels and marked infiltration of IgG4-positive plasma cells in mass lesions. IgG4-related disease of the head and neck region is often associated with the salivary glands, and Küttner's tumor and Mikulicz's disease of the salivary glands may also be IgG4-related diseases. We encountered a patient with an inflammatory pseudotumor (IPT) of the infratemporal fossa, which was histopathologically confirmed to be IgG4-related disease. The patient was a 53-year-old man who presented with tenderness of the right upper jaw, ocular pain, and trismus for approximately 3 months. Imaging revealed a lesion with unclear borders extending from the infratemporal fossa into the right orbit. Soft tissue tumor was suspected and biopsy was performed under general anesthesia. Histopathological examination revealed plasma cell infiltration and fibrosis. Immunohistochemistry revealed prominence of IgG4-positive plasma cells and confirmed the diagnosis of IgG4-related disease. The patient responded well to steroidal therapy, with pain and trismus resolving. Imaging revealed marked reduction in the size of the mass. The patient is currently under observation on low-dose oral prednisolone with no evidence of relapse.

1. Introduction

Although the causes of chronic inflammatory diseases such as autoimmune pancreatitis (AIP) and Mikulicz's disease have been unclear, recent studies have revealed that they are related to IgG4 [1]. Immunoglobulin (Ig) G4-related disease is a newly proposed disease concept according to which these diseases may be caused by the same mechanism. IgG4-related disease is a novel lymphoproliferative disorder that shows hyper-IgG4-γ-globulinemia and IgG4-producing plasma cell expansion in affected organs with fibrotic or sclerotic changes. Patients show systemic inflammatory conditions and various symptoms depending on the affected organ. IgG4-related disease was initially reported in relation to the pancreas, it is now considered to involve the development of fibrotic lesions in multiple organs and tissues, such as the bile ducts, lungs, salivary glands, lacrimal glands, kidneys, prostate, and retroperitoneum. However, the etiology is still unknown.

IgG4-related disease is characterized by elevation of the serum IgG4 level on routine blood tests. IgG4-related disease generally responds well to low-dose steroid therapy, though some patients may be misdiagnosed as having malignant tumors and undergo surgical treatment. Therefore, making a correct diagnosis of this condition is critical [2]. So, we present a rare case of IgG4-related disease in the infratemporal fossa.

2. Case report

On October, 2010, a 53-year-old Japanese man presented to the Department of Oral and Maxillofacial Surgery at Tokai University Hospital, with tenderness of the right upper jaw, ocular pain, and trismus. His symptoms had first appeared in early September. The patient had no past history or family history of pancreatitis and no history of allergy. He had previously been in good health. Physical examination revealed tenderness of the right upper jaw and right eye, as well as trismus. Maximal mouth opening was approximately 20 mm. The cervical lymph nodes were normal. Intraoral examination revealed no swelling or gingival enlargement. Routine blood tests were normal. The total IgG level was 1248 mg/dl (normal < 1695 mg/dl), and the serum levels of IgG subclasses were as follows: 604 mg/dl for IgG1 (48.37% of total IgG, normal <65%), 485 mg/dl for IgG2 (38.84% of total IgG, normal <23%), 35.6 mg/dl for IgG3 (2.85% of total IgG, normal <8%), and 124 mg/dl for IgG4 (9.93% of total IgG, normal <4%). Computed tomography (CT) revealed

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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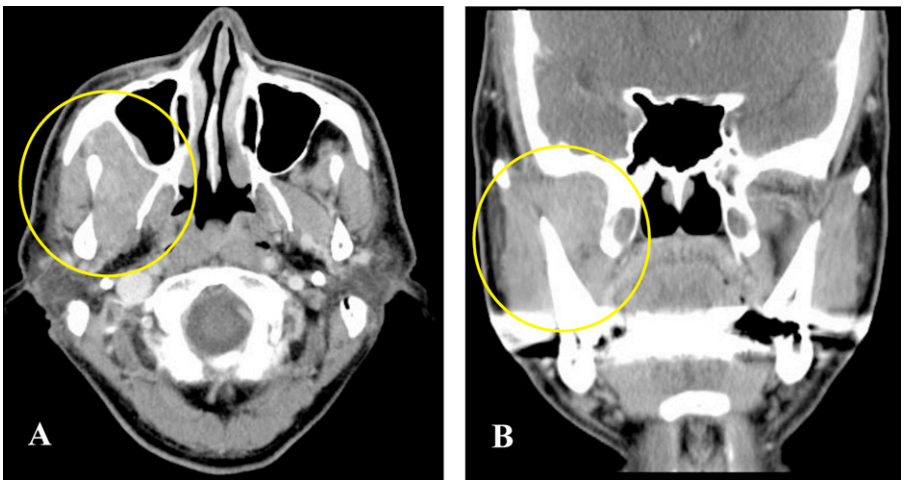

Fig. 1. Axial and coronal computed tomography shows a homogeneous enhancing mass in the right infratemporal fossa.

a slightly enhanced soft tissue mass in the right infratemporal fossa that extended into the posterior part of the right orbital cavity. The external pterygoid muscle and temporalis muscle were markedly enlarged. CT revealed no bone destruction around the tumor (Fig. 1A and B). Magnetic resonance imaging (MRI) confirmed an enhancing mass in the right infratemporal fossa that extended into the orbital region. The lesion was hypointense to fat intensity on T1-weighted images and was isointense with muscle, while it

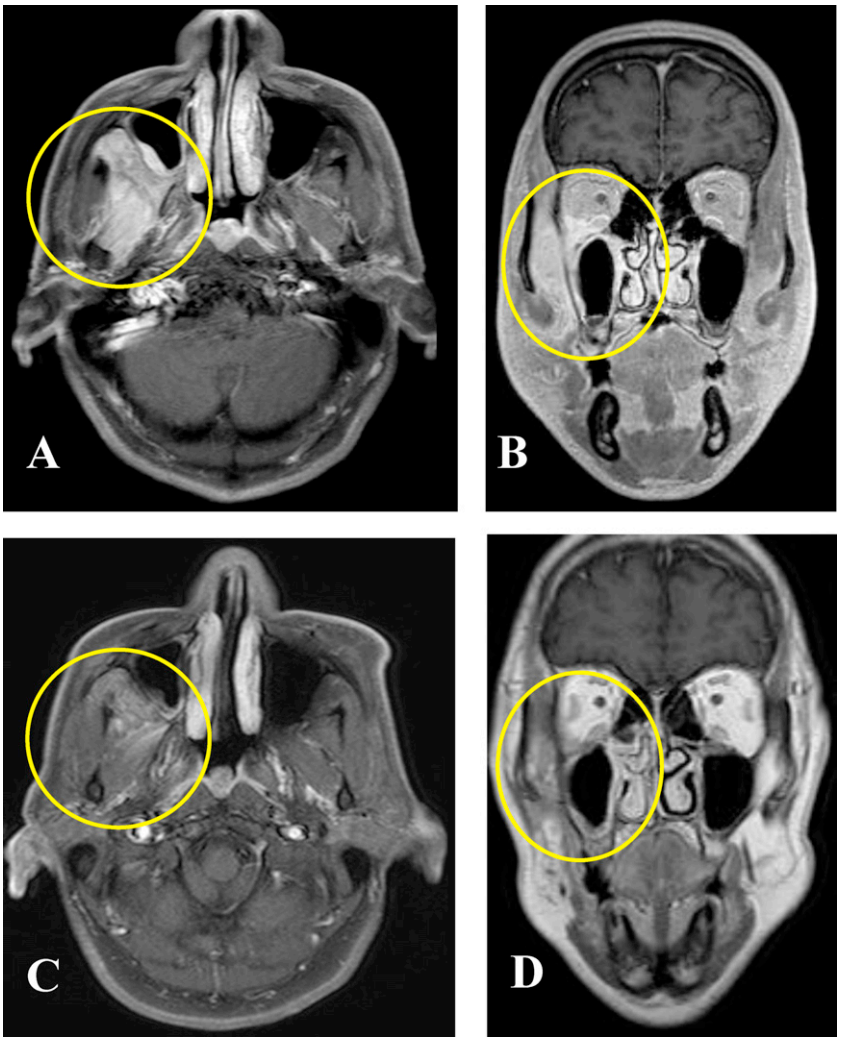

Fig. 2. Magnetic resonance imaging (T1-weighted). (A and B) Before treatment. There is an enhanced mass in the right infratemporal fossa extending into the orbital region. (C and D) After 3 weeks of steroid therapy, the lesion of the right infratemporal fossa has almost disappeared.