A Case of IgG4-Related Disease of the Submandibular Gland

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IgG4-Related disease (IgG4-RD) is a chronic systemic fibroinflammatory disease characterized by dense lymphoplasmacytic infiltrate rich in IgG4-positive plasma cells and fibrosis. IgG4-RD has been described in many organ systems. IgG4-RD is a disease that can affect multiple organs, mainly affecting the digestive system together. Invasion of the head and neck, especially the submandibular gland, is not so much worldwide. In Korea, there are few reports of IgG4-RD sialadenitis.

Case: A 73-year-old man visited the hospital with a fixed, hard, painful Rt. submandibular mass of 2 cm in size that had been palpated since 1 month ago. A hypoechoic mass of 1x1x2 cm was observed on ultrasound (Fig. 1). Excisional biospy was performed and biopsy specimens of the submandibular glands of the patient showed well-preserved lobular architecture with prominent interlobular fibrosis (Fig. 2a). Active lymphoplasmacytic infiltrates in the submandibular gland were observed pathologically (Fig. 2b). In addition, IgG and IgG4 immunostaining showed lymphocytic infiltration and an IgG4/IgG ratio greater than 50% in the patient (Fig. 2c,d). This histologic findings and immunohistochemical profile are consistent with the diagnosis of IgG4-related sialadenitis, and no evidence for lymphoma was found. In laboratory test results, increased serum IgG4 level (372.0 mg/dL), WBC (11,300/mm^3), and moderately elevated AST (42 IU/L), IgG(1499.3 mg/dL) were reported. All other labs, ALT (40 IU/L), Hb (13.6g/dL), RBC (4.30x10^6/mm^3), and PLT (213,000/uL) were within the normal range. Computed tomography (CT) of the neck with contrast (Fig.3) showed subtle heterogeneous parenchymal attenuation and enhancement of remnant right submandibular, left submandibular gland. CT of the abdomen showed mild dilation of EHD and IHD with suspicious focal luminal narrowing at distal CBD, which may indicate IgG4-RD involvement(Fig. 4). The patient started treatment with 40 mg prednisolone daily, followed by tapering of prednisone, currently maintaining 5 mg. Currently, 7 months have elapsed, and the IgG4 level has decreased to normal range (Fig. 5), and there are no remarkable changes on f/u abdominal CT.

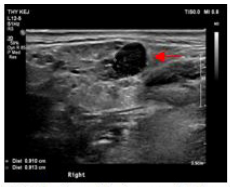

Fig.1 Rt. submandibular mass: 1x1x2cm, hypoechoic (red arrow)

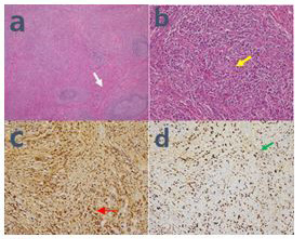

Fig.2
a: (lowpower field) Storiform fibrosis (white arrow)
b: (highpower field) Obliterative phlebitis (yellow arrow).
c: Positive Immunohistochemistry Staining for IgG (red arrow)
d: Positive Immunohistochemistry Staining for IgG4 (green arrow)

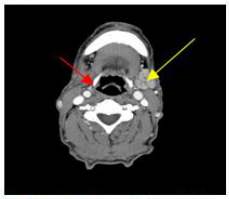

Fig.3 Neck CT, 1 year after Excision. Rt. submandibular mass, removed (red arrow). Subtle heterogeneous attenuation in Lt. submandibular gland (yellow arrow)

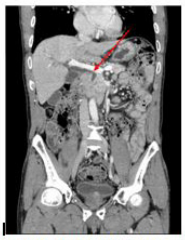

Fig.4 Bile duct stricture (red arrow)

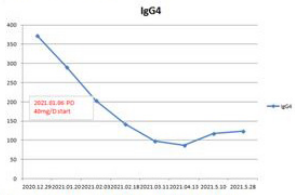

Fig.5 a decrease in IgG4 levels (2021.1.6 PD 40mg/D start)