Two Cases of IgG4-related Mikulicz’s Disease of the Submandibular Glands with Progressive Transformation of the Germinal Centers

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Two patients of IgG4-related Mikulicz’s disease (IgG4-RD) with progressive transformation of germinal centers (PTGC) arising in the submandibular glands are reported. The patients were a 58-year-old male and 39-year-old female who fulfilled the clinico-pathological diagnostic criteria for IgG4-RD, including ① markedly elevated serum IgG4 level, ② bilateral swelling of the submandibular and lacrimal glands, and ③ microscopic finding of an IgG4+/IgG+ plasma cell ratio of more than 40% in the germinal centers.

Histopathologically, PTGCs are characterized by hyperplastic germinal centers composed of mantle zone lymphocytes and remnant of large germinal center cells with follicular lysis. Complete resection is the only treatment for PTGC. However, in patients with IgG4-RD, administration of a corticosteroid might be recommended and efficacious. Because of the possibility of recurrence or malignant transformation after surgery or other treatments, close follow-up is mandatory.

To the best of our knowledge based on a thorough search of the literature, there is only one report other than ours of PTGC arising in the submandibular glands in a patient with IgG4-RD.

Keywords : IgG4-related Mikulicz’s disease, submandibular gland, progressive transformation of germinal center

References

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(Case 1) Findings at the first visit
a. Ultrasonography reveals swelling of the left submandibular gland with a hypoechoic lesion inside. The submandibular gland was partially multilocular.
b. Coronal-section enhanced computed tomography finding reveals bilateral swellings of the submandibular glands (arrows), with small low density areas inside.
c. Horizontal-section enhanced computed tomography finding reveals bilateral swellings of the lacrimal glands (arrows).

(Case 1) Histopathological findings of the submandibular gland
a. (HE-staining, ×40) Many germinal centers exist, which show irregular structures and marked swellings.
b. (HE-staining, ×100) Atrophic salivary tissues are seen with lymphocyte and plasma cell invasion.

(Case 1) Immunohistochemical findings of the submandibular gland
a. (×40) Anti-bcl-2 staining reveals the negative results on the germinal centers and positive ones on the mantle zones (arrow).
b. (×200) Microscopic findings of a IgG4+/ IgG+ plasma cell ratio of more than 70% in the germinal centers.

(Case 2) Histopathological findings of the submandibular gland
a. (HE-staining, ×40) Irregular structures of the germinal centers are found, with follicular lysis (arrow).
b. (HE-staining, ×100) Invasions of lymphocytes and plasma cells into the gland are seen, with storiform fibrosis.