

# Mikulicz's Disease Demonstrating an Enlargement of the Pancreas

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**Abstract :** A 79-year-old female presented with bilateral submandibular and parotid gland swelling and symptoms of oral dryness. Sjogren's syndrome was suspected, but neither anti-SS-A nor anti-SS-B antibodies were positive. The value of serum IgG4 was sufficiently high. Abdominal CT revealed swelling of the pancreas. Immunohistochemical analysis showed abundant IgG4-positive plasma cell infiltration in the biopsied submandibular gland tissue. The patient was finally diagnosed with Mikulicz's disease, and 30mg/day prednisolone was administered. The sizes of the submandibular and parotid glands were reduced immediately. The swelling of the pancreas also decreased. Since autoimmune pancreatitis has been recognized as a distinct entity, a novel clinicopathological IgG4-related systemic disease has been proposed based on the histological and immunohistochemical findings of dense infiltration of IgG4-positive plasma cells with interstitial fibrosis. However, when there is swelling of the salivary glands, it is necessary to consider Mikulicz's disease in the differential diagnosis, and systemic evaluations for complications with other IgG4 diseases is necessary.

**Key words :** Mikulicz's disease, IgG4 related disease, IgG4, Cervical tumor