

DIAGNOSTIC CHALLENGE

Unilateral submandibular swelling

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THE CHALLENGE

A 60-year-old woman sought treatment at the Department of Oral Medicine at Peking University Hospital of Stomatology, Beijing, China. Her main symptom was a painless swelling of the left submandibular area that she had had for 2 months (Figure 1). Other symptoms included dry mouth and difficulty swallowing dry food. She did not complain of any other symptoms, such as fever, fatigue, reduced appetite, or weight loss.

One year before visiting our department at a local hospital, the patient had undergone a partial thyroidectomy. Pathology results revealed a nodular adenomatoid hyperplasia and fibrosis of the thyroid. Shortly afterward, she began to complain of dry eyes and was diagnosed with xerophthalmia at a general hospital. She was not taking any medications and had no history of hypertension, diabetes mellitus, or other systemic disorders.

Physical examination at admission revealed a diffuse, firm swelling of the left submandibular gland and an enlargement in the bilateral sublingual areas, which had a rubbery texture. There was no cervical lymphadenopathy. Her oral mucosa appeared moist, and her unstimulated whole salivary flow rate was measured as 2.0 milliliters for 10 minutes.

Blood test results showed that her complete blood count was normal. Test results of serum proteins showed that the total immunoglobulin G (IgG) level in her serum was 21.73 grams per liter (normal range, 8.00-18.00 g/L), with elevated levels of serum IgG2 (6,460 milligrams per liter; normal range, 1,500-6,400 mg/L), IgG4 (15,700 mg/L; normal range, 80-1,400 mg/L), and total immunoglobulin E (IgE) (322 kilounits per liter; normal range, 0-60 kU/L). The test results were all negative for antinuclear antibodies, anti-double-stranded DNA antibodies, and anti-extractable nuclear antigen antibodies including anti-Sjögren-syndrome-related antigen A (SSA) and anti-Sjögren-syndrome-related antigen B (SSB).

We conducted a sialogram and computed tomography (CT) scan. The sialogram of the left submandibular gland displayed no punctate dilation or obstruction in the gland. The CT scan revealed swelling in the left submandibular gland (Figure 2) and bilateral lacrimal gland. The CT images also showed a round, hyperdense lesion (measuring 2 x 2 square centimeters), which was not apparent clinically, in the left parotid gland.

Incisional biopsy results of the patient's labial glands in the lower lip were nonspecific; however, histopathologic examination of biopsies from the patient's left parotid and submandibular glands showed pronounced inflammation and fibrosis in both. The inflammatory infiltration consisted mainly of lymphocytes and plasmocytes, accompanied by lymph follicle formation with irregular germinal centers. We noted a storiform fibrosis, rich in fibroblasts, mainly in the interlobular connective tissues. We also observed a collagen sheath in the periductal areas (Figure 3). The inflammation and fibrosis resulted in glandular parenchyma atrophy, although the lobular contour was preserved. Immunohistochemical results revealed diffuse infiltration of IgG4-positive plasma cells (Figure 4) with a high ratio (that is, 80%) of IgG4-positive and IgG-positive plasma cells in both the parotid and submandibular glands.