Chronic Painless Parotid Swelling

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A 29-year-old woman with primary Sjögren's syndrome (SS) presented with chronic painless swelling of both parotid glands (Figure 1A). Four years previously, SS was diagnosed based on sicca symptoms, positive anti-SSA/Ro antibody, and lymphocytic infiltrates and sclerotic changes on parotid gland biopsy. Two years ago, she was diagnosed with extranodal marginal zone B-cell lympho-

ma (EMZBCL) of the left conjunctiva, which was treated successfully with radiation therapy. She was referred to our medical center for evaluation of refractory parotiditis. She denied Raynaud's phenomenon.

At presentation, both parotid glands were enlarged without tenderness, heat, or redness (Figure 1A, arrowheads). High-sensitivity C-reactive protein levels and the

Figure 1. (A) Bilateral parotid glands swelling (arrowheads). (B) Enlarged parotid glands with multiple cysts (arrows) and solid nodules on computed tomography. (C) Strong uptake in the bilateral parotid glands on positron emission tomography. (D) B-cell lymphoma with amorphous amyloid aggregates (asterisk) on parotid gland biopsy (H&E, $\times 150$). (E) Infiltration of CD20 positive cells ($\times 150$). (F, G) Lambda chain restriction on immunohistochemical analysis for lambda (F) and kappa (G) light chains ($\times 150$).

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erythrocyte sedimentation rate were 0.03 mg/dL (normal <0.5 mg/dL) and 33 mm/hr (normal <20 mm/hr), respectively. Rheumatoid factor was 93 (normal <15 U/mL). The antinuclear antibody titer was 1:320 with positive anti-SSA/Ro and anti-SSB/La antibodies. Serum protein electrophoresis revealed mild hypergammaglobulinemia but no M-spike was detected. Serum IgG subclass 4 was 32 mg/dL (normal <86.4 mg/dL) and Schirmer's test was 0/0 mm (left/right eye). A salivary gland scan showed a mild reduction in excretory function. Computed tomography (CT) of the salivary gland revealed enlarged parotid glands with multiple cystic (arrows) and solid nodules (Figure 1B). Positron emission tomography revealed increased uptake by the bilateral parotid glands (Figure 1C).

An ultrasound-guided gun biopsy of the right parotid gland revealed non-Hodgkin's B-cell lymphoma, accompanied by homogeneous, amorphous amyloid aggregates (asterisk) on hematoxylin and eosin staining (Figure 1D). Immunohistochemistry revealed infiltration by lympho-plasma cells (Figure 1E), which showed restricted expression of lambda chains (Figure 1F, G). CT of the chest and abdomen was normal. Bone marrow biopsy showed no malignant cells. A recurrent EMZBCL with amyloidosis of the parotid glands was diagnosed [1-3]. Chemotherapy with rituximab, cyclophosphamide, vincristine, and prednisolone was started. Chronic painless swelling of the

salivary glands in SS patients warrants a further investigation to rule out concurrent lymphoma and/or amyloidosis.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

AUTHOR CONTRIBUTIONS

J.K.P. conceived of the study design. J.Y.K. and J.K.P. performed data acquisition and interpretation. All authors prepared and approved the final manuscript.

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