Sjögren’s syndrome is a chronic auto immune disorder manifested by lymphocytic infiltration of the exocrine glands, particularly the salivary and lacrimal glands. The sicca complex of xerostomia and keratoconjunctivitis is the hallmark of Sjögren’s syndrome.1 Its association with lymphoma is well documented and is a severe complication of primary Sjögren’s syndrome, occurring in 5-10% of the patients followed for more than 10 years.2,3 Lymphomas developing in Sjögren’s syndrome are classified as MALT (mucosa-associated lymphoid tissue) lymphomas, which can be furthered subclassified as either low or high grade.4 Our case is that of a 60 year-old female with insidious slowly progressive development of dry eyes and mouth. There was no evidence of underlying rheumatoid arthritis. Clinically, a soft tissue mass was identified in the parotid area. A Computed Tomography (CT) examination at the level of the parotid glands revealed a nodular appearance of both parotid glands, indicating dilatation of the acini. Also noted was a low-density soft tissue mass in the right parotid gland that measured approximately three centimeters at its greater diameter, which histologically proved to be a lymphoma (Figures 1, 2 and 3). The pathophysiology of lymphoma in Sjögren’s syndrome remains unknown. To date, there is no argument favoring a viral infection or deregulation of a unique oncogene or antioncogene.2 CT and MR can demonstrate findings consistent with Sjögren’s syndrome. The parotid glands reveal a punctate or nodular appearance representing globular collections. These findings, however, are non-specific as they can be seen in chronic sialoadenitis and granulomatous diseases.4 A 60-year-old man presented complaining of a painful palatal ulcer that had been present for 6-8 months. Review of systems revealed a 3 year history of xerostomia and xerophthalmia requiring frequent use of lubricating eye drops and artificial saliva. He had a 20-pound

**Figure 1.** H&E stain, original magnification 40x.
Journal of the American Medical Association

Enero-Marzo 2005

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ABSTRACT

Introduction: Sjögren’s Syndrome is a chronic autoimmune disorder manifested by lymphocytic infiltration of the exocrine glands, particularly the salivary and lacrimal glands.

Methodology: Our case is that of a 60 year-old female with insidious slowly progressive development of dry eyes and mouth. A soft tissue mass was identified in the parotid area. A Computed Tomography (CT) examination revealed a nodular appearance of both parotid glands.

Conclusions: CT and MR can demonstrate findings consistent with Sjögren’s syndrome. The parotid glands reveal a punctate or nodular appearance representing globular collections. These findings are non-specific.

Key words: Sjögren’s syndrome, CT, parotid gland.

loss of weight over the prior three months, general malaise, nonproductive cough, and afternoon fevers. He denied nights sweats, hematochezia, melena, or hematemesis.

Physical examination showed a 2-cm nodule in the tail of the right parotid. A deep palatal ulcer with undermining and gray exudates was present and a second ulcer was noted on the upper left gum adjacent to the incisor. A chest X-ray showed multiple bilateral parenchymal nodules, suspicious for neoplasm.

Biopsy of the palate ulcer revealed only acute and chronic inflammation but no evidence of malignancy. Right parotidectomy was performed. Microscopic exam showed a diffuse lymphohistocytic infiltrate between islands of glandular epithelial cells. Areas of fibrosis were noted. This form of chronic sialadenitis is consistent with Sjögren’s syndrome (Figure 1).

There were also foci of necrosis and a monotonous proliferation of larger, atypical lymphocytes. Immunohistochemical staining confirmed and extranodal marginal zone lymphoma with areas suspicious for B-cell transformation (Figure 2).

Figure 2. H&E stain, original magnification 400x.

Figure 3. CT axial sections post-contrast at the level of the parotid glands revealed a punctate and nodular appearance representing globular collections in both parotid glands (arrow). In another section, a low-density soft tissue mass is identified on the right (arrow).

immunohistochemical staining confirmed and extranodal marginal zone lymphoma with areas suspicious for B-cell transformation (Figure 2).
CME Question

Which of the following statements are true as they relate to lymphoma in Sjögren’s syndrome:

(A) Five to ten percent of patients with Sjögren’s syndrome followed for more than 10 years will develop lymphoma.
(B) Lymphomas developing in Sjögren’s syndrome maybe classified as MALT (mucosa-associated lymphoid tissue) lymphomas.
(C) Lymphoma may appear as a low-density mass in a parotid gland with punctate and nodular appearance representing globular collections.
(D) All of the above

Answer: (D) All of the above

References