

## CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

### Oral Medicine and Pathology Quiz – Case 2

A 73-year-old man presented with a painless palatal swelling, which had been progressively enlarging for the last 5 months. The medical history was significant for epilepsy, asthma, and hypertension, and a history of tuberculosis at age 16; the patient was on chronic oxygen therapy and his medications included phenytoin, valsartan, and inhalations with budesonide, salbutamol and ipratropium. Clinical examination revealed a 4.0x2.0 cm ulcerated soft tissue mass on the left soft palate (fig. 1). Head and neck examination did not reveal lymphadenopathy or other pathologic findings. An incisional biopsy of the palatal mass was performed. Histopathologic examination showed that the connective tissue was diffusely infiltrated by atypical lymphocytes, which focally assumed a pseudonodular pattern (fig. 2). The majority of the lymphocytes were of small size and frequently exhibited clear cytoplasm. Immunohistochemical evaluation revealed positivity for CD20, CD79 and DBA44 (45%), while CD5, CD10, CD23, Cyclin D1, Bcl-2 and Bcl-6 were negative. The cell proliferation index Ki-67 was positive in about 30% of the lymphocyte population. Peripheral blood examination and protein electrophoresis rendered normal values. A bone marrow biopsy was negative for the presence of neoplastic cells. Computed tomography of the head and neck, thorax and abdomen did not reveal enlarged lymph nodes.

#### Comment

Lymphomas of the oral cavity are only rarely encountered. They may represent a component of a more widespread disease or, less often, may begin in the mouth without involvement of other sites. Both the oral soft tissues and the jaws may be affected. The palate, especially the junction between hard and soft palate, contains lymphoid tissue which is considered part of the Waldeyer's ring, and may be the site of lymphomagenesis. Various types of non-Hodgkin's lymphomas have been described to affect the palate including extranodal natural killer (NK)/T-cell, plasmablastic, Burkitt's and small B-cell lymphomas. The latter group comprises several distinct entities, such as follicular lymphoma, mantle cell lymphoma, small lymphocytic lymphoma and extranodal marginal zone B-cell lymphoma of mucosa associated lymphoid tissue (MALT). Staging of lymphomas involving the oral cavity is of paramount importance, since the clinical extent of disease, along with the specific histopathologic subtype, guides the selection of the most appropriate treatment and determines the prognosis.

Extranodal marginal zone B-cell lymphomas or MALT lymphomas arise in extranodal and mucosal sites, more commonly affecting the gastrointestinal tract (50%), lungs, head and neck, ocular adnexae, skin, thyroid gland and breast. Predisposing factors for the development of MALT lymphomas include chronic inflammatory and/or autoimmune diseases, such as *Helicobacter pylori* gastritis, Hashimoto's thyroiditis and Sjögren's syndrome. Patients with Sjögren's syndrome have a 44-times higher risk of developing lymphomas, the vast majority of which are categorized as MALT type. Histopathologically, MALT lymphomas comprise a morphologically heterogeneous B-cell population with a predominance

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Figure 1

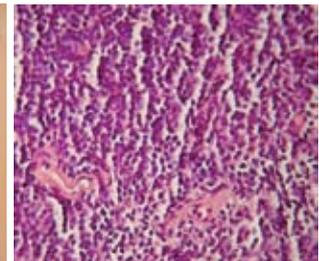


Figure 2

of centrocyte-like cells; involvement of glandular tissue by MALT lymphoma results in the formation of the characteristic lymphoepithelial lesions. Microscopic differential diagnosis includes reactive processes (such as lymphoepithelial sialadenitis) and other small B-cell lymphomas and is based on the careful evaluation of the, sometimes subtle, microscopic differences and immunophenotypic features. Extranodal marginal zone lymphoma is an indolent malignancy which tends to remain localized for a protracted period of time before dissemination period. Most patients present with stage I or II disease and the prognosis is good with an overall 75% 5-year survival.

Besides the various subtypes of lymphoma, a soft tissue swelling or mass of the palate elicits a broad differential diagnosis that ranges from inflammatory conditions (e.g. palatal abscess) to developmental cysts (e.g. nasopalatine duct cyst) to reactive and hyperplastic lesions (e.g. necrotizing sialometaplasia, lymphoid hyperplasia, adenomatous hyperplasia). In addition, several benign or malignant non-lymphoid neoplastic entities may arise in the palate including salivary gland neoplasms (e.g. pleomorphic adenoma and mucoepidermoid carcinoma), and tumors of epithelial (e.g. squamous cell carcinoma), mesenchymal (Kaposi's sarcoma) or melanocytic (e.g. melanoma) origin.

The patient of the present case was admitted to the hospital and received chemotherapy consisted of rituximab, cyclophosphamide, vincristine and prednisolone, which resulted in shrinkage of the palatal mass. He is currently under close follow-up.

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**Diagnosis:** Non-Hodgkin, extranodal marginal zone B-cell lymphoma of mucosa associated lymphoid tissue (MALT lymphoma)