

EXTRANODAL MALT LYMPHOMA PRESENTING WITH ISOLATED HARD PALATE INVOLVEMENT: A DIAGNOSTIC CHALLENGE

SERT DAMAK TUTULUMU İLE SINIRLI EKSTRNODAL MALT LENFOMA: BİR DİAGNOSTİK İNCELEME

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ABSTRACT

Malignant lymphomas of the oral cavity are uncommon lesions and account for 3.5 % of all oral malignancies. In the literature, only a few cases of primary mucosa-associated lymphoid tissue (MALT) lymphoma arising from oral cavity have been described. Here we report a case and diagnostic challenge of a MALT lymphoma of the hard palate and tonsil.

Key words: Malignant lymphoma, Oral, Palate

ÖZET

Oral kavitenin malign lezyonları yaygın değildir ve bütün oral malignitelerin 3.5 %' ini oluştururlar. Literatürde, az sayıda primer olarak ağızda ortaya çıkan mukoza ile ilişkili lenfoid doku (MALT) vakası bildirilmiştir. Bu olgu sunumunda, damak ve tonsilde gelişen MALT lenfomaya tanı konulma aşamalarını anlattık.

Anahtar Kelimeler: Malign lenfoma, Oral, Palatinal

INTRODUCTION

Malignant lymphomas of the oral cavity are uncommon lesions and account for 3.5 % of all oral malignancies.¹ Mucosa-associated lymphoid tissue (MALT) is scattered along mucosal linings. Malignancies that occur in MALT are called MALT lymphomas or MALTomas. In the literature, only a few cases of primary MALT lymphoma arising from oral cavity have been described.^{2,3} Here we report a case and diagnostic challenge of a MALToma of the hard palate and tonsil.

CLINICAL REPORT

A 59-year-old female patient was referred to our clinic for her palatal region swelling. She presented with a three month history of swelling in the middle of the anterior hard palate which was treated by general dentists as an infection but turned to be unresponsive to antibiotics. She denied fatigue, night sweats or weight loss. Her past history and family history were not contributory. The lesion always had been asymptomatic, with no associated pain or

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paresthesia. She had no history of tobacco use and no known allergies and had not undergone any surgeries or received radiation therapy to the head and neck. On examination, the patient appeared well in general. No lymphadenopathy, hepatomegaly, splenomegaly was noticed. There was 2 cm sized nontender swelling on the hard palate (Figure 1). Erythrocyte sedimentation rate (14 mm/h), complete blood count analysis (Hb: 13.1 g/dl, WBC: 7200/mm³, platelets 199,000/mm³) and hemorrhagic diathesis tests were within normal limits. Biopsy from the hard palate was performed. Pathological examination revealed a diffuse, polymorphous proliferation of small and medium-sized lymphoid cells, some with plasmacytic differentiation, and scattered large immunoblasts (Figure 2).



Figure 1. Photograph of the hard palate showing 2 cm sized nontender swelling

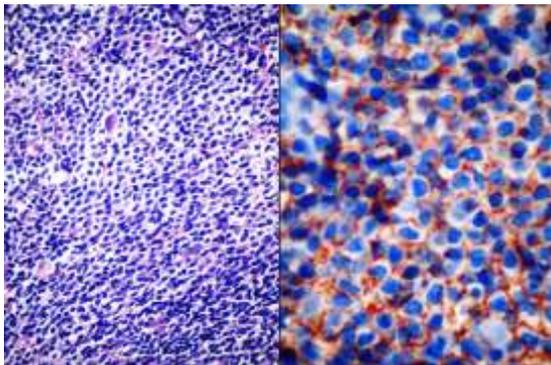


Figure 2. On the left hand side, a neoplastic cellular infiltration composed of small to medium-sized lymphoid cells with slightly irregular nuclei and relatively abundant pale cytoplasm. On the right hand side, positive immunostaining for CD-20 on the neoplastic cells

Immunohistochemical stains demonstrated CD20-positive B cells and negative for CD23, CD10, CD5 and CD3. Staining for immunoglobulin light chains showed that the vast majority of cells contained kappa chain. Ki-67 was positive in approximately 10% of nuclei. The cellular polymorphism, together with the immunophenotypic findings on specimen, supported the diagnosis of MALT lymphoma. Positron emission tomography (PET) CT positron emission tomography/computed tomography (PET/CT) scanning assessed high florodeoxy glucose (fluoro-d-glucose (18F-FDG)) uptake in the hard palate and suspicious uptake in right palatine tonsil (Figure 3). Bone Scintigraphy and PET CT images showed no tumor development another part of the body without hard palate and tonsil (Figure 4). Biopsy specimens obtained by upper gastrointestinal endoscopy did not reveal any lymphomatouse involvement. *Helicobacter pylori* evaluation revealed to be negative. Beta 2 microglobulin was not increased (1.51 mg/L). Protein electrophoresis and serum LDH value were within normal. She was diagnosed as having early stage MALT lymphoma of the hard palate and tonsil. Reevaluation among chronic infection or autoimmune disease including Sjögren syndrome proved to be no another disease association. Irradiation (25Gy) followed by combination chemotherapy (cyclophosphamide, doxorubicin, vincristine and prednisolone plus rituximab) resulted complete regression of swelling. PET-CT showed no evidence of pathological uptake. After three years of the diagnosis she is still well without any symptoms. Due to the nature of this study, it was granted an exemption in writing by the University of Bezmialem IRB. But informed consent was obtained from the patient. I have read the Helsinki Declaration and have followed the guidelines in this investigation.

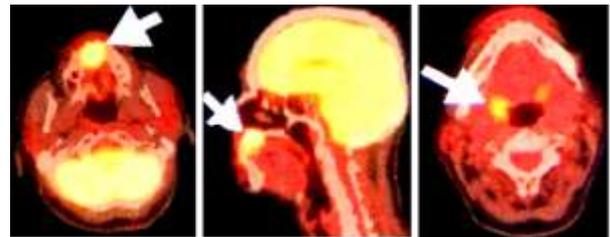


Figure 3. Glucose uptake in the hard palate and tonsil on PET/CT image

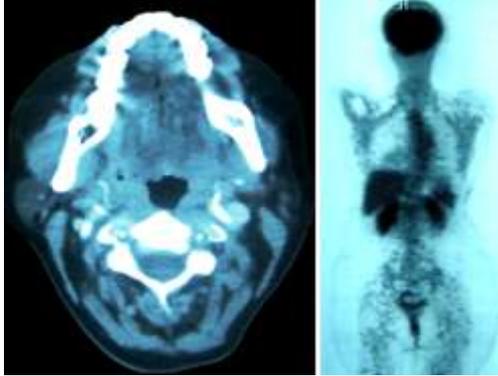


Figure 4. No development of tumor any part of the body without hard palate and tonsil on Scintigraphy and PET/ CT images

DISCUSSION

MALT type lymphoma arising in the oral cavity is rare. MALT lymphomas of the salivary glands are much more common in the parotid gland. Most patients are over 50 years old and predominantly women. MALTomas commonly associated between autoimmune diseases.³ However, our patient did not have any clinical evidence of autoimmune disease.

MALT lymphomas frequently occur in a background of inflammatory disorders, when B cell clones become independent in their growth. In the stomach the antigen that drives the inflammation has been recognized as *Helicobacter pylori*, and in early phases is treated by antibiotic therapy.⁴ Because of tumor primarily arising hard palate and tonsil in our case, it was negative for *Helicobacter pylori* infection. A complete clinical observation is essential for defining the extent of the disease and for planning of a proper treatment protocol. Radiological imaging is essential.⁵ While CT scan is superior in detecting bone destruction, MRI can distinguish tumor tissue from mucosal thickening. Further work-up should include complete physical examination, blood chemistry, PET/CT imaging (of the chest and abdomen), bone marrow biopsy and whole body bone scintigraphy. Histopathological examination must be performed on these lesions to determine the proper treatment and management regimen.⁶ The differential diagnosis of MALT lymphoma must include odontogenic and developmental odontogenic cysts, sarcoidosis, Wegener's granulomatosis, necrotizing sialometaplasia, amyloidosis and carcinomas.⁷

Radiation therapy with or without chemotherapy is useful for treatment of lymphomas. The overall survival of patients with MALT lymphoma

is approximately 85-95 % at 5 years. Late transformation occurs in 10% of cases.⁸ Our patient three years follow up results were not detected recurrence symptoms and she is following regularly for recurrence risk.

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