Primary extranodal marginal zone B-cell lymphoma of hard palate: A case report

Arpan K Shah, Mahesh H Gabhane, Meena M Kulkarni

ABSTRACT

Introduction: MALT (mucosa associated lymphoid tissue) lymphomas comprise a heterogeneous group, originally thought to be derived from the marginal zone B-cells that are found surrounding B-cell follicles and within the adjacent lymphoepithelium. They arise most commonly in extranodal organs such as the stomach, major salivary glands and thyroid. Thus, they can be precisely described as extranodal marginal zone B-cell lymphomas (ENMZL). Here, we report a case of MALT lymphoma arising in the palatal minor salivary glands. Case Report: A 55-year-old woman presented with two years history of a left posterior palatal mass. Clinical investigations of the case included computed tomography (CT) scan for the assessment of bone destruction and possible intra-maxillary extension. Histopathological features and immunohistochemistry findings were consistent with the diagnosis of extranodal marginal zone B-cell lymphoma of MALT type. The lesion was treated by complete surgical excision and followed for two years. Conclusion: Even though oral localization of ENMZL is rare, it should be included in the differential diagnosis of benign-appearing swellings of oral cavity.

Keywords: Extranodal, Histopathology, Lymphoma, MALT, Palate

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INTRODUCTION

Malignant lymphomas of the oral cavity are uncommon and account for 3.5% of all oral malignancies [1]. Lymphomas arising from the mucosa associated lymphoid tissue (MALT) were originally described by Isaacson and Wright in the gastrointestinal tract. They specified them as B-cell lymphomas [2]. MALT lymphomas account for a significant proportion of extranodal lymphomas. In the head and neck region except for the salivary glands the occurrence of these lymphomas is very rare, hence very few cases of primary intraoral MALT lymphoma have been reported so far [3, 4].

In the present paper, we report a case of 55-year-old female with soft tissue swelling involving posterior hard palate. Clinio-radiological profile was suggestive of a chronic inflammatory lesion or a benign neoplasm of palatal minor salivary glands. Microscopic examination
of the lesion was consistent with extranodal marginal zone B-cell lymphoma of MALT type. Clinical work-up, histopathology, immunohistochemical findings, treatment details and follow-up of the case are discussed here.

CASE REPORT

A 55-year-old female presented with a swelling in posterior palatal region since two years. Intraoral examination revealed a swelling involving posterior hard palate on left side, which was soft in consistency (Figure 1). Computed tomography (CT) scan confirmed an oval soft tissue density lesion (19x14 mm) along the left posterolateral aspect of hard palate, partly extending along the soft palate with no evidence of intra-maxillary extension of the lesion (Figure 2). Diagnostic incisional biopsy of the lesion revealed it as benign lymphoid hyperplasia.

After complete surgical excision, microscopic examination of the lesion showed diffuse infiltration of monomorphic small round cells within the connective tissue. Sheets of lymphoid cells were separated from the overlying epithelium by a well-defined band of fibrovascular tissue (Figure 3). Some lymphoid follicles with follicle center and mantle zone were seen. Many minor salivary gland acini and ducts were observed, some of which showed infiltration by lymphoid cells, forming the lymphoepithelial lesions (Figure 4). On higher magnification, the round cells were centrocyte-like, with scant cytoplasm and nuclei containing clumped chromatin. Some plasma cells were also noted. In addition to routine hematoxylin and eosin staining, a panel of immunohistochemical markers was used to arrive at final diagnosis. Leukocyte common antigen (LCA) was used to confirm the neoplasm was indeed composed of lymphocytes. Monoclonal intracytoplasmic immunoglobulin was detected by immunohistochemistry, which confirmed neoplastic origin of the B-cells (Figure 5). The tumor cells were PanCK-, CD5-, CD3-, CD10-, CD20+, and CD23-. Immunohistochemistry for additional markers was not performed because the aforementioned histopathological findings were considered diagnostic for MALT lymphoma. Because fresh tissue was not available for study, flow cytometry and cytogenetic analysis was not performed. Peripheral blood examination and protein electrophoresis were within normal limits. A bone marrow biopsy was negative for the presence of neoplastic cells.

The present case is consistent with the diagnosis of low-grade MALT lymphoma. The patient had uneventful postoperative course.

DISCUSSION

About 20% of oral non-Hodgkin’s lymphomas arise in palatal soft tissues [5]. Extranodal marginal zone lymphomas (ENMZL) constitute a heterogeneous group showing neoplastic cells resembling normal marginal zone B cells [5]. These lymphomas are characterized by their mucosal and glandular tissue localization and commonly referred to as mucosa-associated lymphoid tissue (MALT) lymphomas [6].

ENMZL have peculiar clinicopathological profile that set them apart from other lymphomas. They tend to remain localized for prolonged intervals and the lesion is commonly confused with an inflammatory process (as evident in the present case). However, the presence of monoclonality in these lesions correlates with risk of dissemination and therefore supports their designation as lymphoma [7].

Etiopathogenesis of MALT lymphoma is poorly understood. Chronic inflammatory conditions such as chronic sinusitis, Sjogren’s syndrome, benign lymphoepithelial lesion or myoepithelial sialadenitis (MESA) have been suggested as precursors for the
development of MALT lymphoma in this location [3, 7]. The indolent nature of the disease in these cases is manifested by persistence of the lesion without overt clinical evidence of distant spread even after 14 months [8]. Persistence of the lesion for duration of 24 months in present case is suggestive of MALT lymphoma.

Histopathological distinction between a reactive lymphoid infiltrate and MALT-lymphoma can be difficult. According to Vega et al., larger the lymphoid infiltrate, greater the likelihood of lymphoma [7]. Monomorphous lymphoid population exhibiting centrocyte-like or monocytoid morphology, forming wide zones surrounding epimyoeithelial islands is a useful histological finding suggestive of MALT lymphoma [7]. Kojima et al. described two distinct histopathological patterns of primary oral MALT lymphoma. The first pattern is characterized by occasional follicular colonization and the presence of lymphoepithelial lesions. Another pattern shows a prominent follicular colonization resembling the “floral variant” of follicular lymphoma [4]. Present case showed histological features described in the former pattern.

In contrast to the relatively poor outcome of other B-cell lymphomas, MALT lymphomas have a better prognosis making them amenable to complete surgical removal [3]. Survival in the present case, two years after diagnosis, is consistent with this finding.

**CONCLUSION**

Rarity of occurrence of MALT lymphoma in oral cavity, clinical profile resembling benign process, peculiar histopathology and varied
immunohistochemistry findings make such cases interesting to study. Considering their low-grade malignant behavior and indolent clinical course, ENMZL should be included in the differential diagnosis of benign appearing swelling of oral cavity.

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Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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