Primary Non-Hodgkin's Lymphoma arising from the soft palate: A diagnostic dilemma

Deepak Saharan[1], Amit Kumar Gupta[2], *Akhil Kapoor[3], Ankita Rungta Kapoor[4]

[1]Department of Otorhinolaryngology, Sardar Patel Medical College, Bikaner, Rajasthan, India
[2]Department of Otorhinolaryngology and Head Neck Surgery, Cygnus Hospitals, Delhi-NCR, India
[3]Department of Radiation Oncology, Acharya Tulsi Regional Cancer Treatment & Research Institute, Sardar Patel Medical College, Bikaner, Rajasthan, India
[4]Department of Pulmonary Medicine, North Bengal Medical College, Sushrutnagar, Darjeeling, West Bengal, India
*Email: kapoorakhil1987@gmail.com
Mobile: +919950482121

Abstract

The soft palate houses a wide variety of native tissues that can serve as the source of multiple pathologies. Exact characterization of the lesion is must for the proper management of the disease. We report a case of primary non-Hodgkin's lymphoma arising from the soft palate in a 38-year-old male that was initially misdiagnosed as an inflammatory lesion.

Keywords: Extranodal non-Hodgkin's lymphoma; hard palate; benign lymphoid hyperplasia.

Introduction

The soft palate is a complex sub site of oropharynx that houses a wide variety of native tissues that can serve as the source of multiple pathologies.[1] Besides inflammatory lesions, there is a high possibility of benign and malignant lesions in oral cavity and oropharynx since it is exposed to carcinogens like tobacco. Leucoplakia and erythroplakia are well known benign but premalignant lesions.[2] Squamous cell carcinoma is the most common malignancy of head and neck region. Lymphomas are relatively rare lesions and mimic other pathologies such as periodontal diseases, inflammatory changes or some other malignancy. Lymphoid lesions of the palate can be classified into three main categories: primary lymphoma of the palate, infiltration of the palate as a part of disseminated disease and benign lymphoid hyperplasia (BLH) of the palate.[3] It is imperative to identify the exact variety as the management and prognosis of each of these three is different. We present a case of primary non-Hodgkin's lymphoma arising from the hard palate in a 38-year-old male that was initially misdiagnosed as an inflammatory lesion.

Case Report

A 38-year-old male non diabetic and non hypertensive presented with a swelling in the palatal region from 2 months. The patient gave history of consultation with local doctors and was prescribed a course of antibiotics. However, the lesion did not respond to antibiotics and was progressive. The patient was non smoker and did not have any history of tobacco chewing or betel nut consumption. There was no history of fever, weight loss or night sweating. There was no significant medical or surgical history. On examination, there was a firm mass of size 2.5 X 3 cm with intact overlying mucosa in the region of soft palate [Figure 1]. No ulcer or bleeding was found. No lymph nodes were palpable in the cervical region. Contrast enhanced computed tomography scan revealed a mass confined to the right side of soft palate. The sinuses were free of any disease [Figure 2]. To establish the diagnosis, biopsy was performed from the lesion under local anesthesia. The histopathological examination revealed intact covering of stratified squamous epithelium with underlying diffuse lymphoid cells and follicle-like structures [Figure 3]. Within the follicle-like structures, there were central large cells with a rim of small, round lymphocytes. Inconspicuous nucleolus along with abundant pinkish cytoplasm was present in the large cells. On immunohistochemical study, the large lymphoid cells stained positive for CD20. The surrounding small lymphocytes stained positive for CD5. The staining for CD5, Bcl2 and CD10 were negative. The Ki67 proliferative index was 3%. No other lymph node was found by computed tomography scan thorax and abdomen. The overall picture suggested the diagnosis of low grade B-cell lymphoma of mucosa associated lymphoid tissue (MALT) Stage IA. In view of localized disease, the patient was prescribed local radiotherapy (30 Gy in 15 fractions). The patient had complete remission and is on follow up.

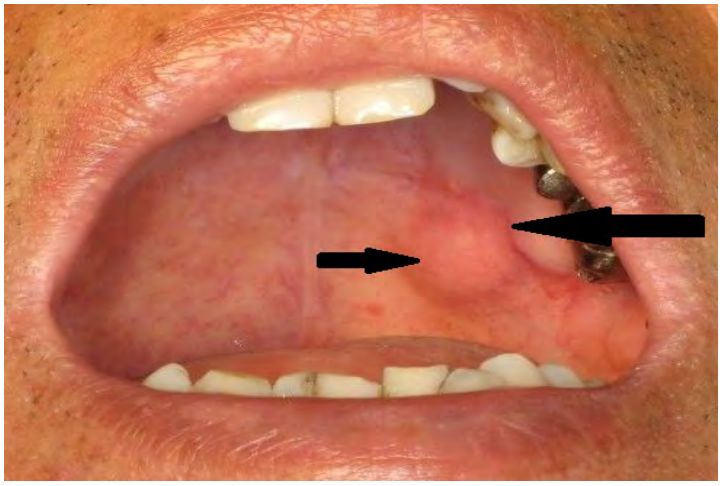

Figure 1: Clinical picture showing a mass with intact overlying mucosa in the region of soft palate

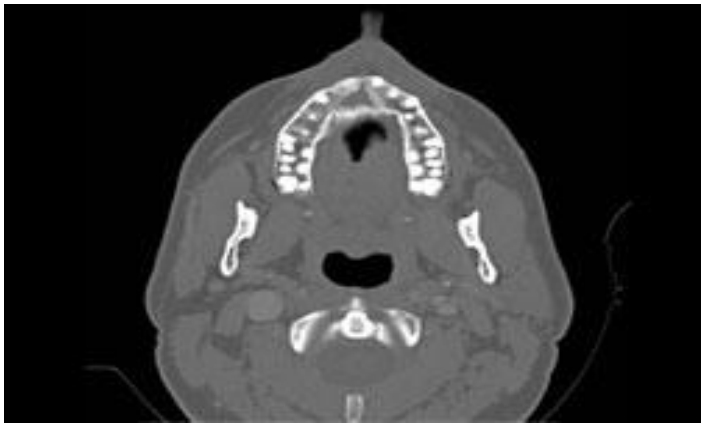

Figure 2: Contrast enhanced computed tomography scan revealed a mass confined to the soft palate

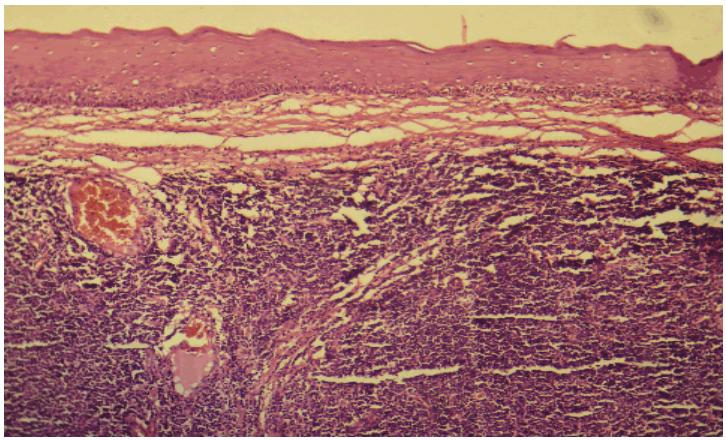

Figure 3: Histopathological examination showing intact covering of stratified squamous epithelium with underlying diffuse lymphoid cells and follicle-like structures

Discussion

Specialized lymphoid tissue is found in association with certain epithelia, in particular the naso- and oropharynx (Waldeyer's ring: adenoids, tonsils), the gastrointestinal tract (gut-associated lymphoid tissue: Peyer's patches of the distal ileum, mucosal lymphoid aggregates in the colon and rectum), and lung (bronchus-associated lymphoid tissue).[4] Collectively, this is known as mucosa-associated lymphoid tissue (MALT). These tissues tend to have prominent B-cell follicles with broad marginal zones but also may have discrete T-cell zones, similar to the paracortex of lymph nodes. MALT is thought to function in responding to intraluminal antigens and the generation of mucosal immunity. Extranodal marginal zone B-cell lymphoma of MALT is an extranodal lymphoma consisting of heterogeneous small B cells, including marginal zone (centrocyte like) cells, monocytoid cells, and small lymphocytes in varying proportions.[5] As these diseases tend to remain localized for

long periods of time, local treatment (surgery or radiation therapy) is effective at long-term control of disease. In particular, low doses of RT (30 Gy) almost always control sites of disease.

In lymphoma, the swelling is non-tender and rarely ulcerated. In this case, the patient had non-ulcerated firm swelling in the soft palate region with overlying intact mucosa. Benign lymphoid hyperplasia (BLH) is a close differential diagnosis of low grade lymphoma in the palate region. In the past, many low grade lymphomas were misdiagnosed as BLH.[6] Extranodal infiltration composed of small round lymphoid cells were usually classified as 'pseudolymphomas' since the disease was shown to pursue an indolent clinical course.[5] However, the term pseudolymphoma or BLH should usually be restricted to reactive lymphoid infiltrate that lack the evidence of clonality on immunophenotyping studies. In BLH, the cells demonstrate all stages of follicular center cell transformation while in case of malignant lymphomas, the range of cells is narrow with nuclear atypia.[6] Since there are no major clinical or histologic differences between BLH and the palatal lymphomas, the immunophenotyping and molecular genetic techniques assume important role in differentiating them. Hence, immunohistochemical study was performed in our patient in whom the large lymphoid cells stained positive for CD20 while surrounding small lymphocytes stained positive for CD5. The staining for CD5, Bcl2 and CD10 were negative with Ki67 proliferative index 3%. These findings confirmed our diagnosis of low grade B-cell lymphoma of MALT.[7] Involved site radiotherapy (ISRT) with radiation dose of 24-30 Gy (2 Gy per fraction) is the most effective treatment for low grade MALToma Stage I and II. Observation alone is an option when the biopsy is excisional and complete disease is removed.[8] However, if the margins are positive post surgery/biopsy, local radiotherapy should be considered. Since our patient had stage I low grade disease, local radiotherapy with 30 Gy was given in 15 fractions (2 Gy per fraction). On completion of radiotherapy, the patient had complete remission and is on follow up.

Conclusions

Primary non-Hodgkin's lymphoma arising from the palate, though a rare entity, should be kept in the differential diagnosis of palatal lesions. Immunohistochemistry may be necessary to differentiate benign lymphoid hyperplasia and low grade lymphomatous lesions.

References

[1] Rooney N, Ramsay AD. Lymphomas of the head and neck 2: The B cell lymphomas. Eur J Cancer B Oral Oncol. 1994;30B:155–9.
[2] Ugboko VI, Oginni FO, Adelusola KA, Durosinmi MA. Orofacial non Hodgkin's lymphoma in Nigerians. J Oral Maxillofac Surg. 2004;62:1347–50.
[3] Mawardi H, Cutler C, Treister N. Medical management update: Non Hodgkin lymphoma. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2009;107:e19–33.
[4] Wolvius EB, van der Valk P, van der Wal JE, van Diest PJ, Huijgens PC, van der Waal I, et al. Primary extranodal nonHodgkin's lymphoma of the oral cavity. An analysis of 34 cases. Eur J Cancer B Oral Oncol. 1994;30B:121–5.
[5] Harris NL, Jaffe ES, Stein H, Banks PM, Chan JK, Cleary ML, et al. A revised European American classification of lymphoid neoplasms: A proposal from International lymphoma study group. Blood. 1994;84:1361–92.
[6] Makepeace AR, Fermont DC, Bennett MH. Non Hodgkin's lymphoma of the nasopharynx, paranasal sinus and palate. Clin Radiol. 1989;40:144–6.
[7] Medeiros LJ, Harmon DC, Linggood RM, Harris NL. Immunohistologic features predict clinical behavior of orbital and conjunctival lymphoid infiltrates. Blood. 1989;74:2121–9.
[8] Adkins KF. Lymphoid hyperplasia in the Oral Mucosa. Aust Dent J. 1973;18:38–40.