Primary Extranodal Non-Hodgkin’s Lymphoma of the Palate: A Diagnostic and Therapeutic Challenge

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ABSTRACT

Lymphomas arising in extranodal sites are intriguing. Oral lymphomas are uncommon and most often mimic other pathological entities, such as dentoalveolar abscess, periodontal abscess, infected dental cyst, or benign jaw tumors. The diagnosis of oral extranodal lymphoma is challenging due to a low index of clinical suspicion. A case of primary extranodal lymphoma presenting as a swelling of the hard palate is presented here. The diagnostic and treatment options of oral lymphomas are also discussed.

Keywords: Extranodal, Oral lymphoma, Palatal swellings.

How to cite this article: Varghese AK, Nair S, Babu SS. Primary Extranodal Non-Hodgkin’s Lymphoma of the Palate: A Diagnostic and Therapeutic Challenge. Oral Maxillofac Pathol J 2014;5(1):453-455.

Source of support: Nil
Conflict of interest: None

INTRODUCTION

Lymphomas are a heterogeneous group of malignant tumors of the lymphoreticular system which are known for their spectrum of behavior ranging from relatively indolent to highly aggressive and potentially fatal. Lymphomas can be classified as Hodgkin’s lymphoma (HL) or non-Hodgkin’s lymphoma (NHL). HL rarely shows extranodal disease (1%) in contrast to NHL (23-30%).

NHL arising from tissue other than lymph nodes and even from sites which normally contain no lymphoid tissue is referred to as primary extranodal lymphoma. The definition of primary extranodal lymphoma, particularly in the presence of both nodal and extranodal disease, remains a controversial issue. Operationally, lymphomas can be considered as extranodal when, after routine staging procedures, there is either no or only ‘minor’ nodal involvement along with a clinically ‘dominant’ extranodal component, to which primary treatment must often be directed. It has recently been demonstrated that NHL on the whole are showing a rapid increase in incidence, and over the past 20 years extranodal disease increased more rapidly than nodal disease. In addition to the AIDS epidemic, predisposing factors such as viral infections, immunosuppressive treatments, or environmental factors, might explain the increased incidence of extranodal lymphomas. Gastrointestinal localizations represent the most common form of extranodal lymphoma. Oral cavity as a primary site constitutes only 2% of all extranodal NHL. The rarity of oral cases of primary extranodal NHL makes diagnosis, understanding of biological behavior and therapeutic options difficult.

CASE REPORT

A 68-year-old female patient consulted the Department of Oral Medicine and Radiology, Pushpagiri College of Dental Sciences, Trivullla, Kerala, India with the complaint of a swelling on the right side of the roof of the mouth since 2 months. She gave a history of noticing the slow growing, painless swelling because of the inability to wear her upper complete denture. Her medical history was positive for diabetes, hypertension and dyslipidemia for which she has been on medication for the past 15 years. The patient had undergone total extraction 12 years ago and subsequent prosthetic rehabilitation with complete dentures. Family history was noncontributory.

EXAMINATION

On extraoral examination, the patient was afebrile and there was no regional lymphadenopathy. On intraoral examination, a 4 x 4 cm diffuse, nonulcerated swelling was seen on the right postero-lateral region of the hard palate extending from the canine region to the tuberosity. Mediolaterally, it extended from the alveolar ridge to 1 cm from the midline (Fig. 1). On palpation, the swelling was nontender, nonfluctuant and firm. No associated sinus or discharge was noticed.

A provisional diagnosis of a salivary gland tumor was made considering the site and nature of the lesion.

INVESTIGATIONS

A panoramic radiograph revealed no bone loss in the region of the swelling. The hemogram and biochemical profile

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were within normal limits. Fine-needle aspiration cytology revealed a mixed inflammatory cell infiltrate and the report was inconclusive. An incisional biopsy was done from a representative area of the lesion.

The histopathology of the H & E stained section showed the presence of an atrophic nonulcerated stratified squamous epithelium overlying connective tissue stroma with a diffuse proliferation of small, round monomorphic cells. The tumor cells were uniform, with single prominent nucleus and scanty eosinophilic cytoplasm. Few mitotic figures were observed (Fig. 2). A diagnosis of a round cell neoplasm suggestive of non-Hodgkin’s lymphoma was made.

Immunostaining with CD45 and cytokeratin showed diffuse positivity of the cells with CD45 (Fig. 3) and was negative for cytokeratin thus, confirming the diagnosis of non-Hodgkin’s lymphoma.

**MANAGEMENT**

The patient was referred to a regional oncology center where a bone marrow biopsy and PET scan was done. Bone marrow examination revealed no evidence of infiltration and the PET imaging failed to detect tumoral site anywhere else in the body.

A final diagnosis of primary extranodal non-Hodgkin’s lymphoma of the palate stage IA (low grade under the working formulation) was made.

The patient was started on chemotherapy and is presently into the second cycle of CHOPR therapy.

**DISCUSSION**

Oral NHL are of significance because of their rarity, nonspecific clinical appearance, and many a times their presentation in the oral cavity as the first identifiable evidence of the disease. It may appear as swelling, ulceration, exophytic masses, delayed healing of extraction sites, or trigeminal neuropathy. Signs and symptoms at presentation depend largely on the localization; generally, patients with extranodal lymphomas tend less often to present B symptoms (systemic symptoms-like fever, unexplained weight loss) than do patients suffering from lymphomas arising in the nodal regions.

The etiology of extranodal lymphomas appears to be multifactorial and includes immune suppression, infections (viral and bacterial), and exposure to pesticides and other environmental agents. The histological spectrum of extranodal lymphomas somehow differs from that of nodal lymphomas. Nearly half of the extranodal cases are of diffuse large cell histology.

The clinical outcome varies among all the specific sites of primary extranodal lymphomas. In fact, the histological
subtype is undoubtedly the main predictor of prognosis in either nodal or extranodal lymphomas.\textsuperscript{7}

In the present case, the patient presented with a palatal swelling which mimicked a benign salivary gland neoplasm and the histopathological appearance also resembled a chronic inflammatory condition. A proper diagnosis could be arrived at by careful histopathological examination aided by immunohistochemical staining.

FDG positron emission tomography (PET) imaging has been shown to be an important technique for both staging and follow-up of nodal and extranodal lymphoma and the same was used to stage the disease in this patient.

Treatment options for head and neck NHL are chemotherapy, radiation, or both. The initial treatment for most patients is known as CHOPR chemotherapy (cyclophosphamide, hydroxydoxorubicin, oncovin, prednisone and rituxan). Radiotherapy is used in early stages for massive tumors. About 45\% of patients may be cured using this protocol.\textsuperscript{8} Survival depends on the extent of the disease, presence of HIV disease, histopathology, and Ann Arbor staging. According to Alexander et al, for extranodal head and neck lymphoma 5-year survival rate is approximately 50\%, whereas median survival rate for stage IE is 10 years.\textsuperscript{9}

CONCLUSION

The extranodal lymphomas represent a challenge in routine lymphoma diagnosis, due to the variety of histological types, molecular abnormalities and clinical pictures that can be present. Correct diagnosis and appropriate treatment of extranodal lymphoma are also complicated by the relative rarity of many of these tumors.

Patients with palatal enlargements are seen very commonly in daily clinical practice, but to differentiate neoplastic lesions from non-neoplastic/reactive lesions is essential in treatment planning. Biopsy of these enlargements, histopathological examination, and immunohistochemical analysis will aid in accurate diagnosis and treatment.

REFERENCES