Case Report

Non-Hodgkin's lymphoma of maxillary sinus: An unusual presentation

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ABSTRACT

Non-Hodgkin's Lymphoma (NHL) are a group of neoplasms that originate from the cells of the lymphoreticular system. Forty percent of Non-Hodgkin's lymphoma arises from extranodal sites. The nasal cavities and paranasal sinuses are rarely affected by primary NHL. Common primary extranodal sites of lymphomas include stomach, liver, soft tissue, dura, bone, intestine and bone marrow. Most patients present with rapidly enlarging masses, often with symptoms both locally and systemically (fever, recurrent night sweats, or weight loss). The vast majority of patients with localized disease are curable with combined modality therapy or combination chemotherapy alone. About 50% patients are cured with doxorubicin based combination chemotherapy and rituximab. An atypical case of extranodal Non-Hodgkin's lymphoma of maxillary sinus is discussed.

Keywords: Chemotherapy, diffuse large B-cell, immunohistochemistry, maxillary sinus, non-Hodgkin's lymphoma

INTRODUCTION

Lymphomas are malignant neoplasm of lymphoreticular cells. Malignant lymphomas of the oral cavity represent 5% of all lymphomas[1] and are most common among male patients between 50-70 years of age.[2] Hematologic malignancies are very often seen in immunocompromised patients.[3] Large B-cell lymphoma (LBCL) is the most common non-Hodgkin's lymphoma. LBCL is a fast growing malignancy that may arise inside or outside of the lymphatic system. Skin, abdomen, lung, central nervous system, and oral cavity are common locations.[4] Lymphoma arising within a single bone, without visceral or lymph node involvement, is a rare condition and constitutes 3.1% of malignant bone tumors and 5% of extranodal lymphomas. The etiology is unknown but it is thought to be due to virus and immunosuppression**.** We present a case reported with swelling on left maxilla which was previously diagnosed as peripheral giant cell granuloma. There is a general agreement that involvement of lymph nodes is common with oral lymphomas.[5] However our patient did not have any lymph node involvement.

CASE REPORT

A 65 year old man presented with a growth in the upper left alveolus with nasal obstruction for the past 6 months. He also complained of a mild intermittent pain associated with sudden ulcerated growth on the left maxillary alveolar ridge. He had undergone extraction of upper left posterior tooth 5 years back. He did not give any contributory medical and family history. He did not chew or smoke tobacco or use betel nut. On extraoral examination a non-tender, firm swelling involving the left side of face extending from infraorbital rim till 2 cms short of lower border of mandible superoinferiorly [Figure 1]. Intraorally the surface of the growth was covered with a yellowish necrotic material and it was slightly tender on palpation. It was extending to the buccal vestibule and the palatal side of alveolus in the region extending from tooth number 25 till left maxillary tuberosity region [Figure 2]. There were no palpable lymph nodes. The clinical differential diagnosis included the most common malignancies in the oral cavity such as squamous cell carcinoma (SCC), minor salivary gland tumor, and carcinoma of the maxillary sinus. Investigations revealed normal complete

hemogram, blood sugar, and liver and kidney function tests. The patient's Enzyme- linked immunosorbent assay for Human immunodeficiency virus and Venereal Disease Research Laboratory were negative. Scrapings taken from the involved region to rule out fungal granulomas did not show any fungi. On radiographic examination in a Waters and Caldwell view, opacification was seen in left maxillary sinus with destruction of anterior and posterolateral wall of maxillary sinus. Computed tomography (CT) scan was performed and sections were taken in the axial, coronal, and sagittal planes. Axial section revealed a large soft tissue density lesion with its epicentre in the maxillary sinus, causing destruction of all walls of the maxillary sinus, extending medially into the lateral wall of the nasal septum, left nasal cavity involving the inferior and middle turbinates, laterally into the pterygopalatine fossa and inferiorly causing destruction of the hard palate and alveolar processes of the left maxilla in the molar region [Figure 3]. Next a biopsy was taken for histopathological evaluation, which was suggestive of peripheral reparative giant cell granuloma. In doubt, a repeat biopsy for immunohistochemistry findings was done. Immunohistochemistry reports were suggestive of non-Hodgkin's lymphoma diffuse large B cell type[6] with tumor cells positive for CD20 [Figure 4]. The patient was staged IE according to the Ann Arbor Staging system. He received 6 cycles of standard (R-CHOP) regimen, Rituximab 375 mg/m² d1 (Day 1),[7] Cyclophosphamide 750 mg/m² d1, Doxorubicine 50 mg/m² d1, Vincristine

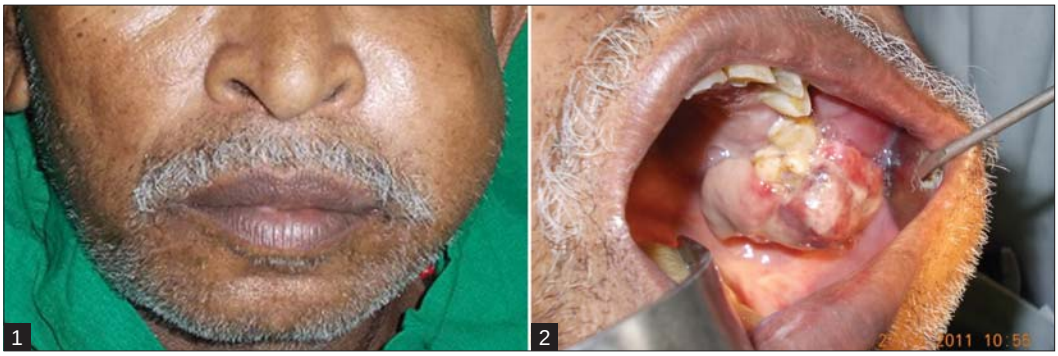

Figure 1 and 2: (1) Extraoral presentation (2) Intraoral presentation

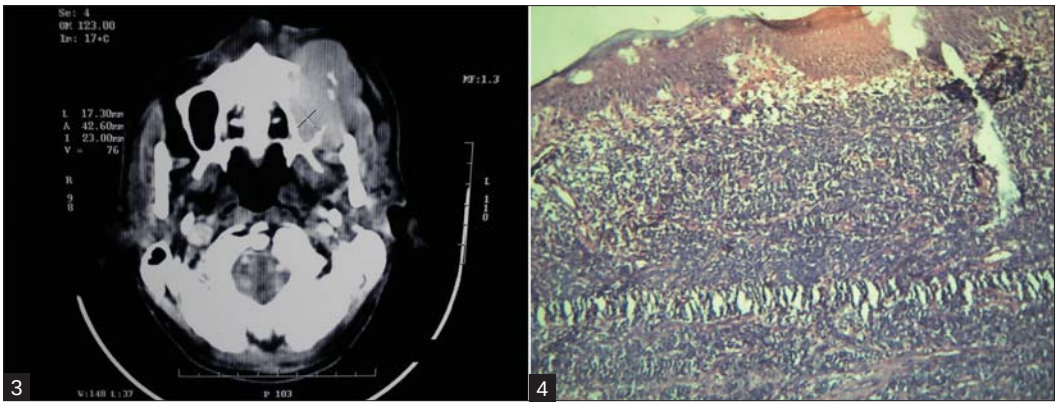

Figure 3 and 4: (3) CT Image (4) Photomicrograph (H & E Stain, Under 100x magnification)

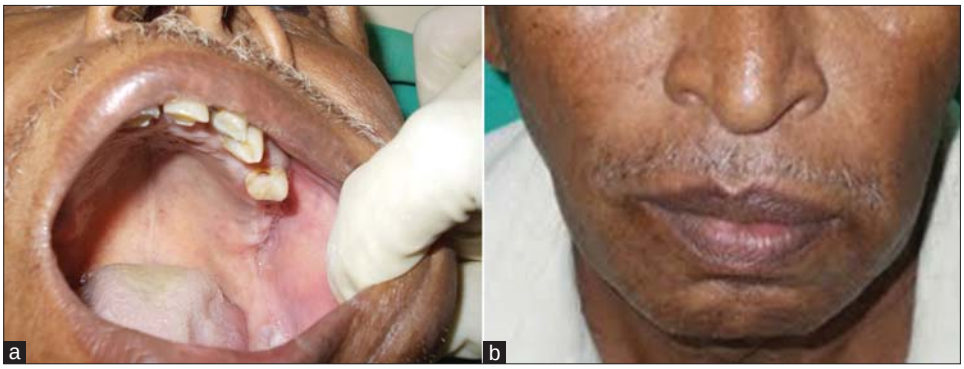

Figure 5: (a) Intraoral presentation post chemotherapy, (b) Extraoral presentation post chemotherapy

1.4 mg/m² d1, and Prednisolone 100 mg (Day 1-5). The response to the treatment was successful [Figure 5a]. Post treatment the patient is in follow up over past 1 year and no recurrence is seen till date [Figure 5b].

DISCUSSION

Non-Hodgkin's lymphoma has a varied manner of presentation, response to therapy and prognosis. A few cases of oral lymphomas were reported in association with acquired immune deficiency syndrome (AIDS), and it might even be the first presentation of AIDS in certain individuals, but in our case it was negative. Soft tissue lymphoma of the oral cavity and primary lymphoma of the jaw are often misdiagnosed. Clinically and radiographically, the manifestation is usually similar to squamous cell carcinoma or to an odontogenic tumor, cyst, or infection. When oral soft tissue lesions first appear, they are relatively soft and often have an overlying ulceration, and are often characterized by absence of other symptoms. If bone is the primary site, tooth mobility and alveolar bone loss are often noted. Pain, swelling, numbness of the lip, and pathologically related fractures may be associated with the bone lesion.[8] Lymphomas are usually submucosal, and on gross appearance, differ from SCC which is usually ulcerative. Our case clinically manifested as an ulcerated growth closely resembling SCC, and it was very difficult to differentiate both of the lesions clinically. Generally histopathological results are assumed to be standard for final diagnosis along with clinical and radiological correlation, but a histopathological diagnosis of peripheral reparative giant cell granuloma was unacceptable due to the fast pace spread of the lesion and also the amount of destruction of surrounding structures towards midline of body which is very atypical for a peripheral giant cell granuloma. Moreover the lesion was spreading on medial as well as lateral sides of left maxillary sinus equally which again created a doubt about its origin. Radiographically and clinically it was suggestive of squamous cell carcinoma and histopathologically it was proved peripheral giant cell granuloma but after immunohistochemistry turned out to be non-Hodgkin's lymphoma which is a very different and rare instance. Histopathology and immunohistochemistry examination should be performed to ensure the accurate diagnosis and histological grading of lymphoma. Management also varies depending on the stage of lymphoma. Paranasal lymphomas have a poor prognosis, which is usually worse than that associated with lymphomas in other sites in the body.[9]

The initial treatment for most patients is combination of chemotherapeutic drugs. Radiotherapy is used in early states for massive tumors.[10] Early diagnosis will improve the prognosis. However, patients older than 60 years, in stages 3 or 4 or having several extranodal places of involvement, may have an unfavourable prognosis. The initial response to treatment is good but this entity shows a prolonged course with remissions and exacerbations. A continuous monitoring and close follow up is recommended.

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