CASE REPORT

A Case Report on Primary Non-hodgkin's Lymphoma of the Mandible

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ABSTRACT

Non-Hodgkin's lymphomas (NHL) are malignancies originating from the lymphoid system. NHL presents with an extra nodal pattern; Intraoral site lymphomas are rarely seen. These tumors frequently resemble other diseases such as chronic osteomyelitis and odontogenic neoplasms, further radiographic evaluations like CT/CBCT and histopathologic examination (Immunohistochemistry) facilitate early appropriate diagnosis. Diagnosis of NHL is challenging without radiographic and histopathological reports. This case report helps a clinician to understand the possibility of complications, and the importance of history taking and X-ray before any extraction. In this case, we report an uncommon incidence of NHL affecting the jaws of a 35-year-old female patient.
Malaysian Journal of Medicine and Health Sciences (2025) 21(SUPP13):159-161. doi:10.47836/mjmhs.21.s14.22

Keywords: Lymphoma, Non-Hodgkin's, Immunohistochemistry, Cone-Beam Computed Tomography

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INTRODUCTION

Primary non-Hodgkin's Lymphoma commonly affects extra nodal sites such as skin, the Gastrointestinal tract, and less often bone. However, Waldeyer's ring, paranasal sinuses, salivary glands, osseous structures, and the oral mucosa are all commonly affected in the head and neck region. The mandible accounts for only 0.6% of isolated malignant non-Hodgkin's lymphomas. Primary non-Hodgkin's lymphoma of the jaws can be difficult to identify owing to its varied clinical appearance, as it can mimic an odontogenic cyst, tumor, or fibro-osseous lesion. (1)

CASE REPORT

A 35-year-old female presented with swelling on the left back side of the lower jaw for one year. She reported that the swelling had been gradually increasing and had suddenly enlarged after tooth extraction 10 days ago. The patient walked in with a normal gait and appeared moderately built and nourished. Her vital signs were within normal limits.

Extra-oral examination revealed facial asymmetry with diffuse swelling on the left side of the face, extending superiorly from the ala-tragus and inferiorly to the lower border of the mandible (Fig. 1a). Anteriorly, the swelling was 2 cm from the angle of the mouth and extended posteriorly 3 cm from the pinna. The swelling was non-tender on palpation, with no local rise in temperature. Intra-oral examination showed an extraction socket in the 36 and 37 regions with granulomatous tissue present (Fig. 1b). There was vestibular obliteration and expansion of the buccal cortical plates, with tenderness on palpation. The neurological examination of trigeminal nerve showed no altered response. No significant findings regarding the periodontal status of 33 and 34 were evident. Based on the extraction history and clinical presentation, a provisional diagnosis of a residual cyst in relation to the 36 and 37 regions and a dentigerous cyst in relation to the 38 region was made. Differential diagnoses of ameloblastoma, odontogenic keratocyst, and odontogenic myxoma were also considered.

An orthopantomogram revealed a multi-locular radiolucency with a soap bubble appearance in the 34 to 37 regions, extending to the mandibular body, involving the inferior alveolar canal suggesting ameloblastoma. There is also evidence of horizontal bone loss in relation to teeth 18, 17, 12, 11, 21, 22, 34, 33, 32, 31, 41, 42 and 43 (Fig. 2). No significant changes were noticed in

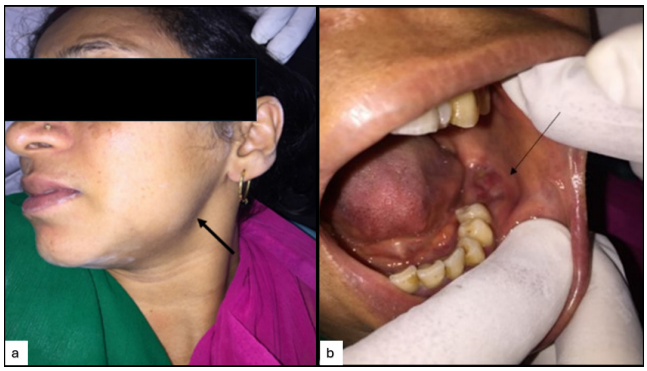

Figure 1: a. Extra-oral image and b. Intra-oral examination showed an unhealed extraction socket in the 36 and 37 regions with granulomatous tissue.

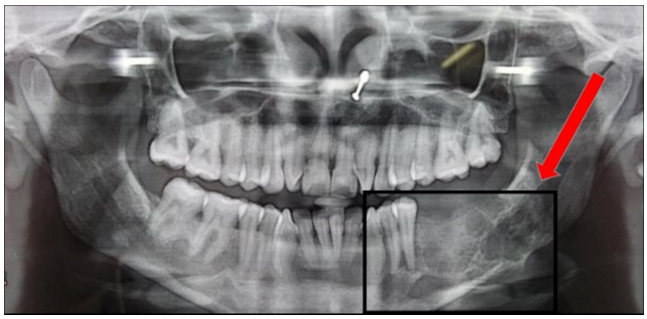

Figure 2: Orthopantomogram showing a multi-locular radiolucency

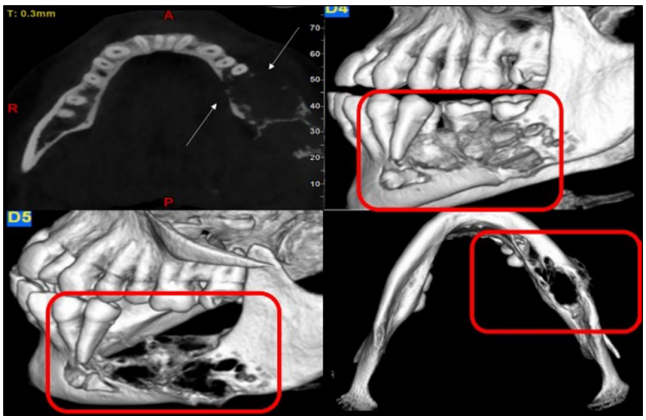

Figure 3: CBCT sections showing the destruction of buccal and lingual cortical plates

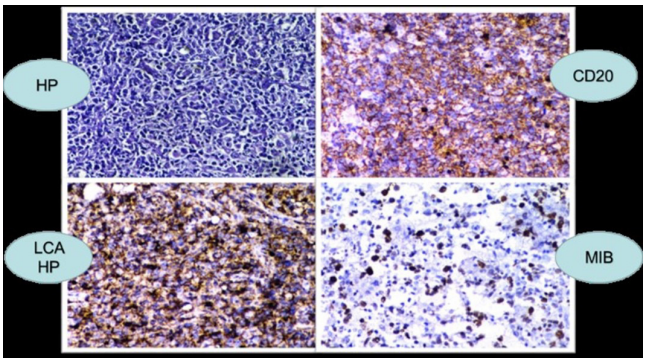

Figure 4: Histopathological sections showing atypical lymphoid cells, IHC showing positive for LCA, MIB and CD20

Maxillary Bone, Temporomandibular Joint and Maxillary Sinus. CBCT sections showed destruction of the buccal and lingual cortical plates on the axial sections. The reconstructed panoramic image revealed a multilocular radiolucency extending from the 34 to 38 regions. In 3D images, bone destruction and a pathological fracture of the inferior border of the mandible on the left side were observed (Fig. 3).

An incisional biopsy was performed, and the specimen was sent for histopathological investigation and immunohistochemistry. Histopathological sections showed atypical lymphoid cells with a polyhedral shape and scant eosinophilic cytoplasm. The nuclei displayed irregular membranes, open chromatin, and prominent nucleoli. Foci of atypical mitotic figures were also observed, with lymphoid cells infiltrating the bone. The immunohistochemistry profile was positive for LCA and CD20, which are classical for non-Hodgkin's lymphoma of the mandible (Fig. 4).

Considering the clinical, radiographic, and histopathological findings, a final diagnosis of primary non-Hodgkin's lymphoma of the mandible was made. A combination therapy was planned, including surgical hemi-mandibulectomy with free fibula flap reconstruction, followed by chemotherapy. However, treatment could not be initiated as the patient was unwilling to undergo any treatment. The patient was referred to the department of psychology for counselling. However after attending three sessions of counselling patient was not convinced for allopathy treatment but considered alternative treatment modalities like ayurveda or any other naturopathy treatments.

DISCUSSION

NHL arises primarily within the lymph nodes, but approximately 24% affect extra nodal locations. (2) Most of the cases initially present as an odontogenic infection. Parrington et al. discussed a case of primary lymphoma of the mandible presenting after tooth extraction, wherein the mucosal lesion had a similar presentation of protruding from the socket during the post-op review. (3) As of this case we had to reconsider many factors inclusive of differential diagnoses for the tumors involving the mandible like multiple myeloma, Ewing sarcoma, Langerhans cell histiocytosis, leukaemia, osteosarcoma, bone metastasis, and osteomyelitis. (4) As of the present case leukaemia and Langerhans cell histiocytosis were considered as differential diagnoses, ameloblastoma, odontogenic keratocyst, odontogenic myxoma, Ewing sarcoma, bone metastasis, and osteomyelitis as radiographic differential diagnoses.

Primary NHLs are frequently misdiagnosed because of their low incidence, vague presentation, and its similarity to other more prevalent dental diseases. When it pertains to the jaw bones, there are no standard clinical presenting signs. These lesions may manifest as cervical lymphadenopathy, pain, numbness, jaw swelling, or tooth mobility. The clinical features usually resemble either as localized osteomyelitis or as an odontogenic lesion. Thus, a thorough clinical, panoramic X-ray and CT examination is required to accurately diagnose a patient with non-Hodgkin lymphoma of the head and neck. This will in turn lead to avoiding of unnecessary invasive dental procedures and prompt to perform a biopsy. Since NHL affecting the mandible as a unique localization is extremely rare and the lack of literature on its classic imaging findings, it is important to perform further investigations in a timely manner to aid in early diagnosis (2,4).

The treatment of NHL is usually custom formulated based on the grade and histological type either using chemotherapy or radiotherapy or a combination of both. (4) In our reported case, we planned combination therapy. The standard chemotherapy includes the CHOP (Cyclophosphamide, Doxorubicin, Vincristine, and Prednisone) regimen. A variant of the same RCHOP, i.e. in combination with Rituximab (anti-CD20 chimeric antibody) has been frequently used in recent times. Radiotherapy in the range of 2400 – 5600 c Gy (35-40 Gy) delivered in 180 c Gy daily fractions has proven successful. Primary lymphoma of bone has an excellent prognosis, more so in stage I and with combination therapy. (5) Unfortunately, we could not start with the treatment since the patient was unwilling to continue any medical or surgical intervention.

CONCLUSION

It never easy to diagnose a case with NHL, knowing the fact that survival rate is very low in such cases, we must strategize management in systematic way so that we can avoid any catastrophe. Psychological counselling plays a major role in treating such diseases, since the systemic illness can also be complimented along with mental illness. With hope, we live!

ACKNOWLEDGEMENT

Informed consent was obtained from the patient for the publication of this case report, including the use of any related data or images. All patient identifiers have been removed or anonymized to ensure confidentiality.

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