CASE REPORT

An unusual case report of primary non hodgkin’s lymphoma of mandible

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ABSTRACT
Mandible accounts for 0.6% for isolated Primary Non Hodgkin’s lymphoma (PNHL). HIV associated PNHL presents with advanced disease and extra nodal involvement in up to 90% of patients. In the mandible main symptoms are pain, swelling, numbness, tooth mobility and cervical lymphadenopathy. Conventional X-Ray and CT scan is required to diagnose and to know the extent of the lesion. However the final diagnosis depends on histopathology and immunohistochemistry. HIV associated NHL has a poor prognosis.

Keywords: Primary Non Hodgkin’s Lymphoma, Mandible, Radiology, Immunohistochemistry.

INTRODUCTION
Primary Non-Hodgkin’s lymphoma (PNHL) involving extranodal sites accounts for 24–45% of the cases1,2. In 1963, the term primary lymphoma of bone was introduced by Ivins and Dahlin. The etiology is unknown, even though virus and immunosuppressant are implicated. The commonest site of extranodal disease is the gastrointestinal tract particularly the small intestine, stomach, and perianal region3. The mandible accounts for only 0.6% of the isolated malignant non-Hodgkin’s lymphoma4. Most common symptom is swelling of the jaw and pain. The diagnosis of lymphoma of the mandible is often delay because of over lapping of radiological features of squamous cell carcinoma. The histopathological diagnosis has to be confirm by immunohistochemistry.

CASE REPORT
History: A 67 year old female presented with swelling over right side of cheek, difficulty in swallowing and low grade fever for two months. No past history of tobacco chewing. Examination: A 10x10 cm size swelling was present on right side of cheek with overlying tense skin, numbness over right cheek, palpable lymphnodes in right submandibular (level 1B) and upper jugular (level 2). Investigation: Haemoglobin 11.2 gm%, total WBC count 8200 cell/cu.mm, ESR was within normal limit. Patient was HIV +2 positive. Chest X ray is normal. X-ray skull with mandible showed large soft tissue radio-opacity mass lesion in right side of mandible with osteolytic destruction of underlying right ramus and right side of body of mandible. Multiple osteolytic lesions in the skull vault (Figure-1). On CT scan neck, a 83 (Crinocaudal) x 75(Transverse) x 81(Antero- Posterior) mm size well defined soft tissue density lesion was noted in right side of mandible with destruction of underlying right ramus and body of mandible. Lesions shows heterogenous post contrast enhancement (pre contrast HU of 47 and post contrast HU of 86) with non enhancing necrotic area within. (Figure-2) Superiorly lesion erode the right maxilla and extend up to the level of right temporalis muscle. Medially lesion displaced tongue towards the opposite site. Another osteolytic lesion was also noted in left side of body of mandible. Multiple permeative type of osteolytic lesions noticed in skull. (Figure-3) Right side level 1b and level 2 lymph nodes appeared enlarge. On CT scan Chest and Abdomen, multiple small intra pulmonary nodules found in right upper lobe and left lower lobe. Osteolytic lesion noted in body of C6, C7, T4 and T5 vertebra. (Figure-4) Few bilateral axillary lymph node noted. In liver well defined homogenously enhancing lesion noted in segment VI. (Figure-5) Histopathological study showed pleomorphic and atypical lymphoid cells with atypical mitotic figures, area of sclerosis and haemorrhage. (Figure-6) The Immunohistochemistry profile showed positivity for leukocyte common antigen (LCA), CD 45 and CD 20. Base on the histopathological and immunohistochemistry, a diagnosis of Non Hodgkin’s lymphoma was made than patient was referred to oncologist for further management.

Figure 1: X-ray skull with mandible (AP and Lateral view)

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DISCUSSION

Primary non-Hodgkin’s lymphoma (PNHL) involving extranodal sites accounts for 24–45% of the cases. The commonest site of extranodal disease is the gastrointestinal tract particularly the small intestine, stomach, and perianal region. The mandible accounts for only 0.6% of the isolated malignant non-Hodgkin’s lymphoma. PNHL demonstrates a preponderance for male patients in the fifth to seventh decades of life. In the era that preceded the introduction of highly active antiretroviral therapy (HAART) in 1996, NHL was diagnosed approximately 60 times more often in the HIV positive population than in the general population. Patients with HIV associated NHL more frequently present with advanced disease and/or extranodal disease than immunocompetent patients in up to 90% of patients. PNHL of the mandible can involve the canal by two mechanisms: (1) it can develop and arise in the bone, wrap around the mandibular canal and infiltrate it later; (2) it can arise from lymphoid tissue in the mandibular canal, grow with neural and perineural spread, and later slowly expand and destroy the surrounding bone.

Presentation symptoms of PBL include localized bone pain and sometimes the presence of a palpable mass. In the mandible, the main symptoms of PBL are pain, swelling, numbness, tooth mobility and cervical lymphadenopathy. Limb et al. suggested that the mean time to achieve a correct diagnosis of PBL is 8 months. Usually, biopsy is carried out in the non-healing extraction site after repeated treatment for non-responding odontogenic infection. Histopathology usually shows pleomorphic and atypical lymphoid cells. Lymphoma was suspected and immunohistochemistry was performed for the following markers: LCA (CD45), CD15, CD20, CD30, PAN-CK, EMA, ALK1 and CD3. The result was positive for LCA (CD45) and CD20. Therefore, based on the immunohistochemistry profile, a diagnosis of non-Hodgkin’s lymphoma was made.

When tissue diagnosis is confirmed as NHL of the mandible, determination must be made regarding orientation and spread of tumor. Overall assessment is essential to rule out nodal and visceral involvement. A CT scan can be carried out to look out for extranodal involvement. Laboratory studies are not specific, although elevated lactate dehydrogenase is observed as a poor diagnostic factor. Direct mandibular involvement by squamous cell carcinoma of the oral cavity occurs in two main patterns: erosive and infiltrative. Erosive involvement occurs when cortical bone recedes before a pushing tumor border. In this form of involvement, there is often a scalloped excavation of underlying medullary bone. In the infiltrative pattern of tumor involvement, cancer diffusely spreads throughout the cancellous, medullary bone. So on imaging differentiation is difficult, biopsy is required to confirm diagnosis. The 5-year survival rate for NHL of the maxillo-mandibular region is reported to be approximately 50%. Since the development of NHL is associated with a relative risk of death of 20-fold. HIV associated NHL caused up to 16% of all deaths attributable to AIDS.
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REFERENCES