

Challenges in the Diagnosis of Extranodal Non-Hodgkin's Lymphoma of the Base of Tongue: A Rare Case Report

Dika Wisnu Prabawa, and Renny A Rena*

Department of Internal Medicine, Faculty of Medicine
Universitas Udayana, Prof IGNG Ngoerah Hospital, Bali, Indonesia

*Corresponding author details: Renny A Rena

ABSTRACT

Introduction: Primary non-Hodgkin's lymphomas (NHLs) of the oral cavity, especially in the tongue, are extremely rare and account for 1% of all malignant tumors of the oral cavity, accounting for 7% of all primary Waldeyer's ring NHLs. Clinical features are non-specific ulcerative lesions that do not heal, making its diagnosis challenging. Fine-needle aspiration biopsy was helpful in the early diagnosis of the intraoral NHL involving a cervical lymph node. **Case Report:** we present a rare case of extranodal NHL of the base of tongue in a 55-year-old female who presented with a mass in the base of the tongue with cervical lymphadenopathy. The diagnosis was confirmed as primary NHL diffuse large B cell type by histopathological and immunohistochemistry results. **Conclusion:** A careful clinical evaluation supported by histopathological and radiologic investigations will help identify the disease early, resulting in a better prognosis.

Keywords: non-Hodgkin's lymphoma; tongue; diffuse large B cell lymphoma

INTRODUCTION

Non-Hodgkin's lymphoma (NHL) is the third most common group of malignant lesions in the oral cavity after squamous cell carcinoma (SCC) and salivary gland neoplasms. [1,2] Lymphomas that arise at the base of tongue are very rare; only about 7% of all NHL occur in the Waldeyer ring. [3,4] NHL at the base of tongue affects patients between the fourth and eighth decades of life, with a mean age between 50 and 60 years. [5,6] Diagnosis of lymphoma in the oral cavity, especially the base of tongue, can be complicated by the low level of clinical suspicion.

We report a case of extranodal NHL of the base of tongue in a 55-year-old female patient. Through this case report, it is hoped to increase the level of awareness of clinicians about NHL, especially NHL at the base of tongue, which is a rare case; early detection and early intervention can be carried out in patients.

CASE ILLUSTRATION

A 55-year-old woman came to the hospital complaining of a node appearing at the base of her tongue, which had been enlarging for the last five months. Complaints accompanied by fever that has come and gone in the previous five months, night sweats, and drastic weight loss in the last two months. The lump is painless, but the patient feels a node when the patient swallow's food, and the voice is said to be getting hoarse. In the last one month, a new lump appeared on the right and left neck, which increased. History of other illnesses such as hypertension and diabetes were denied. No family had similar complaints. Smoking and drinking alcohol habits are denied.

The patient was alert; vital signs were obtained with a blood pressure of 130/80 mmHg, temperature of 37.5°C, pulse rate of 98 times/minute, and respiration rate of 18 times/minute. On examination of the oropharynx, a mass appeared in the posterior 1/3 of the right side of the tongue, hyperemic, irregular border without pain with a

size of 1x2 cm, which seemed to be attached to the base of tongue and moved when the tongue was moved. On the neck, multiple masses of soft, solid consistency were palpable on the right and left sides, with the largest size being 3x6 cm. Abdomen and extremities were within normal limits.

The complete blood count was within normal limits. Electrolyte, renal, and liver function tests were also within normal limits. PA chest x-ray examination (Figure 1) showed suspicion of calcification in the lymph nodes. We performed an FNAB examination of the dextra colli nodules and showed a cytomorphological picture of the distribution of single atypical-pleomorphic large cells sized of undetermined origin among lymphoid tissue with a differential diagnosis of metastatic undifferentiated carcinoma (Schminckle type) and malignant lymphoma.

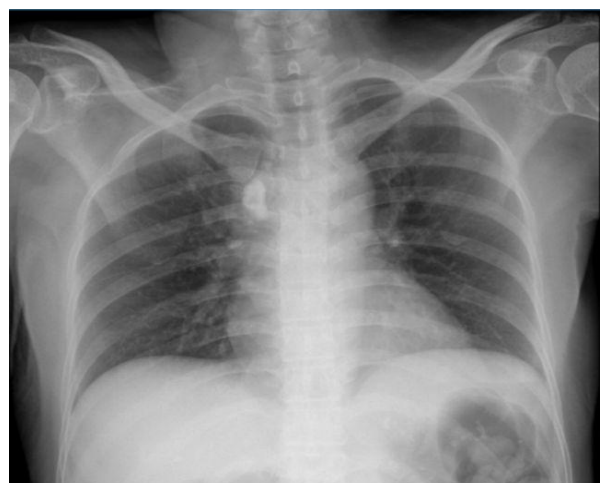


FIGURE 1: Chest x-ray showed suspicion of calcification in the lymph nodes.

From histopathological examination of the base of tongue (Figure 2), it was found that some showed the morphology of Non-Hodkin Lymphoma, diffuse, predominantly large cells.

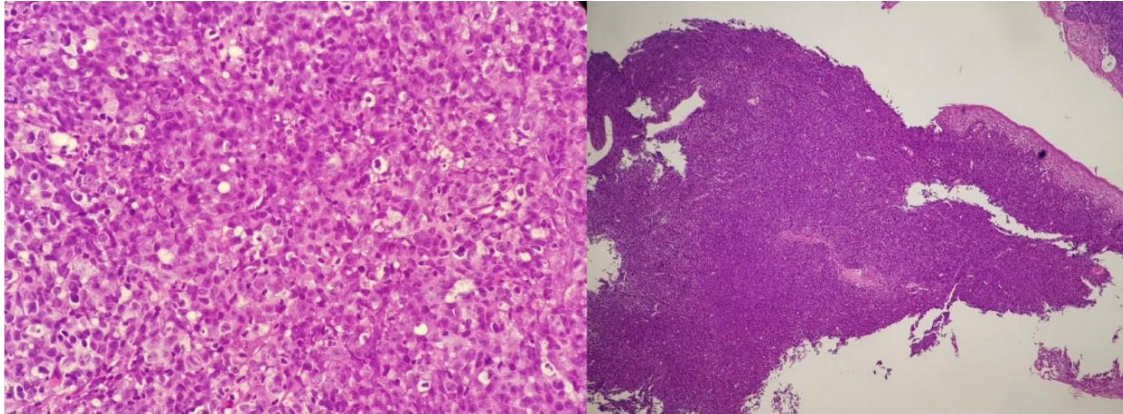


FIGURE 2: Histopathology of the base of tongue showing squeezing and crushed artefacts and necrosis. There is a diffuse infiltration of atypical, round cells with a slight scattering of mature and polymorphonuclear lymphocytes among them. The nuclei appear polymorphic round oval with mostly vesicular chromatin, consistent with the appearance of Centro blasts.

The abdominal ultrasound examination was within normal limits. The results of the MSCT examination of the mid-face and colli region with contrast showed a mass in the right tonsil, accompanied by multiple lymphadenopathy of the colli area, suprasternal, bilateral paratracheal, staging T2N3Mx without metastases in the brain parenchyma. (Figure 3).

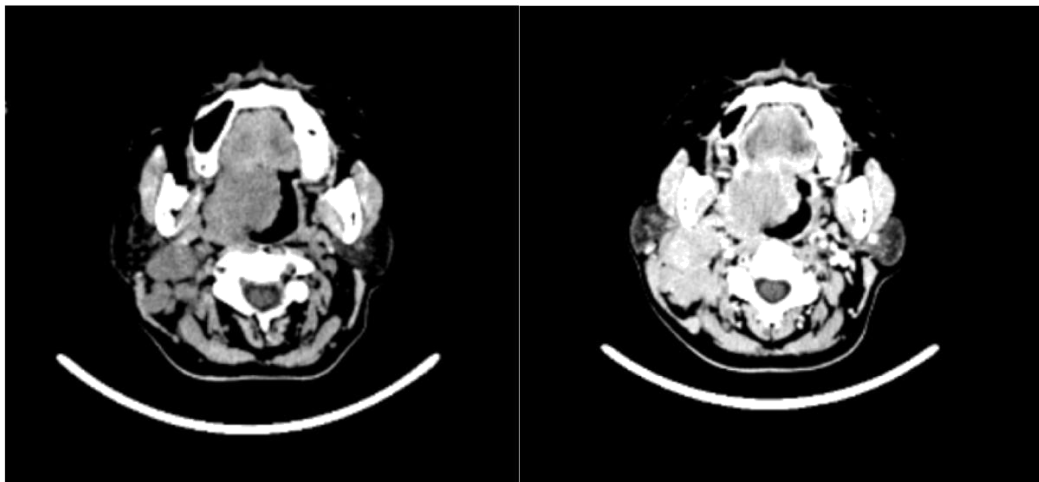


FIGURE 3: CT Scan of the midface showed compaction of the tonsils in lobulated shape, irregular edges, measuring 3.6x3.6x3.6 cm which extended to the oropharynx and had sharp contrast. Multiple enlarged lymph nodes were seen in the region colli, suprasternal, right and left para trachea with the largest diameter 3.8x6.0x6.9 cm, lobulated in shape pressing on the right parotid gland and right jugular vein.

Based on the clinical and supporting examinations that had been carried out, the patient was diagnosed with stage IIE non-Hodkin's lymphoma of the base of tongue, and chemotherapy was planned with three to six cycles of rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone (R-CHOP).

DISCUSSION

Approximately 3-5% of extra-nodal NHL arise in the oral and perioral areas, especially in the Waldeyer ring, which includes the palatal tonsils, nasopharyngeal lymphoid tissue, palate, and tongue base. Lymphoma that arises at the base of tongue is very rare; only 7% of all NHL occur in the Waldeyer ring. [8,9,10] In this case, a 55-year-old patient with NHL of the base of tongue is reported, which is a rare case.

Chromosomal translocations play an essential role in genome damage leading to lymphoid malignancies. Other risk factors such as viruses commonly associated with NHL

We performed the immunohistochemical examination, and the results showed that CD 20 was stained positive in most of the cell components that make up the lesion.

include Epstein-Barr virus, human T-cell lymphotropic virus-1 (HTLV), human herpesvirus-8 (HHV8), human type C retrovirus, HIV, SV40, and Helicobacter pylori.[11,12] Other risk factors for NHL are immunosuppression, ultraviolet radiation, autoimmune and chronic inflammatory disorders (rheumatoid arthritis, Sjogren's syndrome, and SLE), and occupational exposures (pesticides such as phenoxy acids, organophosphates, and organochlorines).[7,9]

Clinically, NHL in the base of tongue may present with local swelling, pain or discomfort, voice changes, dysphagia or odynophagia due to ulceration, and upper airway obstruction. Most cases of NHL of the base of tongue also present as peripheral lymphadenopathy. [14,15] Cervical lymphadenopathy is the most common head and neck presentation of NHL, characterized by multiple painless nodes. These lesions are not as hard as metastatic nodules and are not fixed to the skin or internal planes.[12] Some patients have later-stage symptoms, such as fever, night sweats, or significant weight loss. [5,7]

NHL in the oral cavity has a clinical picture similar to other common benign oral lesions.[11] Possible differential diagnoses include Squamous Cell Carcinoma (SCC), the most common malignancy of the base of the tongue, salivary gland malignancies (adenoid cystic carcinoma or mucoepidermoid carcinoma), and infection process such as tuberculosis. [12,14]

Defining the stages of oropharyngeal lymphoma and other nodal lymphomas also requires CT of the head, neck, chest, abdomen, and pelvis. [4,13] In the patient's case, the patient's stage is IIE due to the presence of a lymph node area (nodal) with an extra-nodal site at the base of tongue with the presence of lymphadenopathy and signs of unexplained weight loss unexplained fever, and night sweats. In this case, no metastases were found based on the CT image.

In general, therapy for NHL consists of specific treatment to suppress lymphoma cells and supportive treatment to improve the patient's general condition or to overcome the side effects of chemotherapy or radiotherapy. Specific therapy for NHL can be given through radiotherapy, chemotherapy, immunotherapy, and stem cell transplantation.[15]

Radiotherapy is still used primarily to maintain the remission phase after chemotherapy, but it is still used as the sole treatment modality in particular situations. Until now, the CHOP regimen (Cyclophosphamide, doxorubicin, vincristine, prednisone) is still the first-line chemotherapy regimen for diffuse large B cell lymphoma. The CHOP regimen will be combined with the monoclonal antibody rituximab, a chimeric monoclonal antibody directed against the CD20 antigen expressed by all B lymphocyte cells. Chemotherapy of three to six cycles of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) has been found to increase complete response rates and prolong survival, event-free, and overall survival in patients with DLBCL, without increasing clinically significant toxicity.[3,9] Other chemotherapy recommended as second line is the EPOCH regimen (Etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin) and combined with rituximab. However, from the development of therapy to date, the RCHOP chemotherapy regimen is more often used and has been proven more effective than other combination chemotherapy. [15,16] The patient was given first-line chemotherapy with the RCHOP regimen and followed the recommended therapy.

Treatment of primary extra-nodal lymphoma of the tongue is not clearly defined. Several studies have shown that NHL of the base of tongue responds to chemotherapy treatment followed by radiotherapy or combination therapy in patients with early-stage DLBCL. Treatment recommendations in cases of NHL of the base of tongue are generally identical for nodal and extra-nodal disease. In the patient's case, chemotherapy was planned with the RCHOP regimen for six series, and then monitoring and evaluation were carried out based on the target lesion's response to chemotherapy.

The prognosis of patients with extra-nodal NHL mainly depends on the tumor stage, degree of aggressiveness, and response to treatment. Ren et al.'s research on seven cases NHL of the base of tongue showed that three patients used the R-CHOP regimen and showed complete and partial remission results.[9]

CONCLUSION

NHL of the base of tongue is a very rare type of oral NHL manifestation. Early detection, diagnosis, and correct disease staging can result in complete recovery and better long-term survival. This case report presents a rare case of extra-nodal NHL of the base of tongue in a 55-year-old woman. In this case, careful clinical evaluation is essential, supported by histopathological and radiological examinations that will help identify the disease early.

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