

CASE REPORT

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# Extranodal natural killer T cell lymphoma of the oropharynx: case report



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## Abstract

**Background:** T cell lymphoma of the upper aerodigestive tract is a rare entity usually presenting as midfacial ulcerative lesion. It is extremely rare to present as non-healing erosive lesion of the oropharynx.

**Case presentation:** A 29-year-old female patient presented with odynophagia and trismus for several months. An “inflammatory” oropharyngeal ulcer was diagnosed and she was treated with repeated courses of antibiotics and corticosteroids without response. Recently, she had some blood stained saliva and vomited fresh blood. When seen, she had extensive painful ulcer eroding the right side of the soft palate and right tonsillar area. The ulcer had a whitish floor and ragged border without any tendency to heal. A second biopsy was taken and proved the lesion to be extranodal natural killer T cell lymphoma (ENKTL). The patient was treated with a modified SMILE (steroid, methotrexate, ifosfamide, L-asparaginase, and etoposide) protocol.

**Conclusions:** Extranodal natural killer T cell lymphoma can manifest first in the oropharynx. If left untreated it may lead to deep erosive lesion and major oral bleeding. SMILE chemotherapy protocol was used in our patient with good early response.

**Keywords:** Lymphoma, Extra-nodal, Palate, Ulcer

## Background

Extranodal natural killer T cell lymphoma-the nasal type (ENKTL-NT) usually presents as an erosive lesion of midfacial structures. Rarely, the oropharynx may be affected first in the form of indolent ulcerative lesion [1–3]. Persistent non-specific throat symptoms and vague systemic symptoms may be present for several months before diagnosis.

The aim of this report is to present a rare case of T cell lymphoma presenting as oropharyngeal ulcer in 29-year-old female patient. Clinical features, diagnostic workup, and treatment of this tumor are to be discussed.

## Ethics considerations

The local ethics committee approved the study. Written consent was obtained from the study participant and she was informed about the procedures to be done and the

expected result. Also, a written consent for study publication was obtained from her.

## Case presentation

A 29-year-old female patient without history of chronic illnesses presented to our hospital with persistent odynophagia of increasing severity for several months associated with significant weight loss. She had no nasal discharge, no epistaxis, no nasal regurgitation of liquids, and no nasal tone of voice. She had no ear problems. She gave history of intermittent fever reaching up to 38.5°, morning arthralgia and bone aches.

She received repeated courses of antibiotics and oral steroids without response. She had a histopathology report of a biopsy taken under local anesthesia from the lesion. Only nonspecific inflammatory granulation tissue was reported by the pathologist.

Two months before coming to our hospital, she started to have bleeding per mouth and sometimes she vomited fresh blood.

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On examination, she was pale and looked ill. She had moderate degree of trismus and through the limited mouth opening, we could see an extensive ulcer on the right side of the soft palate eroding the uvula and extending laterally to the palatoglossal fold and the tonsillar fossa on right side down to the lingual tonsil (Fig. 1).

With transnasal flexible endoscopy, we could see the ulcer involving the nasal side of the soft palate. Going down with the scope, we could see a blood clot adherent to the right side of the tongue base.

Neck examination was unremarkable except for enlarged mildly tender jugulodigastric lymph nodes on the right side.

Laboratory workup showed microcytic hypochromic anemia (Hb = 9 g/dl) with mild leucocytosis and absolute monocytosis, normal coagulation profile, high ESR (1st hour 74 and 2nd hour > 100) high C-reactive protein, high lactate dehydrogenase. Tests were negative for rheumatoid factor, antinuclear antibody, C-ANCA, P-ANCA, and anti-ds DNA antibodies. Hepatitis C virus antibodies and HIV markers were also negative.

Contrast-enhanced CT showed erosion of the soft palate on the right side and deep erosion of the right palatine tonsil (Figs. 2, 3).

A decision was made to take another biopsy from the lesion under local anesthesia. This time, biopsy showed atypical lymphoid cells with high anaplasia. Some tumor cells had angiocentric spread. With immunotyping, cells were positive for CD56, CD3, CD43, and CD8. Ki 67 was high 80%. Cells were negative for CD7 and CD4. This was consistent with angiocentric ENKTL.

We requested PET-CT scan as part of the lymphoma workup. Fortunately, it showed no metabolically active

lesions other than the oropharyngeal ulcer and the ipsilateral jugulodigastric lymph nodes. Following Ann Arbor Staging system, the patient was classified as stage IIE with B symptoms.

Patient was referred to the Medical Oncology department and a decision was made to start a modified SMILE (steroid, methotrexate, ifosfamide, L-asparaginase, and etoposide) protocol. While writing this report, patient already received 2 cycles of this protocol and she had less pain and no oral bleeding.

## Discussion

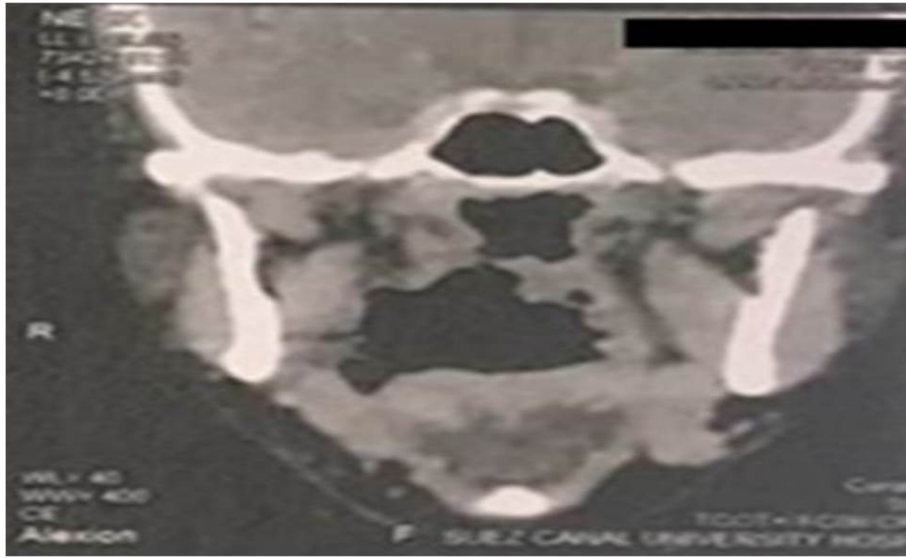
Differential diagnosis of oropharyngeal erosive lesions that fail to heal encompasses a myriad of pathologic entities: carcinomas, lymphomas, vasculitides, e.g., granulomatosis with polyangiitis and fungal granulomas. Diagnosis depends on biopsy taken from the appropriate site and of sufficient depth [4, 5].

The age of our patient and the clinical picture are not typical of squamous cell carcinoma (SCC) which is a disease of elderly. Smoking and alcohol are important risk factors in SCC. However, human papilloma virus positive tumors have been diagnosed in relatively young patients who are non-smokers [6]. Pharyngeal lymphoma of B cell type is not uncommon and it usually presents as tonsillar enlargement or as a nasopharyngeal mass [7].

ENKTL is a rare tumor that presents mainly as ulcerative lesion of the nose and paranasal sinuses. Rarely, it manifests as an oropharyngeal lesion [8, 9]. Diagnosis needs to be expeditious to prevent erosion of the palate and tonsillar area with a possibility of massive bleeding from the lateral pharyngeal wall or tongue base.



**Fig. 1** Oropharynx of the patient: the lesion eroded the right tonsil and the right side of the soft palate and the uvula. Patient had trismus



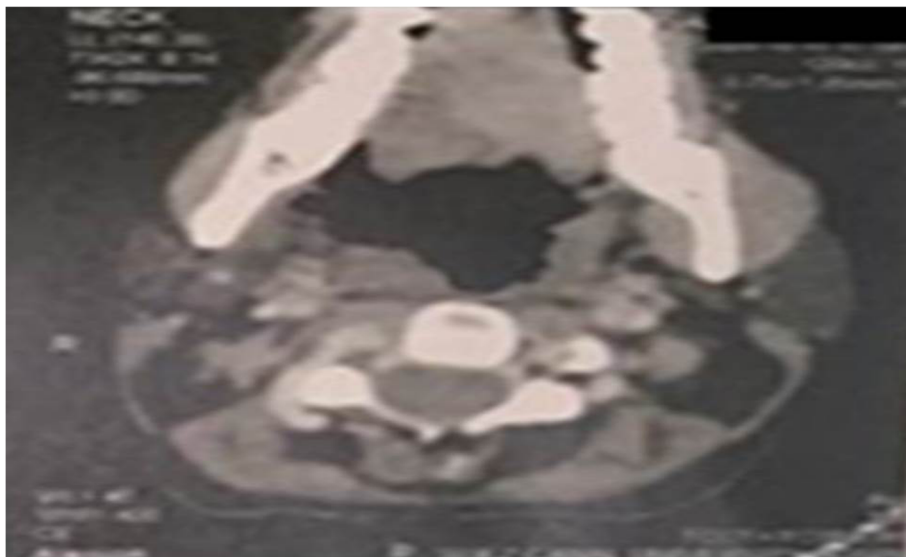
**Fig. 2** Coronal CT showing the oropharyngeal defect in the right tonsillar area extending to the tongue base and eroding the palate

In the past, treatment of ENKTL-NT was mainly radiotherapy to stop erosion of facial soft tissue and facial bones. The tumor is known to be resistant to anthracycline based chemotherapy regimen [10]. Expression of P-glycoprotein in this tumor is responsible for its multidrug resistance [11]. Therefore, CHOP protocols (cyclophosphamide, doxorubicin, vincristine, and prednisone) used for B cell lymphoma are usually not effective in this tumor [12].

We sent our patient to Medical Oncology department to receive the SMILE chemotherapy protocol. This is used now in many centers as the standard protocol for patients with advanced ENKTL-NT [13]. L-Asparaginase-

based regimens give outstanding response in these patients because tumor cells cannot synthesize L-asparagine and die when their stores of this amino acid are depleted by L-asparaginase.

Phase II of SMILE chemotherapy regimen showed an excellent anti-tumor activity in ENKTL-NT, with an overall response rate of 80% and a 1-year survival rate of 55% [14]. Cycles should be repeated every 28 days followed by re-staging after 2 cycles. Patients in whom incomplete response is obtained may benefit from involved field radiotherapy (IFRT) before proceeding to complete 6 courses [15].



**Fig. 3** Axial CT showing the oropharyngeal defect involving the right tonsillar area, palatoglossal fold, and extending toward the tongue base

## Conclusions

Extranodal natural T cell lymphoma can manifest first in the oropharynx. If left untreated, it may lead to deep erosive lesion and major oral bleeding. SMILE chemotherapy protocol was used in our patient with good early response.

## Abbreviations

NK: Natural killer; ENKTL-NT: Extranodal NK/T cell lymphoma; SLE: Systemic lupus erythematosus; CRP: C-reactive protein; LDH: Lactate dehydrogenase; ANCA: Antineutrophil cytoplasmic antibodies; HIV: Human immunodeficiency virus; HCV: Hepatitis C virus; CECT: Contrast enhanced computed tomography; MRI: Magnetic resonance imaging; PET-CT: Positron emission tomography-computed tomography; ICU: Intensive care unit; NHL: Non-Hodgkin lymphoma; HC: Hematopoietic cancers; PS: Performance status; IFRT: Involved field radiotherapy

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None

## Authors' contributions

AS revised the clinical data and plan of management, IH analyzed and interpreted the patient data evaluation, MH followed up patient and collected the patient data, MR is a major contributor in writing the manuscript. All authors read and approved the final manuscript.

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## Availability of data and materials

Not applicable

## Declarations

### Ethics approval and consent to participate

Study approved by the local ethics committee (Faculty of Medicine Suez Canal University under the number 816 (date of approval—April 2021). Written consent was obtained from study participant and she has been informed about the procedures to be done and the expected results.

### Consent for publication

Written consent for study publication was obtained from the study participant.

### Competing interests

The authors declare that they have no competing interests.

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