

NK/T-Cell Lymphoma on the Upper Lip

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Abstract

Extra-nodal natural killer/T-cell lymphoma, nasal type is a distinct entity of non-Hodgkin's lymphoma. This case report describes a rare presentation of NK/T cell lymphoma on the upper lip. However, lung metastasis was confirmed by CT scans after 2 cycles of chemotherapy. The patient then died of infection and multiple organ failure. This case indicates that Oral Medicine specialists should be aware of this disease, and multiple diagnostic techniques should be applied in order to make an accurate and timely diagnosis.

Keywords: Lip; NK/T-cell lymphoma

Introduction

Extra-nodal natural killer (NK)/T-cell lymphoma, nasal-type (ENKTL) is an uncommon type of lymphoma with involvement of extra-nasal sites, such as visceral organs and soft tissues, as the primary manifestations [1]. Reports of localized NK/T cell lymphoma on the upper lip were extremely rare.

Case Report

A 42-year-old man was referred to our clinic by his physician in November 2014. He had been suffering from progressive ulceration of the upper lip with pain and itching for four months. The patient had a history of second- to third-degree fire burn ten years ago, **involving 80% total body surface area**. He had also been taking benazepril pills to control high blood pressure for ten years.

Physical examination revealed an ill-defined, 3.5 cm x 2.5 cm ulcerative lump with irregular and raised border on the upper lip (Figure 1).

No other oral or extra-oral lesion was observed. Routine laboratory tests, including whole blood cell count, coagulation test, urine test, as well as liver and kidney function panels were normal. Tests for human immunodeficiency virus and syphilis were negative.

An incisional biopsy was taken from the lump. Routine histopathological examination revealed dense infiltration of medium-sized lymphocyte-like cells (Figure 2).



Figure 1: Clinical manifestation.

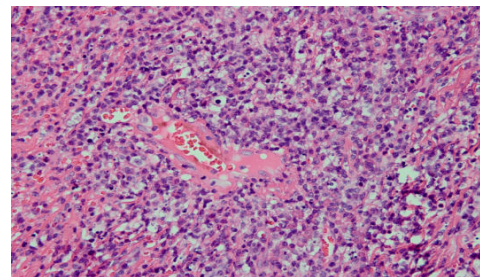


Figure 2: HE 400.

Immunohistochemistry was performed and the findings were as follows: CD56 (±)CD20 (-), Granzyme B (+), MPO (-), CD4 (-), TdT (-), CD34 (-), CD123 (-), and CD3ε (+) (Figure 3).

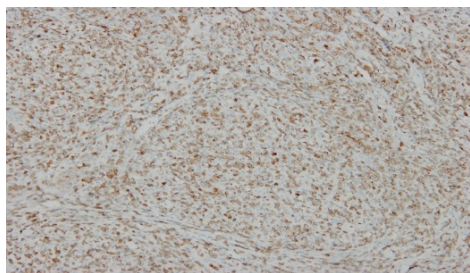


Figure 3: CD3 epsilon 200.

The positive rate of Ki67 was 40%. *In situ* hybridization (ISH) revealed positive nuclear staining of EBER 1/2 in the lymphocyte-like cells (Figure 4).

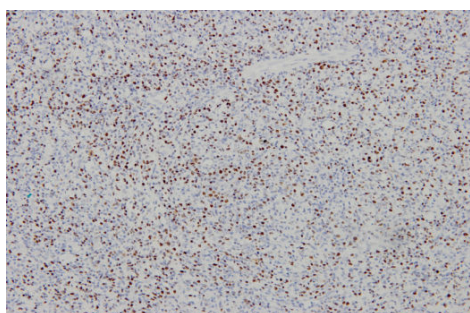


Figure 4: EBER 400.

Following bone marrow aspiration examination and contrast-enhanced CT scans of the nasopharynx, neck, chest and abdomen didn't reveal any pathological change.

The lesion enlarged quickly after the biopsy. The size of the lump was around 5 cm X 3 cm at the second review after two weeks.

Based on the histopathological findings, a diagnosis of ENKTL (invasive, WHO) was made, although ulceration of the upper lip as the primary clinical manifestation of this disease was rarely reported.

The patient received the THP-COP regimen containing pirarubicin, cyclophosphamide, vincristine and prednisolone in another hospital.

After two cycles of THP-COP chemotherapy, the size of the lesion didn't change. Furthermore, lung metastasis and mediastinal lymph node involvement was confirmed by CT scans. The treatment plan was then switched to a regimen with gemcitabine, pegaspargase and oxaliplatin. Partial remission was achieved after two cycles of chemotherapy. However, the patient died of infection and multiple organ failure in May 2015, six months after the diagnosis was confirmed.

Discussion

ENKTL is a distinct entity of Non-Hodgkin's lymphoma, which is extremely rare in Europe and North America but more prevalent in Asia and Latin America. Epstein-Barr virus (EBV) has been proved to be involved in the pathogenesis of ENKTL. Clinically, NK/T-cell lymphoma is divided into nasal and extra-nasal (non-nasal) subgroups according to the sites of the primary lesion. Clinical manifestations of nasal NK/T-cell lymphomas include nasal obstruction, epistaxis and palatal perforation. In the current case, none of the above symptoms was observed at the first or any of the following visits. The involvement of nasal cavity was further ruled out by additional examination of contrast-enhanced CT. The patient chose not to do PET/CT due to lack of funds.

For non-nasal lymphomas of any stage, systemic chemotherapy with involved-field radiotherapy is preferred [2]. NK/T-cell lymphomas of the non-nasal type are more aggressive than their nasal counterparts and usually have poor prognosis.

We presented a rare case of ulcerative lump on the lip as the primary manifestation of ENKTL. This case indicates that Oral Medicine specialists should be aware of this disease, and multiple diagnostic techniques should be applied in order to make a correct diagnosis.

Funding Sources

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