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Case Report

Extranasal T/NK lymphoma with a fatal outcome: A case report

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ABSTRACT

Extranodal T/NK lymphoma (LTNKEN) is a rare and aggressive form of non-Hodgkin lymphoma of high-grade malignancy. A distinction is made between the nasal forms, characterized by primary involvement of the nasal cavity, and the extranasal forms. Primary skin involvement is highly rare. Diagnosis is based on immunohistochemical studies. Treatment includes chemotherapy and radiotherapy. This type of lymphoma has a poor prognosis even with treatment. Herein, we report a Moroccan case of cutaneous T/NK lymphoma in a young patient with a primary cutaneous localization of fatal evolution.

Key words: Lymphoma; Extranodal lymphoma; T/NK lymphoma; Non-hodgkin lymphoma

INTRODUCTION

Extranodal T/NK lymphoma (ENTNHL) is a high-grade, non-Hodgkin, EBV-induced lymphoma [1]. It is a rare lymphoma that affects, in particular, the populations of East Asia and southern South America. There are two types: the nasal forms with primary involvement of the oropharyngeal sphere and the extranasal forms, also known as the nasal type, in which skin involvement is the most common location in terms of frequency, reported in 10% of cases [2]. The prognosis of these lymphomas remains poor because of late diagnosis and an aggressive evolution, complicated by visceral involvement. Herein, we describe a case of cutaneous, nasal-type LTNKEN revealed by a primary cutaneous localization with a fatal outcome in a young patient.

CASE REPORT

A 27-year-old, non-immunocompromised patient presented for four months with a rapidly progressive skin nodule of the leg in a context of fever, night sweats, and a profound alteration in the general condition.

An examination revealed a 20-cm long, violet, erythematous, necrotic skin tumor on the outer surface of the right leg (Fig. 1) associated with a mass of homolateral inguinal adenopathies and hepatosplenomegaly. A workup showed an inflammatory syndrome, functional renal failure, hepatic cytolysis, hypertriglyceridemia, hypercholesterolemia, and pancytopenia. A diagnosis of macrophagic activation syndrome (MAS) was retained.

A skin biopsy revealed a dense and atypical lymphocytic infiltrate (Figs. 2a and 2b). Immunohistochemistry revealed the expression of NK cell markers and cytotoxic molecules (TIA-1, granzyme B, and perforin), as well as the presence of Epstein–Barr virus (EBV) RNA in the tumor cells.

A cervico-thoraco-abdomino-pelvic CT scan revealed a free cavum and the integrity of the laryngo-pharyngeal carrefour. A homogeneous hepatosplenomegaly was also noted. There were no visceral metastases.

The diagnosis was extranasal ENKTL complicated by MAS. Systemic corticosteroid therapy was initiated

How to cite this article: Ammar N, Benzekri L, Omari M, Znati K, Senouci K. Extranasal T/NK lymphoma with a fatal outcome: A case report. Our Dermatol Online. 2023;14(2):191-193.

Submission: 21.08.2022; **Acceptance:** 06.11.2022

DOI: 10.7241/ourd.20232.14

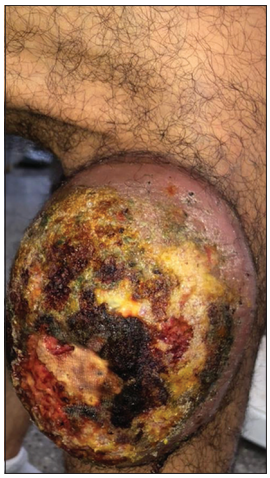

Figure 1: Erythematous and violaceous, nodular tumor, necrotic on the external surface of the left leg.

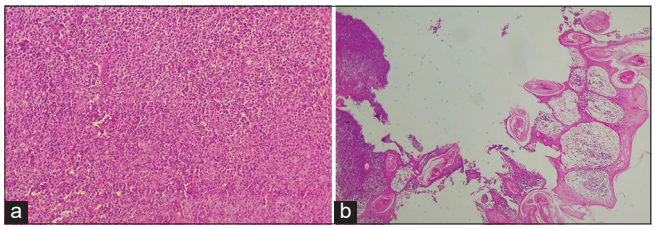

Figure 2: (a and b) Standard histology shows a malignant proliferation of round, spindle-shaped cells of lymphoid appearance with an angiocentric arrangement.

as an emergency, yet the patient died by multivisceral failure.

DISCUSSION

LTNKEN accounts for 5–18% of non-Hodgkin lymphomas [2]. It occurs mainly in Southeast Asia and Central and South American populations, and is rare in Europe and the U.S. A male predominance is observed. It is a high-grade, malignant lymphoma with a poor prognosis [4].

Two entities of extranodal T/NK lymphoma are distinguished according to the initial location of the lesions. The nasal forms (nasal LTNKEN), which represent 80% of cases, begin in the nasal cavity or adjacent structures (orbits, oral cavity, sinuses, upper aerodigestive tract). The extranasal forms, known as the nasal type, represent 20% of cases. Skin involvement may be related to the metastatic extension of nasal LTNKEN to the skin or may be primary in the case of extranasal LTNKEN. Primary skin involvement is reported in 10–26% of cases of LTNKEN, the spleen and liver are the primary sites in 5% of cases, while initial lymph node involvement is exceptional [5].

A longer diagnostic delay and a different visceral involvement explain the poorer prognosis of the extranasal forms. Because of the structures invaded, the nasal forms are more symptomatic at an early stage (nasal obstruction, chronic rhinitis and sinusitis, epistaxis) and are, therefore, generally diagnosed at the localized stage, unlike the extranasal forms.

Skin involvement usually presents as ulcerated tumors, yet also as vasculitis, panniculitis, or cellulitis.

Histology reveals a dense infiltrate of lymphocytes of variable size associated with histiocytes, plasma cells, and eosinophils. This infiltrate is both dermohypodermal and epidermotropic, and frequently presents aspects of necrosis with angiocentricity and angiodestruction [6].

On immunohistochemistry, tumor cells typically express CD2, CD56, and cytotoxic proteins. There is intracytoplasmic CD3E yet no membrane expression of CD3. LMP-1 protein expression is inconsistent, but EBV may be detected by *in situ* hybridization in difficult cases [6].

The prognosis of these lymphomas is poor, particularly in the extranasal forms, due to a longer diagnostic delay and more extensive visceral involvement. In advanced stages, treatment is based on multidrug therapy with L-asparaginase, followed by autologous or even allogeneic hematopoietic stem cell transplantation [7,8].

CONCLUSION

The diagnosis of LTNKEN remains difficult and requires an anatomopathological and clinical confrontation. The cutaneous involvement in this type of lymphoma is often atypical and infrequent, which may lead to a diagnostic delay. The prognosis is generally poor and the diagnosis must be suspected early.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be

published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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Source of Support: This article has no funding source,

Conflict of Interest: The authors have no conflict of interest to declare.