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Case Report

Extranodal NK/T-cell lymphoma, nasal type: a case report

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ABSTRACT

Extranodal NK/T-cell lymphoma, nasal type (ENKTL-NT), is a rare and aggressive malignancy associated with Epstein-Barr virus (EBV). Its clinical presentation includes midfacial destruction, ulceration, and nasal obstruction, often leading to misdiagnosis. A 43-year-old male presented with a rapidly progressive nasal lesion. Clinical evaluation, imaging, and histopathological analysis with immunohistochemistry confirmed ENKTL-NT. Treatment includes radiotherapy and chemotherapy, improving survival rates, though prognosis remains poor in advanced cases. Early diagnosis is crucial for better outcomes.

Keywords: NK/T cell lymphoma, EN-NK/T-NT, ENKTL, Nasal obstruction, Epstein Barr virus

INTRODUCTION

Extranodal NK/T-cell lymphoma, nasal type (ENKTL-NT) is a rare and aggressive neoplasia with a higher prevalence in Asia and Latin America.[6] It originates from NK cells or, to a lesser extent, T lymphocytes, and is closely associated with Epstein-Barr virus (EBV) infection. Its clinical presentation usually includes destructive lesions in the midface region, which can make its differential diagnosis with inflammatory or infectious diseases difficult[1]. The diagnosis of ENKTL-NT is based on characteristic histopathological findings, such as atypical lymphoid infiltrates with angiocentric and angiodestructive arrangement, and coagulative necrosis as well as on the expression of immunohistochemical markers such as CD2, CD56 and cytotoxic proteins.[1,2] In therapeutic terms, ENKTL-NT is highly radiosensitive, so radiotherapy is the treatment of choice in early stages; however, its exclusive use is associated with high recurrence rates. Therefore, a combined approach with chemotherapy is recommended.[1] This article presents a clinical case of ENKTL-NT, describing its clinical manifestation and diagnosis, and comparing it with the available literature. It seeks to provide relevant information on this pathology of difficult diagnosis and aggressive evolution, highlighting the importance of a multidisciplinary approach for its timely management.

CASE REPORT

A 43-year-old male patient presented with a nodular lesion on the right nasal ala, which progressively increased in size of up to double the volume in a period of 10 days within 2 months prior to his assessment. He reports treatment with the administration of an unspecified antibiotic for 7 days, without improvement. Subsequently, it progresses with discharge of serous exudate and subsequent evolution to scabs, accompanied by intense pain (VAS 10/10). He mentions improvement after the administration of non-steroidal anti-inflammatory drugs and intramuscular dexamethasone. He is consulted in the dermatology service, where the patient is found to have a localized dermatosis that affects the head and face at the level of the nasal dorsum and right nasal ala, consisting of an infiltrated plaque of approximately 5×5 cm with erythema, blood crusts and some areas with necrosis, with irregular, well-defined edges. The ipsilateral nasal mucosa is seen with edema, erythema and some blood crusts, as well as the discharge of abundant serous material, as well as a stomatosis affecting the hard palate, consisting of two ulcers of approximately 3×3 cm and 1×1 cm, both with a

dirty bottom with abundant fibrin content. The rest of the skin and appendages have palpable right preauricular adenopathy, of medium consistency, mobile, not adhered to deep planes, of approximately 2×2 cm. A diagnosis of a probable lymphoproliferative process is integrated, to rule out nasal T/NK cell lymphoma. The patient is subsequently admitted to hospitalization. A computed tomography of the head is performed, with the finding of an infiltrative-looking lesion in soft tissues of the nasal region, extending to the right periorbital region, upper labial region and nasal cavity losing interface with the lower turbinates, which show bone erosion. The lesion presents heterogeneous enhancement in contrast phase, some irregular hypodense images are observed, and some internal vascular structures are evident.

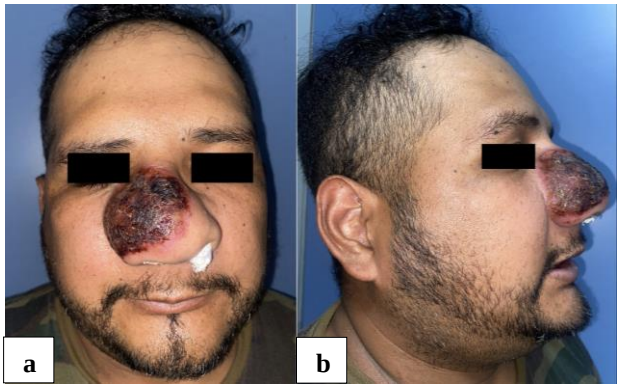

Figure 1 (a and b): Clinical images of patient with a histopathological diagnosis compatible with NK/T cell lymphoma (front and lateral view).

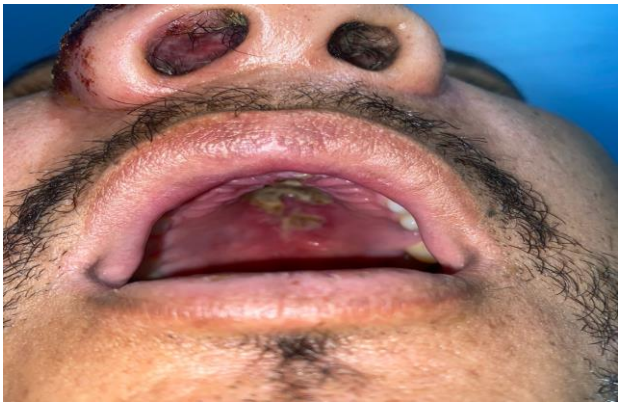

Figure 2: Necrotic ulcer in the palate associated with NK/ T cell lymphoma.

In addition, involvement of the right tear sac is demonstrated, without ruling out the possibility of infiltration into the tear duct in its proximal third. A biopsy was taken, reporting a stratum corneum with areas of necrosis and serous deposits with cellular remains corresponding to scab. Mild irregular acanthosis with areas of ulceration. From the papillary dermis to the deep reticular dermis where the cut reaches, an atypical inflammatory infiltrate is observed with an interstitial and perivascular arrangement that is composed of lymphoid cells of different sizes with pleomorphic nuclei, some kidney-shaped, others rounded, with dispersed chromatin and formation of perinuclear halos that are arranged in a hemorrhagic stroma with destruction of the vascular walls, edema and areas with Azzopardi phenomenon, with a histopathological diagnosis compatible with NK T lymphoma immunophenotype CD2 +, CD3 +, perforin +, CD30 +, CD56 +, with a proliferation index of 60%, BCL2 negative, BCL6 negative. In this case, in situ hybridization was not performed to properly classify the lesion, because the hospital does not have such a resource.

DISCUSSION

ENKTCL is the most common subtype of NK/T-cell lymphoma in Asia and Latin America, especially Guatemala and Brazil.[6] Extranodal NK/T-cell lymphoma, nasal type, is considered an aggressive subtype of T-cell lymphoma with an estimated overall survival of less than 2 years from diagnosis.[8] It has a higher incidence in men, with a mean age of diagnosis of 50 years.[6] Certain strains or mutations of EBV can preferentially infect NK/T cells and evade immune surveillance, which could explain the geographic distribution of the disease.[7]

General manifestations of non-T-cell T lymphocyte disease include signs and symptoms located mainly in the face and neck region: facial pain, diplopia, visual impairment, protrusion of the eyeball, eyelid ptosis, pupillary abnormalities, nasal obstruction, refractory sinusitis, velopalatine motor disturbances, cranial nerve neuropathies, infraorbital and intrasinusal masses. Other associations consist of respiratory failure and enlargement of the liver and spleen.[4] The classic clinical presentation usually involves palatal perforation.[3] In the case of our patient, from the time of evaluation he was found to have clear involvement of the hard palate.

Patients with ENKL are divided into three risk groups based on four criteria: age >60 years, tumor stage III or IV, involvement of distant lymph nodes and non-nasal type disease. Three-year survival rates are 81%, 62%, and 25% for patients with zero, one, and two or more risk factors, respectively[5]. Most patients with nasal ENKTCL-NT present with stage I or II according to the Ann Arbor classification. However, 10-20% of cases progress to disseminated disease, involving the skin, gastrointestinal tract, lymph nodes, or central nervous system.[7]

It typically causes vascular damage and tissue destruction. Bone marrow involvement and B symptoms (i.e., fever, night sweats, weight loss) are seen in approximately 10-35% of patients, respectively.[3] It is important to recognize the existence of highly aggressive lymphomas and classify them appropriately due to the rarity of cases. Within the clinical evaluation, it is important to assess for "B" symptoms and the physical examination should include visual inspection of the nasal and oral cavities and palpation of the buccal and gingival mucous membranes,

floor of the mouth, tongue, tonsillar fossae, palate and posterior pharyngeal wall.

Regarding the pre-diagnostic and staging approach, de Oliveira et al recommends the diagnosis of ENKTCL is based on clinical and histopathological characteristics and the expression of standard cytotoxic molecules and CD56.[6] Quantitative EBV DNA can indicate tumor burden, with elevations associated with a worse prognosis. Diagnostic delays are common due to inconclusive initial biopsies due to necrosis.[5] Imaging studies such as CT are also recommended to evaluate the nasal cavity, hard palate, anterior fossa and nasopharynx.

More recent therapeutic options include radiotherapy, chemotherapy or a combination of both.[1] Since ENKTL-NT is highly radiosensitive, radiotherapy is generally used as a first-line treatment, however, its use as monotherapy is associated with a high rate of relapses, both local and distant. Therefore, a combined therapeutic approach is recommended, based on the CHOP regimen (cyclophosphamide, doxorubicin, vincristine, prednisone), which has been shown to increase the 5-year survival rate from 20% to 80%.[4] Other treatment options include immunotherapy and autologous or allogeneic hematopoietic stem cell transplantation.[1]

CONCLUSION

ENKTL-NT is a rare and aggressive neoplasia and its diagnosis is challenging due to its nonspecific clinical presentation and its similarity to inflammatory or infectious processes. Its rapid and destructive evolution highlights the importance of an early diagnosis, which should be based on a detailed clinical evaluation, imaging studies and histopathological confirmation with immunohistochemistry. This case highlights the importance of a comprehensive and multidisciplinary approach for the identification and management of this entity. Although ENKTL-NT is highly radiosensitive, the use of radiotherapy alone is associated with high recurrence rates, justifying the need for combined therapeutic strategies. Modern L-asparaginase-based regimens have been shown to improve survival compared to the traditional CHOP regimen, although their access and toxicity remain challenges to consider. Given the guarded prognosis of this disease, it is essential to continue researching new therapeutic strategies, including immunotherapy and targeted therapies, to improve outcomes in patients with advanced disease. Furthermore, this case underlines the relevance of clinical suspicion and early recognition of the characteristic signs of ENKTL-NT, which can significantly impact the evolution and prognosis of patients.

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