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NK/T cell lymphoma, nasal-type (rare, rapidly evolving, mutilating, and highly lethal lymphoid neoplasm): a case report

Linfoma de células NK/T tipo nasal (neoplasia linfoide rara, de rápida evolução, mutilante e de alta letalidade): relato de caso

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ABSTRACT

NK/T cell lymphoma is a rare, aggressive, non-Hodgkin tumor that is closely related to the Epstein-Barr virus. It has a poor prognosis and poor response to treatments. We report the case of a 91-year-old woman with a history of injury in the nasal region for three months. Histological study showed diffuse infiltration of the dermis by small and atypical lymphoid cells and positive immunohistochemistry for Ki-67, CD30, and CD3 (cytoplasmic). Due to the fast growth of the lesion and the high morbidity of the neoplasm, the patient was referred to the hospital for assistance but died before starting treatment.

Keywords: Epstein-Barr Virus Infections; Lymphocytes; Medical oncology; Extranodal T-NK Cell Lymphoma; Lethal Midline Granuloma

RESUMO

O linfoma de células NK/T é um tumor não Hodgkin, raro, muito agressivo e intimamente relacionado ao vírus Epstein-Barr (EBV). Possui prognóstico ruim e resposta pobre aos tratamentos. Trata-se de uma paciente feminina, 91 anos, com história de lesão em região nasal há três meses. Estudo histológico evidenciou infiltração difusa da derme por células linfoides pequenas e atípicas e imuno-histoquímica positiva para Ki-67, CD30 e CD3 (citoplasmático). Devido ao rápido crescimento da lesão e à alta morbidade da neoplasia, a paciente foi encaminhada para internação para medidas de suporte, mas evoluiu para óbito antes do início do tratamento.

Palavras-chave: *Infecções por Vírus Epstein-Barr; Linfócitos; Oncologia; Linfoma Extranodal de Células T-NK; Granuloma Letal da Linha Média*

Case report

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INTRODUCTION

NK/T cell lymphoma is a non-Hodgkin tumor commonly described in Asia and Latin America. It is rare (about 1% of cutaneous T-cell lymphomas), very aggressive, and closely related to the Epstein-Barr virus[1,2,3,4] (the infection can happen before or simultaneously with the tumor genesis).[2] The precursor cell has lymphoid lineage, and in 95% of cases, it comes from the NK cell (cytolytic function).[2]

It constitutes the group of non-mycosis fungoid/non-Sézary syndrome cutaneous lymphomas (10% of cases),[4] with a rapid, mutilating, and highly lethal evolution (five-year survival rate <5%).[1]

The tumor is divided into non-nasal and nasal subtypes (former lethal midline granuloma[2,3,4,5]). It affects the nasal or nasopharyngeal region in 80% of cases.[2,4] However, it can also involve the aerodigestive tract,[4] testicles,[5] muscles, and the uterus, or evolve contiguously to the orbit, salivary glands, and paranasal sinuses. Many non-nasal cases probably had their subclinical onset in the nose (primary site).[2]

It has a poor prognosis and poor response to treatments.

CASE REPORT

A 91-year-old woman, white, hypertensive, presented a history of a nasal lesion for three months and weight loss (5 kg in 30 days). An erythematous and edematous lesion was found in the left nasal region, with local ulceration and an area of necrosis, at the initial dermatological examination. The diagnostic hypotheses of invasive squamous cell carcinoma (SCC), NK/T cell lymphoma, leishmaniasis, and mucormycosis were raised. The investigation was then started.

One week after the initial evaluation, the lesion worsened considerably in size and appearance, and antibiotic therapy was then introduced. Laboratory tests showed hypochromic and microcytic anemia, leukocytosis, thrombocytosis, and increased ESR and CRP (28 and 138, respectively). Cranial tomography revealed an area of dermal ulceration from the left nasal/malar region to the adjacent bone limits without signs of invasion, chronic osteomyelitis, or organized collections. An anatomopathological study (Figures 3 and 4) showed diffuse infiltration of the dermis by small and atypical lymphoid cells. Immunohistochemistry (Figures 5 and 6) was positive for Ki-67 (estimated at 80%), CD30, and CD3 (cytoplasmic), confirming the diagnosis of high-grade (CD30+) T lymphoma (CD3e+) or NK/T lymphoma nasal-type.

Due to the rapid evolution of the neoplasm (Figures 1 and 2) and the patient's clinical worsening (decrease in general condition, adynamia, difficulty swallowing), she was referred to the hospital for support measures and palliative care, under the supervision of the Dermatology, Clinical Oncohematology, and Head and Neck Surgery (HNC) teams. The patient died on the fifth day of hospitalization.

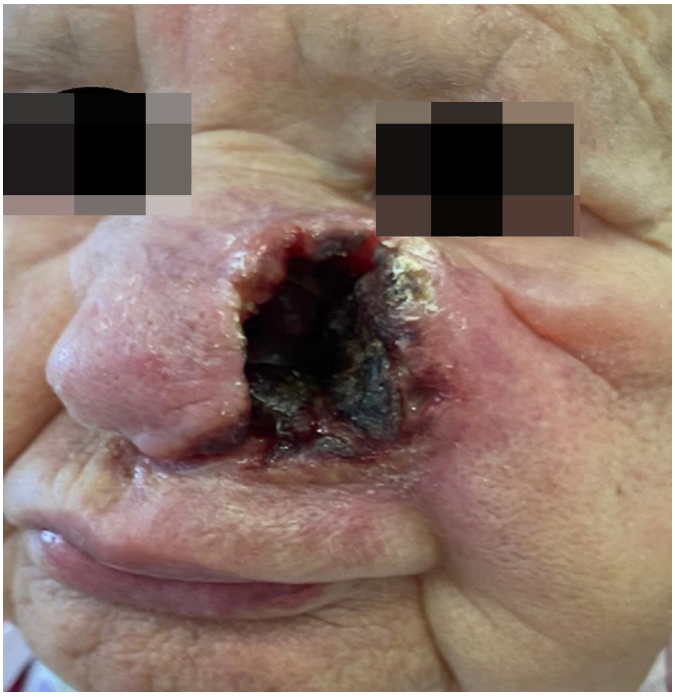

Figure 1: INITIAL EVALUATION (lesion with 4 cm in the largest diameter): ulcer with erythematous and infiltrated edges and background with hematic crust and necrotic material in the left nasal region. Perilesional erythema and edema. Involvement of a small portion of the nose on the left (nasal wall and ala)

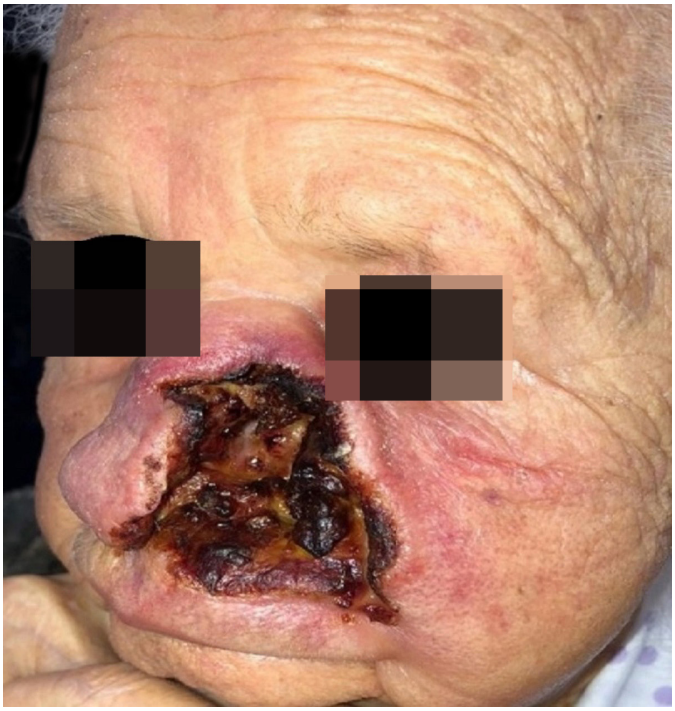

Figure 2: EVALUATION AFTER ONE WEEK (lesion with 7 cm in the largest diameter): ulceration in the left nasal and malar regions, with infiltration of edges and bottom covered by fibrinonecrotic material. Perilesional edema and erythema. Involvement of the entire left nasal wall and ala and part of the nasal tip, causing disfigurement of the central portion of the face

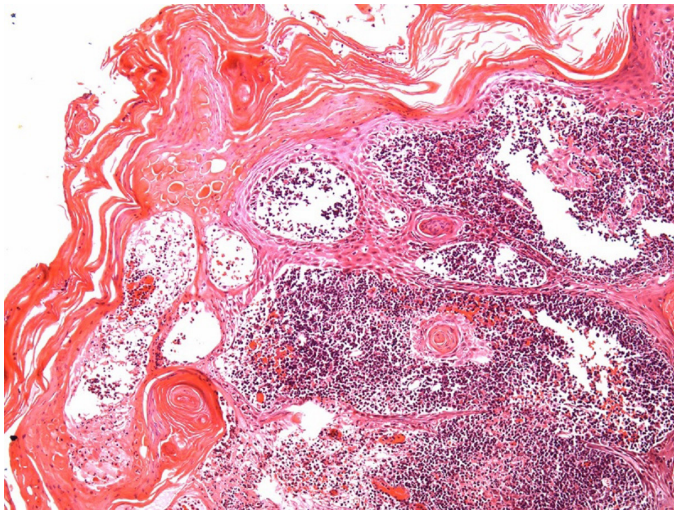

Figure 3: Hematoxylin & Eosin, 100x - Pseudoepitheliomatous hyperplasia, hyperkeratosis, and corneal pseudocysts. Dense lymphocytic infiltrate in the dermis

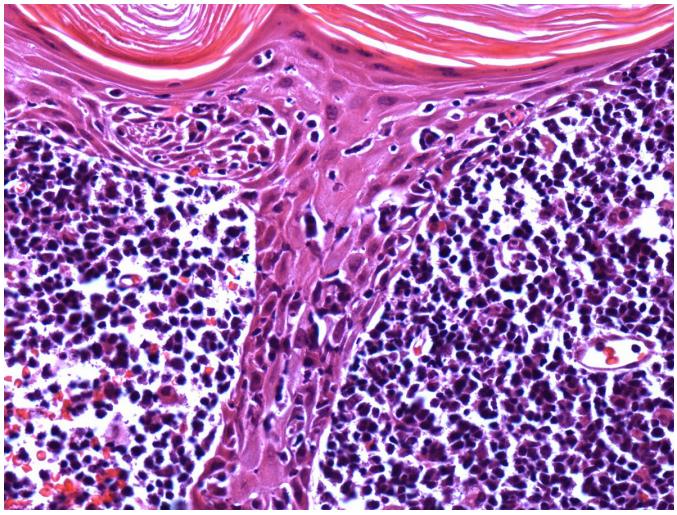

Figure 4: Hematoxylin & Eosin, 400x - Epidermotropism and microabscesses. Atypical lymphocytes with hyperchromatic nuclei. An intimate relationship with vessels

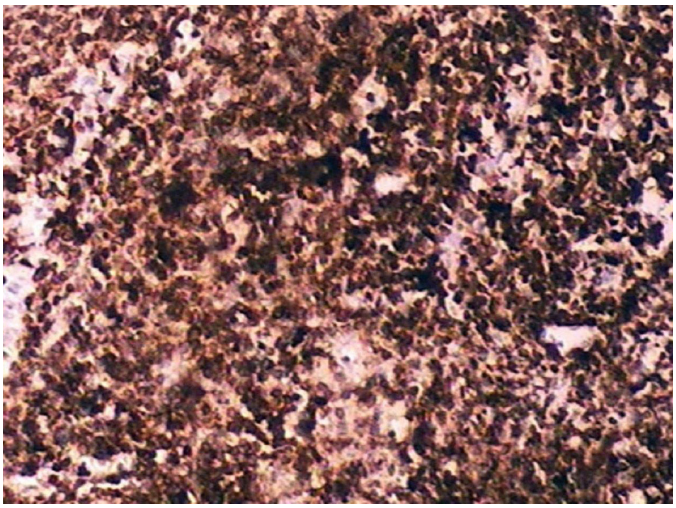

Figure 5: Immunohistochemistry - positive cytoplasmic CD3

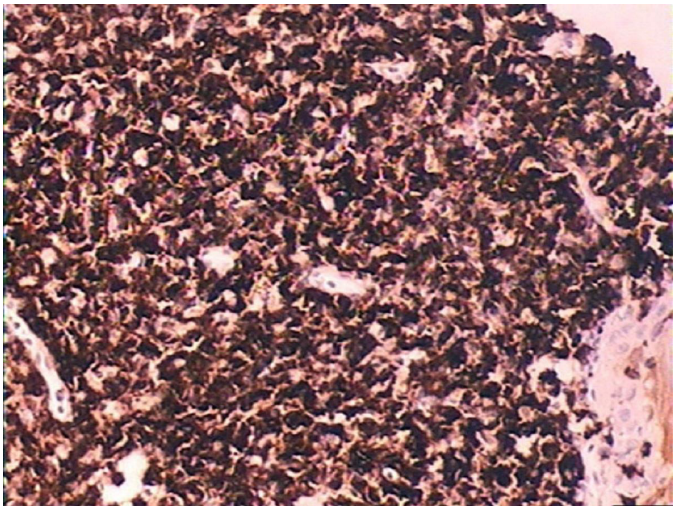

Figure 6: Immunohistochemistry - CD30 positive

DISCUSSION

Nasal-type NK/T lymphoma may present clinically with epistaxis, nasal tumor with perilesional edema, and hard palate perforation (causing communication between the nasal and oral cavities). Also, the patient may complain of nasal obstruction and/or secretion.[4] The case reported manifested similarly to the cases described in the literature, focusing on the extensive area of necrosis.

The anatomopathological study revealed infiltration of atypical lymphoid cells, neutrophils, and eosinophils, characterizing the so-called polymorphic reticulosis.[2,5] Another histopathological characteristic of this tumor is its close relationship with blood vessels (wall invasion and vascular occlusion).[2,3]

Immunohistochemistry is generally positive for CD3 (surface: negative; cytoplasmic epsilon chain: positive), CD56, CD2, and cytotoxic molecules (granzyme B, perforin, and TIA1).[2,3,5,6,7] Positive Ki-67 indicates a high rate of cell proliferation.

The quantification of EBV-DNA in blood plasma (prognostic indicator) and *in situ* hybridization to assess tumor medullary bone invasion by detecting the virus genetic material can also be used.[2,3,5]

Regarding imaging tests, in addition to computed tomography, Pet-scan (PET-CT)[5,7,8] and soft tissue magnetic resonance imaging (MRI) are considered, mainly to assess extension and complications. PET-CT is especially important in non-nasal cases as the absence of hypermetabolic areas (the tumor lymphoid cell has a high avidity for 18-fluorodeoxyglucose) in the nasal/nasopharyngeal region excludes the possibility that the primary site is the face.[2]

Regarding treatment,[2,5,6,8,9] combination of radiotherapy and chemotherapy is considered the gold standard. However, new protocols and therapies (targeted therapy, stem cell transplantation, immunotherapy targeting the EBV, among others) have been discussed.[6,9] In the case described, as in many others of this type of lymphoma, there was no time for treatment, only comfort measures. ●

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