BJCR
Brazilian Journal of Case Reports

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Publicações Científicas de Acesso Aberto

Case Report

Extranodal Nasal-Type T/NK Lymphoma with Primary Oral Manifestation and Fulminant Course: A Case Report

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Abstract: Extranodal nasal-type T/NK lymphoma (ENKTCL-NT) is a rare and aggressive non-Hodgkin neoplasm originating from natural killer (NK) cells or, less frequently, cytotoxic T lymphocytes. This subtype shows a high prevalence in Latin American populations and a strong etiological association with the Epstein–Barr virus (EBV). The present study reports the case of a 61-year-old patient with multiple comorbidities who presented with progressive ulcerated lesions in the oral cavity. Initially managed as a common infectious process, the condition proved refractory to treatment, progressing to extensive tissue necrosis and recurrent bleeding. Following multidisciplinary investigation and immunohistochemical review, the diagnosis of ENKTCL-NT was confirmed. Due to the biological aggressiveness of the neoplasm and the patient's clinical instability, the outcome was fatal before the initiation of specific oncologic therapy. This case underscores the need for deep and repeated biopsies in destructive midline lesions.

Keywords: Extranodal T/NK-cell Lymphoma; NK-cell Neoplasms; Differential Diagnosis; Oral Pathology; ENKTCL-NT; Mucormycosis.

Citation: Müller Filho FJ, Locatelli IP, Antunes PEH, Lemos BD, Eugênio GBR, Albuquerque ALT. Extranodal Nasal-Type T/NK Lymphoma with Primary Oral Manifestation and Fulminant Course: A Case Report. Brazilian Journal of Case Reports. 2026 Jan-Dec;06(1):bjcr161.

https://doi.org/10.52600/2163-583X.bjcr.2026.6.1.bjcr161

Received: 16 January 2026
Accepted: 8 February 2026
Published: 10 February 2026

1. Introduction

Extranodal nasal-type T/NK lymphoma (ENKTCL-NT) is a distinct clinicopathological entity characterized by an angiocentric and angioinvasive pattern that results in massive ischemic tissue necrosis [1,2]. Although the most common primary site is the nasal cavity and paranasal sinuses, extranodal involvement, particularly within the oral cavity, may occur and presents significant diagnostic challenges [1,3,5].

Clinically, lesions may mimic inflammatory or infectious processes (fungal or bacterial), as well as other granulomatous diseases such as granulomatosis with polyangiitis (Wegener's granulomatosis) and, crucially in diabetic patients, invasive fungal infections such as mucormycosis [4,6]. The literature indicates that superficial tissue necrosis often masks lymphomatous infiltration in initial biopsies, leading to diagnoses of "nonspecific chronic inflammation" and delaying the initiation of chemotherapy, which directly impacts survival [3,6].

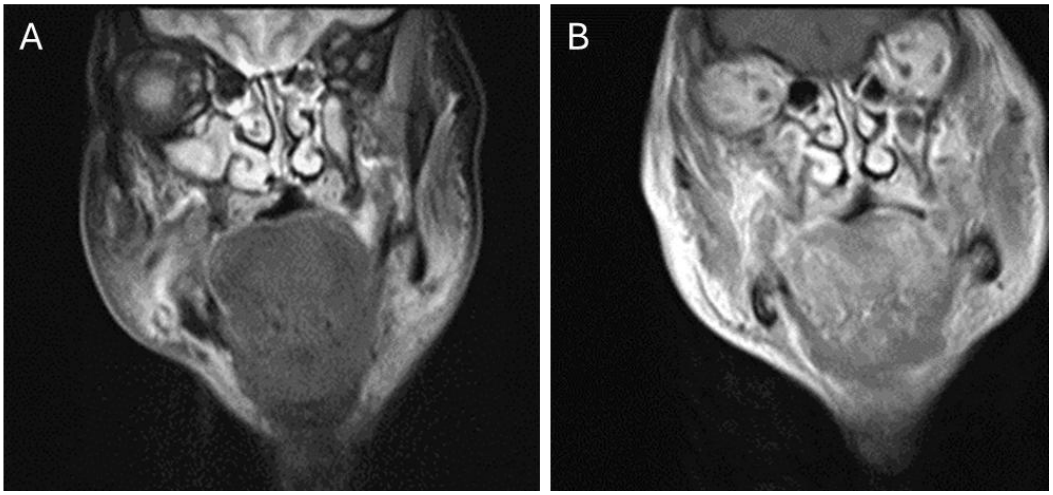
This study aims to report a rare and aggressive case of neoplasm with primary oral presentation, highlighting the diagnostic challenges, rapid clinical progression, and the importance of early recognition to optimize therapeutic possibilities and patient prognosis [3,5,7].

2. Case Report

A 61-year-old female patient with a medical history of systemic arterial hypertension, type 2 diabetes mellitus (insulin-dependent), hypothyroidism, and schizophrenia under pharmacological control presented in April 2025 with edema of the upper lip associated with painful ulcers on the hard palate and buccal mucosa. She also reported otalgia, purulent discharge, and fever spikes. Initial evaluations at an emergency care unit suggested bullous myringitis and stomatitis of viral or fungal etiology. The patient was admitted on April 28, 2025, due to persistence and worsening of the lesions.

Intraoral examination revealed extensive ulcerated areas on the hard palate with bone exposure and soft tissue infiltration. Nasal endoscopy performed at admission showed no lesions in the nasal mucosa or nasopharynx. Magnetic Resonance Imaging (MRI) of the face demonstrated thickening and heterogeneous enhancement of the left hard palate mucosa, extending to the nasolabial sulcus (Figures 1A and 1B), suggesting an infiltrative process, while confirming the absence of occupation or bone destruction in the paranasal sinuses and nasal cavity, supporting a primary oral presentation (Figure 3). Systemic staging with Computed Tomography (CT) of the chest, abdomen, and pelvis revealed no evidence of distant disseminated disease.

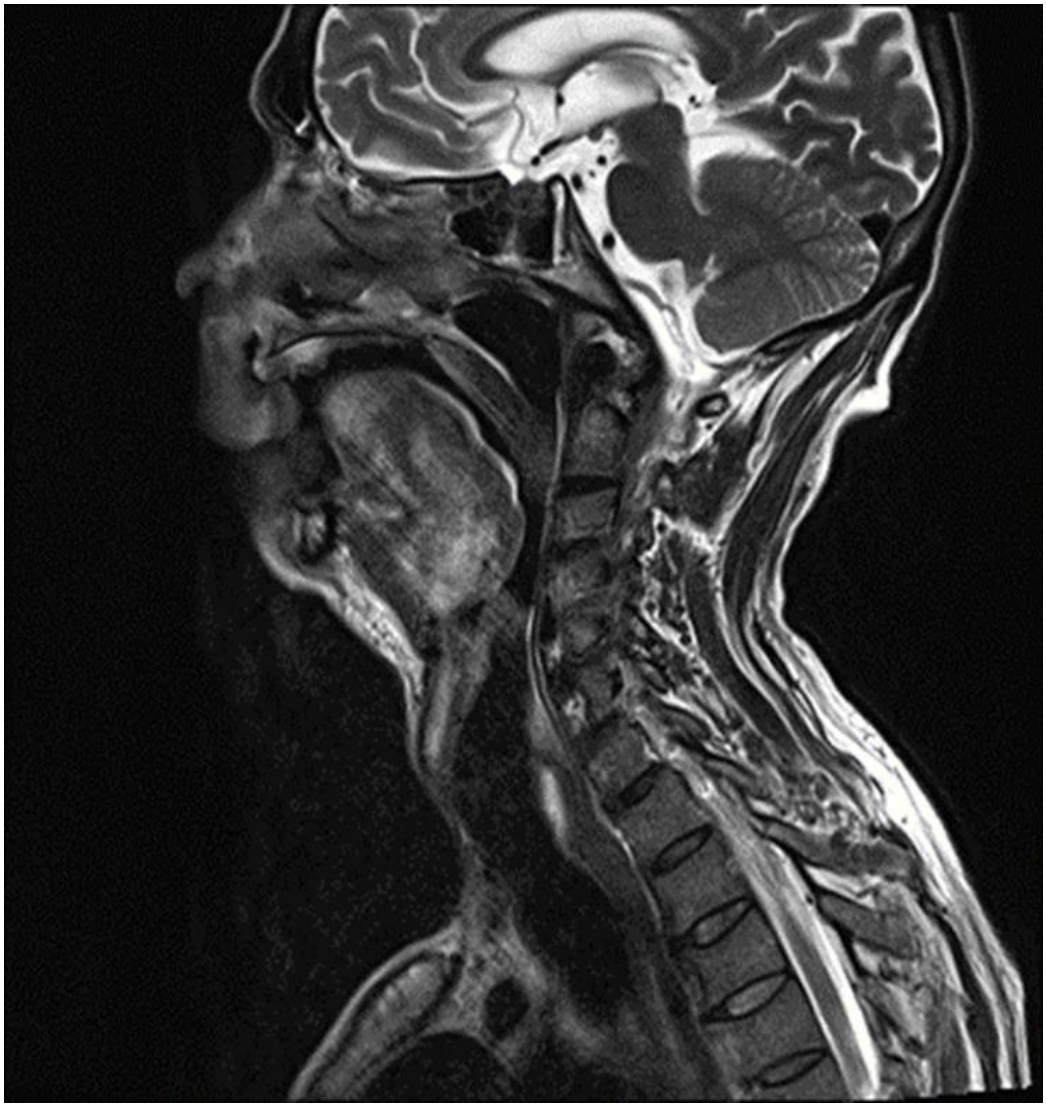
Figure 1. A. Coronal STIR image demonstrating asymmetry of the hard palate. B. Coronal T1 FS DIXON post-contrast (C+) image showing subtle blurring of the hard palate.



Initial incisional biopsies were inconclusive, reporting only necrotic tissue and chronic inflammatory infiltrate; special PAS and Grocott stains were performed to exclude a fungal etiology (mucormycosis), yielding negative results. Given the rapid progression and destructive nature of the lesion, a new deep biopsy and immunohistochemical study were performed. The panel revealed positive markers for CD3 (cytoplasmic), CD56 (focal), granzyme B (cytotoxic protein), and CD43. Testing for EBER (EBV-encoded RNA) could not be completed in time due to clinical deterioration; however, the morphology and immunophenotype were considered diagnostic. The Ki-67 proliferative index was 80%, indicating high mitotic activity.

Despite intensive support and surgical debridement to control infectious foci, the patient developed hemodynamic deterioration and respiratory instability. The 33-day interval between MRI and definitive diagnosis, exacerbated by the difficulty in obtaining an adequately deep sample during the initial approach, prevented interventions such as palliative radiotherapy. The patient died on June 20, 2025, without clinical conditions to undergo chemotherapy protocols (such as SMILE or asparaginase).

Figure 2. Sagittal T2-weighted image demonstrating irregularity of the hard palate.



3. Discussion

The diagnosis of extranodal nasal-type T/NK lymphoma (ENKTCL-NT) in the oral cavity represents one of the greatest challenges in clinical pathology [3,6]. The pathognomonic feature of this disease is its angiocentric pattern, in which neoplastic cells infiltrate the walls of blood vessels, causing luminal occlusion and resulting in extensive ischemic coagulative necrosis [1,2]. This mechanism explains the "diagnostic pitfall" observed in the present case, in which superficial biopsies yielded false-negative results containing only necrotic tissue [3,6]. The initial failure to detect the disease was not solely due to its rarity, but rather to the insufficient depth of the first biopsy, which is critical for distinguishing septic necrosis from an underlying lymphoproliferative process [6]. In insulin-dependent diabetic patients, the mandatory exclusion of invasive fungal infections, such as mucormycosis, through Grocott and PAS staining is essential, as the clinical presentation is virtually indistinguishable [4,6].

From a technical standpoint, immunohistochemical differentiation is crucial. The expression of cytotoxic proteins, such as granzyme B, confirms the aggressive cytotoxic lineage [1,2]. Although in situ hybridization for EBER is considered the gold standard and flow cytometry is ideal for confirming clonality in cases with focal CD56 expression, the combination of cytoplasmic CD3 positivity, cytotoxic marker expression, and angiocentric growth was sufficient to establish the diagnosis in this urgent clinical scenario [2,5,6]. In

the present case, a Ki-67 index of 80% further supports the fulminant biological behavior of the disease [2,3].

Another relevant aspect is disease management in patients with psychiatric comorbidities. Schizophrenia impaired accurate communication of early symptoms, contributing to a delay of more than 30 days between hospital admission and definitive therapy [6,7]. Although radiotherapy is the cornerstone of treatment for localized disease (stage I/II), the rapid deterioration of the patient's performance status (ECOG) ultimately contraindicated any therapeutic modality [2,3].

6. Conclusion

This report underscores that persistent, destructive, and refractory oral ulcers should undergo immediate deep biopsy with immunohistochemical analysis and fungal staining. ENKTCL-NT is a rapidly progressive disease; prompt clinical suspicion and imaging-based exclusion of an occult nasal primary site are the only factors that may enable timely therapeutic intervention.

Funding: None.

Research Ethics Committee Approval: We declare that the patient approved participation in the study by signing an informed consent form, and that the study followed the ethical guidelines established by the Declaration of Helsinki.

Acknowledgments: None.

Conflicts of Interest: All other authors declare no conflicts of interest.

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