

Extranodal NK/T-Cell Lymphoma, Nasal Type (Angiocentric T-Cell Lymphoma): A Case Report

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Abstract

Case Report

NK/T lymphoma is a rare type of non-Hodgkin's lymphoma. It corresponds to a clinical entity with necrosis that starts preferentially in the nasal cavities and the nasopharynx and extends to the centofacial bone structures. Its anatomopathological diagnosis is not always obvious. Modern immunophenotyping and molecular biology tools have made it possible to isolate this type of lymphoma. Treatment is based on radiotherapy and chemotherapy. Nasosinus localization of T/NK lymphoma is rare. This is the case report of a 18-year-old female patient who presented with nasal obstruction and foul smelling, edema of the right hemiface and an ulcerative lesion over the palate of 4 months duration, which had been treated with antibiotics without success. After performing a number of diagnostic tests, it was found histologically and confirmed by immunohistochemical analysis that the patient had an ENKL, nasal type (also known as angiocentric T-cell lymphoma). Our Purpose of the presentation is To discuss the particularity of this localization, the diagnosis difficulties and therapeutic modalities.

Keywords: Extranodal NK/T-cell lymphoma, nasal type, chemotherapy.

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INTRODUCTION

Extranodal NK/T-cell lymphoma, nasal type (ENKTL) is a rare type of lymphoma that is endemic to East Asia and parts of Central and South America. Most (80% to 90%) patients present with nasal obstruction, sinusitis, ulcer, and epistaxis due to a destructive mass involving the midline facial tissues [4, 1]. ENKTL most often presents as a localized disease, clinical stage I-II; however, wide spread dissemination can occur in a subset of patients. Occasionally, patients with ENKTL present with only extranasal sites of disease, most often skin, lung, and gastrointestinal tract, but a variety of other extranasal sites have been reported [1, 2, 5]. Accurate diagnosis of ENKTL can be challenging, especially in small biopsy specimens or in frozen sections, as the neoplastic cells are often admixed with inflammatory cells and necrosis.

CASE REPORT

A 18-year-old female, without any previous history, presented a budding formation, bleeding on contact, filling the entire right nasal cavity.

Examination of the oral cavity showed ulceration of the palate with right jugal infiltration with homolateral palpebral edema obstructing the palpebral fissure with purulent secretions. The rest of the clinical examination was normal, especially the lymph nodes. The nasosinus CT scan showed a nasal tumor process invading the homolateral maxillary sinus, with osteolysis of the alveolar border of the maxilla and the hard palate and lysis of the nasal septum and nasosinus walls. Histopathological study of biopsy samples from the right nasal cavity and the bony palate taken under local anesthesia showed a diffusé undifferentiated process with large round cells, immunohistochemistry made the diagnosis of certainty of nasal T/NK lymphoma; lymphoid elements were positive for CD3, CD2 and CD56. The extension workup was negative. She underwent multidrug therapy (4 courses of CHOEP: adriamycin, vincristine, cyclophosphamide, etoposide, and prednisone) followed by locoregional external radiotherapy at a dose of 50 Gy in 25 sessions (2 Gy/fr; 5 fr/week). She is in complete clinical and radiological remission after 15 months.



Fig-1: Coronal CT section showing an infiltrated tissue of the right nasal cavity, necrotic and intensely enhanced after contrast. It obstructs the middle turbinate, It also lyses the internal wall of the right maxillary sinus with the beginning of hypodense filling of the ethmoidal cells.



Fig-2: A poorly limited ulcerating lesion with irregular contours and a necrotic ulcer surface extending from the right surface to the malar region



Fig-3: A loss of mucosal substance of the hard palate, the edges were inflammatory

DISCUSSION

Malignant lymphomas of the head and neck region that originate in the nasal cavity, paranasal sinuses and hard palate form an interesting and frequently diagnostically difficult group [1]. In the past, these lymphomas have been confused with a number of infectious, autoimmune, or inflammatory designations, most of which we now know represent peripheral T-cell lymphomas or angiocentric immunoproliferative lesions [2].

Nasal T/NK lymphoma is an aggressive form of non-Hodgkin's lymphoma with specific clinicopathological characteristics. The combination of chemotherapy for advanced stages does not seem to improve survival compared to radiotherapy alone which remains the treatment of choice especially for localized stages [3]. The nasal location of T /NK lymphoma is rare [4]. The diagnosis is based on immunohistochemical study. The treatment includes chemotherapy and radiotherapy [5, 2]. This type of lymphoma has a poor prognosis, even with treatment. A better management of T/NK lymphoma requires multidisciplinary collaboration between ENT doctors, radio-chemotherapists and nutritionists in order to improve the prognosis of this pathology.

CONCLUSION

Nonspecific nasal symptoms often predate the appearance of mucosal ulceration and tissue necrosis by one year or more. The ambiguous nature of these symptoms can result in a delay in diagnosis. Representative biopsy material and good interaction with the pathologist is important. Although not always possible, a diagnosis should be sought prior to commencing a treatment course. In conclusion, clinicians should consider extranodal nasal lymphoma as a rare cause of midline destructive lesions. They should be aware of the difficulties of obtaining histological diagnosis despite apparently adequate biopsies.

Conflict of interests

The authors declare no conflicting interests.

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