

Non-Hodgkin's Lymphoma of Nasal Cavity and Paranasal Sinuses: A Case Report

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Abstract

Case Report

Non-hodgkin lymphoma (NHL) is rare tumors of the jaws. Their locations in the air spaces of the face coming in second row. We related the case of a primitive NHL located in two maxillary sinuses with an extension to the nasal and orbital fossa in a patient of 41 year, which illustrates the diagnostic difficulties encountered face a little suggestive symptoms. The diagnosis was confirmed by histological examination. In this observation, we will review the literature in order to review the clinical, radiological, histological, and therapeutic and outcome of this rare disease.

Keywords: Lymphoma, non-Hodgkin, maxillary sinus.

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INTRODUCTION

Non-Hodgkin's malignant lymphomas (NHML) represent the set of monoclonal malignant proliferations beginning in lymphoid tissue affecting B or T cells and which do not meet the definition of Hodgkin's disease. The mode of revelation of NHML is lymph node in 2/3 of cases and extra-ganglionic in 1/3 of cases. The localization in the nasal cavity and paranasal sinuses comes after that of the Waldeyer's ring (20). Its frequency is estimated between 7 and 22% of ORL lymphomas. The diagnosis of these locations, which is difficult to assess on clinical examination, is more difficult and requires CT imaging (21).

Histologically, the diffuse large B-cell forms are predominant in paranasal locations and, in particular, in the maxillary sinus (14). Centrofacial involvement is of T origin and has a poorer prognosis (1).

The case of a primary NHL localized to the maxillary sinus discovered after a facial swelling, proptosis and unilateral blindness has been reported, it illustrates the diagnostic difficulties encountered when faced with a symptom that is not very suggestive.

CASE REPORT

This is a 41-year-old man, with no particular medical and surgical history, who presents with an inflammatory right cheek swelling that has been evolving for two years, complicated by exophthalmos

and blindness of the left eye that have developed since 2 months.

Clinical examination showed a bilateral inflammatory jugular swelling more marked on the right side associated with exophthalmos of the left eye.

The buccal cavity examination revealed an alveolar arch, going from tooth 15 to the tuberosity of the maxilla, and which overhangs the adjacent palate without filling the vestibule, this arch was cracked along the alveolar arch (figure 1). Palpation revealed a firm and very sensitive consistency with bilateral cervical lymphadenopathy.

The CT scan revealed a voluminous aggressive tissular mass centered on the two maxillary sinuses, causing osteolysis of their bony walls and of the right bony palate with infiltration of the opposite soft tissues, in particular the pterygoid muscles with extension to the nasal fossa. Left eye thus responsible for grade III proptosis; associated with endocranial extension (figure 2).

The craniofacial and orbital MRI showed a tumor of the two maxillary sinuses and of the left nasal fossa with iso signal on T1 and hyper signal on T2 with a heterogeneous enhancement after contrast media injection, with extension to the deep spaces of the face, to the maxillary bone, to the palate, the nasopharynx, left orbit and base of the skull (Figure 3).

These clinical and radiological elements suggested a malignant tumor. With this in mind, a biopsy was carried out revealing: a malignant tumor proliferation, the cells are large or medium in size arranged in diffuse layers, associated with large foci of ischemic-looking necrosis.

A complementary immunohistochemical study was carried out, it objectified:

- Anti Bcl-6: positive in 10% of tumor cells.
- Anti CD20: positive
- Anti-Ki-67 Antigen: positive in 20% of tumor cells.
- Lack of expression of anti CD3 and anti CD10.

All of these elements are characteristic of non-Hodgkin's large cell malignant lymphoma of the B phenotype.

An extension assessement including chest x-ray, abdominal ultrasound, and thoraco-abdominal CT was performed and did not identify other secondary locations.

We therefore concluded to a primary non-Hodgkin's type B malignant lymphoma localized in the maxillary sinuses and classified as stage IV according to the Ann Arbor classification.

The patient was referred to the oncology department for further treatment.



Fig-1: buccal examination: arching of the palatal arch going from tooth 15 to the tuberosity of the maxilla, with cracking of the bony palate

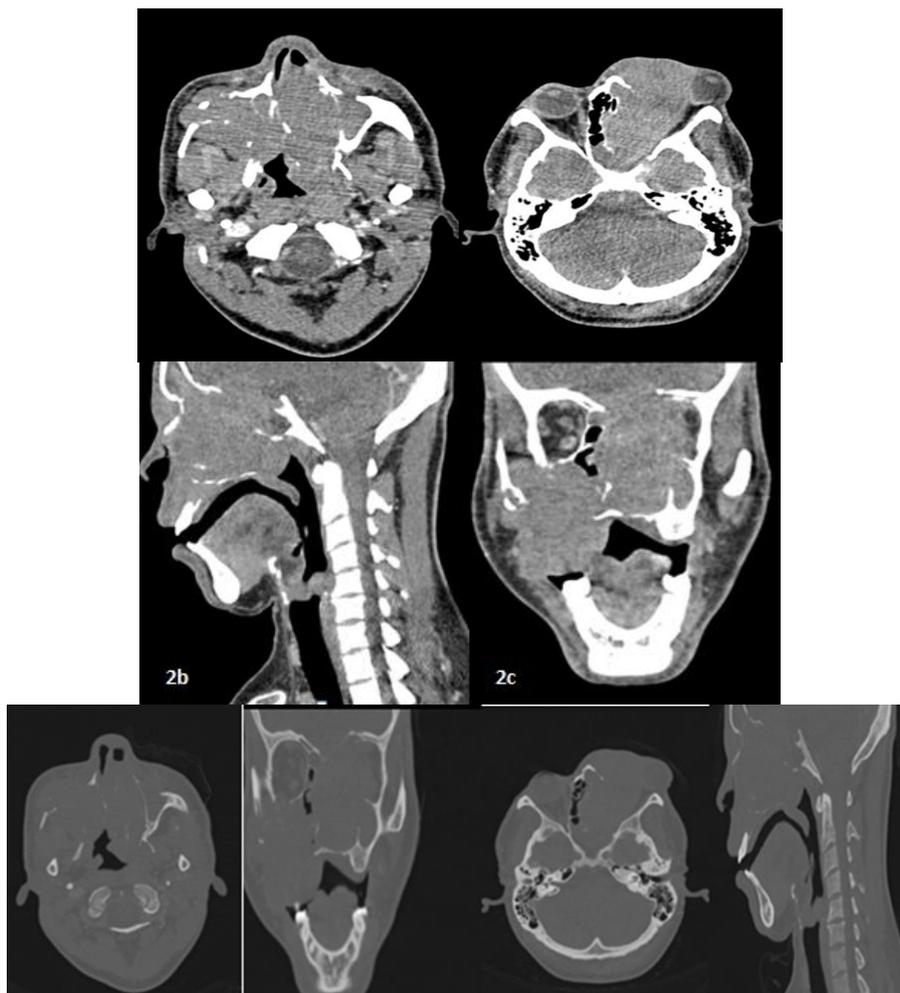


Figure-2 and 3: Facial CT: axial slices (a), coronal and sagittal reconstructions (b) and (c) in mediastinal and bone window: showing a voluminous aggressive masses centered on the two maxillary sinuses and causing an osteolysis of their bone walls , from the bony palate and the sphenoid body with endocranial extension, to the nasal cavity and to the left eye

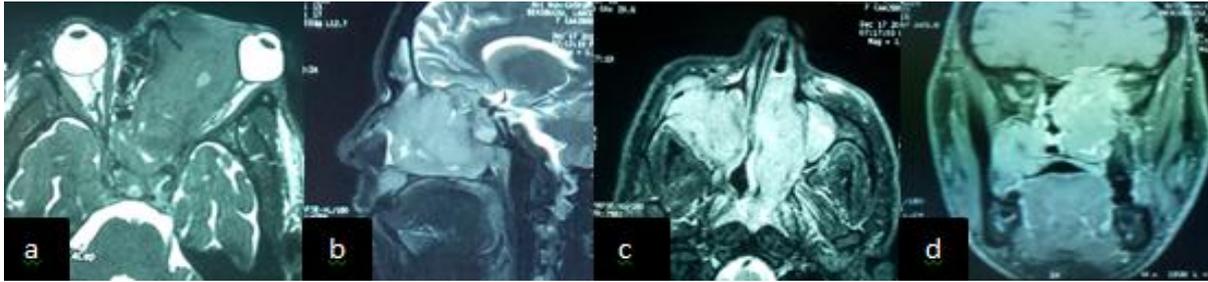


Fig-4: Craniofacial and orbital MRI, axial and sagittal T2 sequence (a, b) and axial and coronal T1 sequence with Gadolinium injection (c, d): demonstrating a tumor process of the two maxillary sinuses and the left nasal fossa that is hypersignal on T2 with heterogeneous enhancement and extension to the deep spaces of the face, the jawbone, the bony palate, the nasopharynx, the left orbit and at the base of the skull

DISCUSSION

Non-Hodgkin's malignant lymphomas constitute a heterogeneous set of lymphoid proliferations affecting B or T cells. The mode of revelation is lymph node in 2/3 of cases and extra lymph node in 1/3 of cases.

In the cervicofacial extremity, Waldeyer's lymph ring is the most common site of NHL, followed by the major salivary glands, in this case the parotid gland and the submandibular gland [1]. In contrast, NHL of the nasal cavity and paranasal sinuses remains exceptional and represents only 0.17% of all lymphomas [2] and only 3% of cervicocephalic neoplasias [3]. The most frequent histological type is type B, while type T remains exceptional [3].

These lymphomas most commonly occur in the sixth decade of life, with a slight male predominance [4]. The clinical manifestations are not specific [5]. If the general signs are rare, the local signs are mainly represented by an often old nasal obstruction, more rarely a rhinorrhea or recurrent epistaxis [5]. Finally, the ophthalmological signs can be rarely revealing such as an exophthalmos (1.5 to 5). % [6, 7], decreased visual acuity or limited eye movements.

The differential diagnosis arises mainly with other tumors of the jaw, in particular with squamous cell carcinoma [8]. The differential diagnosis also includes mucocoeles, infectious aetiologies (acute and chronic sinusitis), antrochoanal inflammatory polyp and tumors (benign, malignant and metastatic) [2].

In conventional imaging, the images observed reflect bone invasion and/or tumor extension. The bony localization of a malignant lymphoid proliferation takes on the appearance of a diffuse radiolucency with disappearance of the lamina dura in the dentate sectors and from the bottom sinus border to the maxilla [9]. However, the radiological appearance of a malignant epithelial tumor is that of frank and massive osteolysis [10].

Computed tomography examination is essential for assessing bone invasion and for estimating the size of the lesion for its TNM classification [10]. Lymphomatous lesions are usually parenchymal in appearance, isodense, with invasion of neighboring cavities. The bone walls are most often eroded decalcified, lysed, with a honeycomb-like areolar appearance. In fact, the late onset of bone involvement compared to the onset of the disease, the mildness of the bone involvement contrasting with extensive tumor formation may be suggestive of lymphomatous etiology [4, 5].

Magnetic resonance imaging (MRI) has an interest in the assessment of extension to adjacent structures and also allows differentiating the tumor process from an inflammatory lesion [11]. Lymphoma appears iso intense before injection, moderately enhanced by the contrast product and differentiating from retention fluids on T2 sequences by its relative iso signal [12]. Intraorbital extension results in a mass, surrounding and/or infiltrating the retro-orbital nerve, fatty and muscular structures, which can lead to exophthalmos.

The definitive diagnosis can only be established by the histological definition of tumor proliferation and the characterization of its phenotype by immunohistochemical study [13]. These immunohistochemical criteria are essential for therapeutic management because, along with the stage of the disease, they are the two main prognostic factors [14, 15].

The most widely used classification system for staging NHL is the Ann Arbor system, which considers the number of tumor sites (lymph node and extranodal) and their location, as well as the presence or absence of B symptoms [16-17].

The treatment of these lymphomas is based on polyvalent chemotherapy isolated or associated with external radiotherapy (30 to 50 Grays) in the most advanced cases (T3, T4) [18]. The isolated clinical forms (stage IA E) are usually of good prognosis with a survival ranging from 70% to 100% at 5 years [1, 14].

Overall 5-year survival is 30-60% in stages I-II and drops to 30% for more advanced stages [19].

CONCLUSION

Nasosinus lymphomas represent a rare entity, characterized by an initial symptomatology often rough, delaying the diagnosis. Imaging is nonspecific, and the diagnosis remains histological. Chemotherapy adjuvant to radiotherapy gives good results. The prognosis depends on the histologic type, the Ann Harbor stage at diagnosis and the therapeutic possibilities for each patient.

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