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# **Primary Nasal Lymphoma: A Case Report**

#### Abstract

**Objective:** Describe the modalities for the management of Primary nasal lymphoma.

**Materials and methods:** We report the case of a 63-year-old man who presented with Primary nasal lymphoma.

**Case report:** This is a 63-year-old patient admitted to the emergency department. He was consulted for progressive nasal obstruction, first unilateral right then bilateral, complete on the right and partial on the left, with rhinorrhoea mucopurulent streaked with blood. On inspection, there is a swelling in the right nasal fossa protruding through the nostril opening and effacing the nasolabial fold, with the facing skin inflamed. A biopsy of the mass with pathological examination and immunohistochemical study (IHC) confirmed the diagnosis of plasma cell lymphoma. The remainder of the extension record was normal. The patient received 5 courses of adjuvant chemotherapy according to the CHOP protocol (adriamycin + vincristine + cyclophosphamide + prednisone), followed by locoregional external radiotherapy (sinuses and nasal cavities) at a dose of 48.6 Gy in 27 sessions.

**Conclusion:** Nasosinus lymphomas represent a rare entity, characterized by initial symptomatology often rough, delaying the diagnosis. Radiology is non-specific, and the diagnosis remains histological. Chemotherapy adjuvant to radiotherapy gives good results.

Keywords: Lymphoma; Sinonasal; Diagnosis; Treatment

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### Introduction

The primary localization of nasosinusal lymphomas is rare, representing less than 1% of non-Hodgkin lymphomas [1]. This entity currently includes Centro facial malignant granulomas and certain forms of Wegener's disease which have traditionally been treated with long-term corticosteroids or alkylating agents. These lymphomas are characterized by their clinical polymorphism, their binding to Epstein-Bar virus (EBV), and the poor prognosis of the extensive forms [2]. Therapeutic advances have been obtained by the more frequent use of chemotherapy combined with radiotherapy, especially for large and/or aggressive tumors [3].

#### **Case Report**

A 63-year-old patient consulted for a progressive nasal obstruction, first unilateral right then bilateral, complete on the right and partial on the left, with blood-streaked mucopurulent rhinorrhea.

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On clinical examination, the patient appeared to be in fairly good general condition, afebrile. On inspection, there is a swelling in the right nasal fossa protruding through the nostril opening and effacing the nasolabial fold, with the facing skin inflamed. On endoscopic examination, which was very difficult, there was a budding fleshy polypoid formation filling the entire right nasal fossa, with a free middle meatus, after aspiration of purulent secretions and pushing back the middle turbinate and the nasal septum to the left; this formation bleeds easily on contact. On the left side, there was an incomplete nasal obstruction related to the deviation of the septum in its anterior part. The remainder of the physical examination was normal, particularly the lymph node areas **(Figure 1).** 

A nasosinusal computed tomography showed an opacity of tissue density occupying the anterior two-thirds of the 2 nasal cavities more important on the right with sinuses of the free face and, with lysis of the internal wall of the maxillary sinus and the nasal

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Figure 1 A photographic image of the patient's face (A) and profile (B) showing tumefaction of the right nasolabial fold obstructing the homolateral nasal fossa, with a little inflamed opposite.

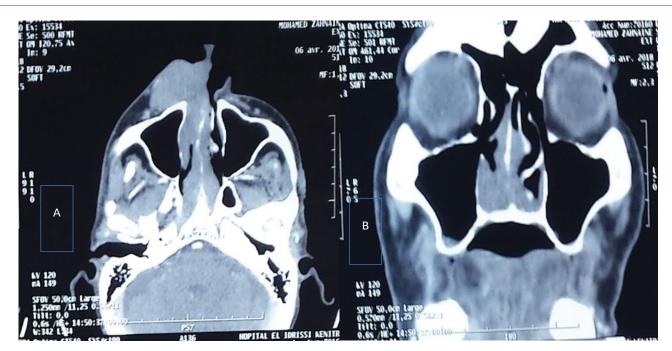


Figure 2 Image of the facial CT scan with an injection of the contrast product in axial (A) and coronal (B) section showing opacity of tissue density occupying the anterior two-thirds of the 2 larger nasal cavities on the right with free facial sinuses, with lysis of the wall internal of the maxillary sinus and the nasal septum, and infiltrating the soft parts under the skin.

septum, and infiltrating soft tissue under the skin. This mass was enhanced after the injection of the contrast product and showed no calcification (Figure 2). A biopsy of the mass, which was friable, at the level of the right nasal fossa was carried out: the pathological examination of the inclusion block objectified the existence of a malignant tumor formation characterized by a diffuse cellular layer formed large lymphomatous elements, pleomorphic in appearance, with clear plasmacytic differentiation and the presence of numerous plasmablasts. Mitoses were numerous.

Immunostaining with anti-CD3 antibodies and anti-CD56 antibodies showed diffuse labeling of tumor cells, anti-Ki 67 antibodies showed nuclear labeling of 80% of tumor cells, and anti-CD20 antibodies showed labeling of a few rare B lymphocytes, while that with the anti-AE1AE3 antibody was negative, concluding in a plasma cell lymphoma.

Once the diagnosis of lymphoma was made, an extension workup was performed with a clinical neurological examination, ophthalmology, and in particular of the lymph node areas, which was normal. The laboratory examinations were normal, the sternal puncture was negative and the thoracic-abdominopelvic scanner was normal. This lymphoma was classified as stage IE according to the Ann Arbor classification.

The patient received 5 courses of adjuvant chemotherapy according to the CHOP protocol (adriamycin + vincristine + cyclophosphamide + prednisone), followed by locoregional external radiotherapy (sinuses and nasal cavities) at a dose of 48.6 Gy in 27 sessions. This radio chemotherapy was well tolerated by the patient. At the end of treatment, the clinical symptoms disappeared and endoscopic examination was strictly normal apart from a deviation of the nasal septum and its perforation caused by tumor lysis. The evolution was favourable, without recurrence with a follow-up of 4 years.

#### Discussion

Lymphomas of extranodal origin constitute 10% to 58% of non-Hodgkin lymphomas (NHL) [4]. Primary paranasal sinus lymphomas are rare, constituting only 3% of cervicofacial neoplasias [4]. This nasosinusal location remains exceptional and represents only 0.17% of all lymphomas [5]. The most frequent histological type is type B, while type T remains exceptional [4].

These lymphomas occur with predilection during the sixth decade of life, with a slight male predominance [6], as in the case of our patient. The clinical manifestations are nonspecific, and their intensity is often less than the observed tumor volume would suggest [7]. If the general signs are rare, the local signs are mainly represented by an often-old nasal obstruction, more rarely recurrent rhinorrhea, or epistaxis [7].

The cervicothoracic CT scan is essential for the study of lesions, which have certain peculiarities: they are generally homogeneous opacities with polycyclic, regular outlines, most often located at the level of the intersinusomaxillary septum; the bone walls

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are most often eroded, decalcified, lysed, with a honeycomblike areolar appearance. 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 [6,7]. This is the case with our patient, who presented with a large tumor while the destructive phenomena were less important. Radiological control studies have shown the persistence of sequelae thickening or fibrosis [4,7]. Magnetic resonance imaging (MRI) has an interest in the assessment of extension to adjacent structures and also allows to differentiate the tumor process from an inflammatory lesion.

The definitive diagnosis is based on histological examination and the IHC, the latter also making it possible to determine the type of lymphoma (B or T). Among the differential diagnoses, mucoceles are the most common. The differential diagnosis also includes infectious aetiologies (acute and chronic sinusitis), antrochoanal inflammatory polyp, and tumors (benign, malignant, and metastatic) [5].

The initial treatment of nasosinusal lymphomas is mainly based on radiotherapy [8]. The role of surgery in the management of these lymphomas has never been validated in terms of tumor control or survival [8]. The indications for adjuvant chemotherapy from the outset are still debated [9,10]. Regarding the therapeutic attitude, it is still a source of controversy. Not all teams have the same attitude for Stage I [9,10]. Some teams continue to offer radiotherapy alone for stage I since the response to treatment is complete in 95 to 100% of cases [9]. However, some authors have shown that the course, when it is not favorable in stages I, most often corresponds to a course of the disease away from the initial focus, and therefore recommend adjuvant chemotherapy from stage I. radiotherapy [10]. This attitude is the one we adopted in our patient, who received chemotherapy according to the CHOP protocol, followed by radiotherapy. Encouraging results have been obtained with the use of an anti-CD20 monoclonal antibody (rituximab) in combination with CHOP-type chemotherapy [11,12]. The course is particularly pejorative when the lymphomatous cells express the EBV genome, the CD5, or the NK antigen, if a hemophagocytic syndrome appears, angiocentric or angioinvasive images, or a high grade of malignancy [11,12]. Overall, 5-year survival is 30-60% in stages I-II and drops to 30% for more advanced stages.

# Conclusion

Nasosinusal lymphomas represent a rare entity, characterized by initial symptomatology often rough, delaying the diagnosis. Radiology is non-specific, and the diagnosis remains histological. Chemotherapy adjuvant to radiotherapy gives good results.

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