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Uncommon Presentation of Non-Hodgkin's Lymphoma of Maxillary Sinus: A Case Report

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Abstract Non-Hodgkin's Lymphoma is a group of neoplasms that originate from the cells of the lymphoreticular system. In 40% of cases, it arises from extranodal sites. The nasal cavities and paranasal sinuses are rarely affected by primary Non hodgkin's lymphoma. We report the case of a 60-year-old man admitted for Non-Hodgkin's Lymphoma of maxillary sinus. CT scan demonstrated an extensive lesion destroying the medial and anterior walls. The patient received 6 cycles of the chemotherapy. The follow-up for the last 1 year was normal without recurrence. The difficulty encountered in the diagnosis of this clinicopathological entity owing to its radiological and clinical expression, particularly the evolution was discussed. Though rare, the signs and symptoms of the case suggest that Non hodgkin's lymphoma should be considered in the differential diagnosis of nasal cavity lesions.

Keywords: *lymphoma, maxillary, clinical, radiological, treatment*

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1. Introduction

Malignant lymphomas are a neoplasm originating from lymphocytes or cells generated during the multipotential differentiation of a stem cell [1]. They are relatively uncommon, representing less than 1% of all head and neck malignancies [2]. The most common subtype, approximately 80% of lymphomas, is represented by large B-cell lymphomas (LBCL) [3]. It is a fast growing malignancy arising inside or outside of the lymphatic system. Skin, abdomen, lung, central nervous system, and oral cavity are common locations [4]. The nasal cavities and paranasal sinuses are rarely affected by primary Non-Hodgkin's lymphoma (NHL). Its overall incidence is 8 % of paranasal malignancies [5]. We report an unusual case of a NHL of maxillary sinus. We highlight the epidemiological, diagnosis and treatment of this uncommon disease in order to illustrate the diagnostic difficulties due to the anatomical situation very complex and its late diagnosis.

2. Case Report

A 60-year-old male, presented in our ENT department; with a six months history of a firm mass in the left cheek, which slowly increased over. History revealed a left unilateral nasal obstruction with nose bleeding. There is no history of smoking or alcohol drinking. Also, he didn't have any history of medical surgery. Physical examination of the facial region revealed a painless swelling of the left cheek, measuring about 4 cm/ 3cm (Figure 1).

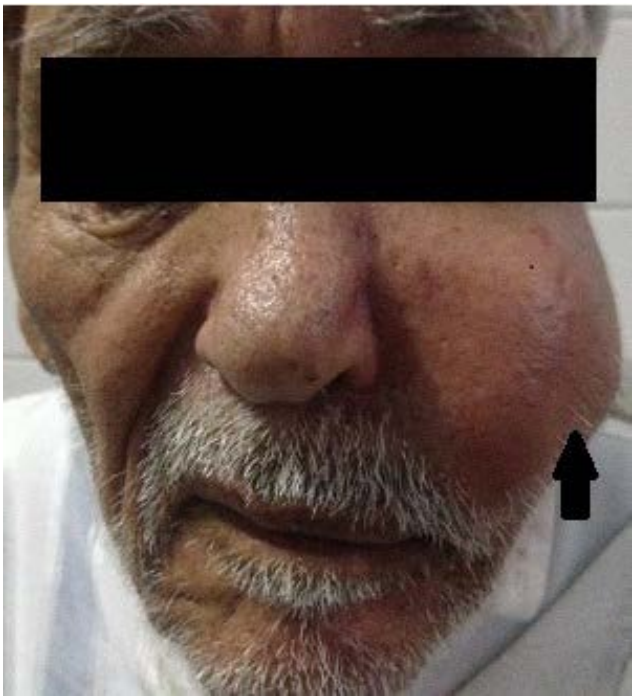

Figure 1. Extra-oral examination shows a mass on the left cheek

This lesion was firm in consistency, non-erythematous with a healthy skin. There was no paresthesia or anesthesia of the face. Endobuccal examination revealed a tumefaction of the left upper gum with palatal and vestibular extension (Figure 2).

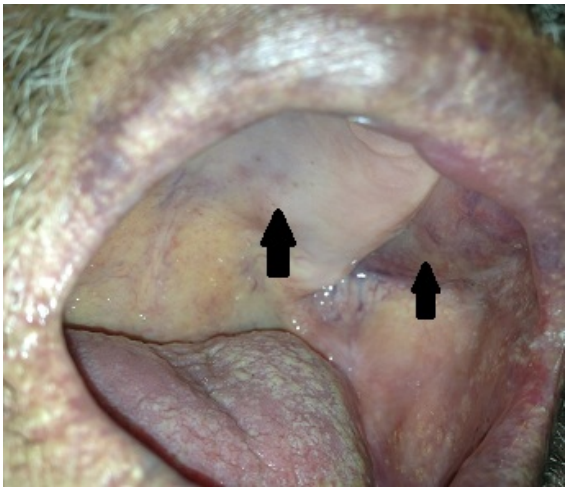

Figure 2. Intra-oral examination shows a mass on left upper gum with palatal and vestibular extension

Table 1. table summarizing the patient characteristics and clinicopathological information

	Yes	No
Left nasal obstruction	**x**	
Left nose bleeding	**x**	
History of alcohol drinking		**x**
History of smoking		**x**
History of surgery		**x**
Swelling of the left cheek	**x**	
Paresthesia or anesthesia of the face	**x**	
Endobuccal tumefaction	**x**	
Lymph nodes		**x**

There were no palpable regional lymph nodes. Visual function was preserved and visual acuity was measured as 10/10 in each eye. A systemic examination was unremarkable. Nasal endoscopic examination found mucopurulent discharge and tumefaction in the left nasal cavity. An axial computed tomography (CT) scan revealed a heterogeneous large lesion in the left maxillary sinus destroying the medial and anterior walls with extending medially into the nasal septum and inferiorly into the hard palate and alveolar processes of the left maxilla in (Figure 3).

A biopsy with histopathological examination and immunohistochemistry analysis found positivity of CD20 marker (Figure 4).

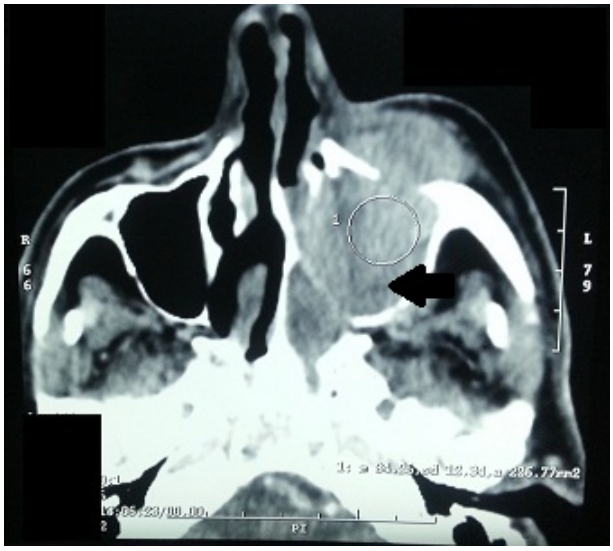

Figure 3. Axial CT scan showed a heterogeneous expansive lesion in the left maxillary sinus

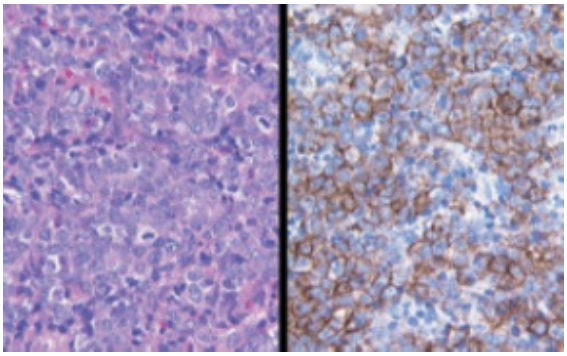

Figure 4. lymphoma diffuse large B-cell with high CD20 immunoreactivity

The diagnosis of LBCL is confirmed. The patient was then referred to a hematologist and an oncologist for further evaluation. Staging was composed of CT scan of the head and neck, lung, abdomen, pelvis, and of bone marrow biopsy. Full blood counts, a coagulation screen, blood chemistry studies, serum lactate dehydrogenase levels were normal. An immunodeficiency test (HIV) was negative. His disease was staged at IE according to the Ann Arbor staging system.

Table 2. table summarizing the patient investigations and assays performed

Investigations and assays performed	Report
Nasal endoscopic examination	Mucopurulent discharge and tumefaction in the left nasal cavity
Computed Tomography (CT)	Heterogeneous expansive lesion in the left maxillary sinus with local extension
Histopathological examination and immunohistochemistry	LBCL with positivity of CD20 marker
CT scan of the head and neck, lung, abdomen, pelvis	Normal
Bone marrow biopsy	Normal
Full blood counts	Normal
Coagulation screen	Normal
Blood chemistry studies	Normal
Serum lactate dehydrogenase levels	Normal
HIV test	Negatif

The patient received 6 cycles of the CHOP regimen (cyclophosphamide 750 mg/m^2, adriamycin 50 mg/m^2, vincristine 2 mg and Prednisolone 100 mg by day for five consecutive days in each cycle). After one year follow-up, the endoscopic and CT control showed tumor disappearance (Figure 5).

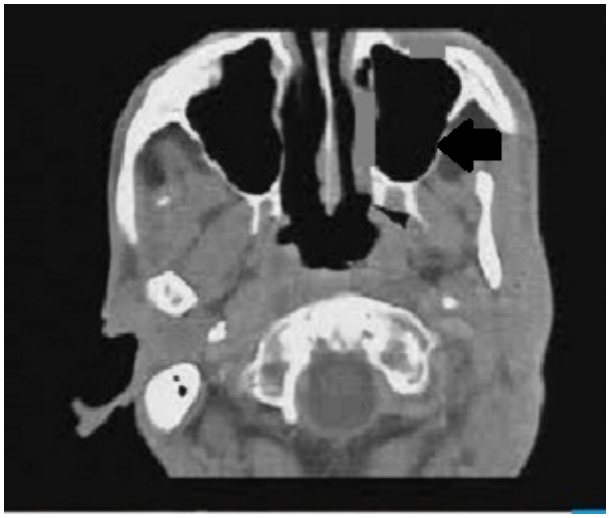

Figure 5. CT Image post chemotherapy

3. Discussion

In lymphoma, the tumoral invasion is frequent in the head and neck region. Cervical lymph-node involvement is found in 39–72% of cases [6]. On the other hand, Waldeyer's ring represents the most common location for extranodal involvement [7]. However, paranasal sinuses are considered rare locations for extranodular lymphomas (0.2–5% of cases) [8]. The maxillary sinus is the most common site of involvement, followed by the ethmoid, sphenoid and the frontal sinus. They can occur at any age, but usually there is a prevalence of middle to older ages, with a male predominance [9]. In Caucasian populations, LBCL is the prevalent type, while in Asia and South America nasal NK/T lymphoma predominates [8]. The symptoms are non-specific and depend greatly on the exact location and extent of the lesion. Revelation is mostly by classic sinonasal functional signs: nasal obstruction, mucopurulent rhinorrhea, epistaxis and anosmia. However, these signs were no specific but the unilateral character can suspect a malignant lesion. Sometimes a considerable number of patients present non-rhinologic signs due to tumor volume (endo-oral swelling, exophthalmus, diplopia, neurologic deficit) [10]. Radiographically, CT scan is the most useful preoperative study in detailing the initial dimensions of the mass, detecting a locoregional extension or a lymph node metastasis [11]. CT images show homogeneous unilateral sometimes with spontaneous uptake or heterogeneous opacity, with frequent bone destruction. These findings are not specific to lymphoma and a malignant neoplasm must be suspected [12]. Histopathology and immunohistochemistry examination should be performed to confirm the diagnosis and histological grading of lymphoma. Also, treatment varies depending on the stage of lymphoma. Paranasal lymphomas have a poor prognosis, which is usually worse than that associated with lymphomas in other sites in the body [13]. Generally, patients with a NHL tumor of ≥ 5 cm in diameter in the head and neck appear to have a worse prognosis than those with smaller tumors [14,15]. Chemotherapy and radiotherapy are the two main pillars of therapy of NHL depending to stage and prognosis factors. Radiotherapy is used in localized forms with large tumor volume [10,16]. Chemotherapy is the treatment of choice in disseminated forms and the most usual protocol being is CHOP. Some authors recommend systematic complementary craniofacial radiotherapy, even in disseminated forms [9,16]. Early diagnosis will improve the prognosis. A continuous monitoring and close follow up is recommended.

4. Conclusion

Despite NHL of maxillary sinus is rare, otolaryngologists should keep this diagnosis in mind within the range of tumours of the paranasal sinuses. Prognosis depends on histologic type, Ann Arbor grade. Early treatment will improve the prognosis and the patient survival.

Conflict of Interest

The authors have no competing interests.

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