

NASOPHARYNGEAL NON HODGKIN LYMPHOMA WITH INTRACRANIAL EXTENSION

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ABSTRACT

Non-Hodgkin's lymphoma (NHL) is the second most common neoplasm found in the head and neck region after squamous cell carcinoma. Within this region, primary NHL of the nasopharynx is rare. We reported a 65-years-old woman patient with progressive worsening of chronic sinusitis with nasal obstruction, mucopurulent rhinorrhea, headache and neuralgia of the right hemiface. On physical examination, a bulky mass was observed in the nasopharynx. Neck computed tomography revealed a diffuse soft tissue mass with mild peripheral enhancement involving both nasopharyngeal walls. The cervical-facial MRI showed a bulky expansive lesion occupying the entire nasopharyngeal lumen, involving the both nasal hemifossa, ethmoidal cells, sphenoidal sinus, cavernous sinus, and pterygopalatine fossa, with intracranial extension. Biopsy revealed a diffuse large B cell lymphoma. Based on the data, chemotherapy with regiment CHOP (cyclophosphamide, hydroxydaunorubicin, oncovin/vincristine, and prednisone) was given. A good therapeutic outcome is dependent on early diagnosis and a multidisciplinary approach.

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INTRODUCTION

Non-Hodgkin Lymphoma (NHL) accounts for 5% of malignancies of the head and neck[1]. Most NHL in the head and neck arise in the extranodal lymphatic system of the waldeyer's ring. Within the waldeyer's ring, the nasopharynx is the second most common site of disease after the tonsil. Primary nasopharyngeal lymphoma is much less common occurring in only 8% of all NHL of the head and neck, usually high grade, aggressive and has a strikingly poor prognosis compared to other extranodal lymphomas [2]. Therapeutic progress has been achieved through the more frequent use of chemotherapy associated with radiotherapy [3]. The purpose of this work, through a clinical case of NHL of the nasopharynx of phenotype B with intracranial extension, is to describe the clinical, histological, therapeutic and prognostic aspects of this disease.

Case report

A 65-years-old woman with a history of type 2 diabetes mellitus and high blood pressure presented with progressive worsening of chronic sinusitis with nasal obstruction, mucopurulent rhinorrhea, headache and neuralgia of the right hemiface of 8 months duration. Five months before his hospital admission, she had complained of right otalgia, with tinnitus and hearing impairment. No fever, pruritus, night

sweat or weight loss were noted. Nasal endoscopy revealed obstruction of both posterior choanae by a huge nasopharyngeal mass accompanied with necrotic materiel. Lymphadenopathies were detected in the right lateral cervical, and supraclavicular groups. Neurological examination on admission revealed right trigeminal nerve dysfunction, hypoesthesia of the right hemiface, and limited eye movement. Neck computed tomography revealed a diffuse soft tissue mass with mild peripheral enhancement involving both nasopharyngeal walls (Fig.1). The cervical-facial MRI showed a bulky expansive lesion occupying the entire nasopharyngeal lumen, involving the both nasal hemifossa, ethmoidal cells, sphenoidal sinus, cavernous sinus, and pterygopalatine fossa, with intracranial extension (fig.2). Blood count and biochemical tests were all normal. The chest CT did not show a pulmonary focus. Under general anesthesia, an endoscopy was performed and several biopsies were performed in the nasopharyngeal formation. The histopathological examination of the material obtained with the biopsies demonstrated a diffuse proliferation of large lymphoid cells with quite abundant basophil cytoplasm and pale perinuclear ring, oval nuclei with dispersed chromatin, and one or more nucleoli (A, hematoxylin-eosin 400x). Immunohistochemical reactions were performed and the neoplastic cells were diffusely positive for CD20 (B) and weakly positive for BCL6 (C). The proliferation index, evaluated with K167 (D), NUM1 clone, washigh: about 87% (Fig.3).

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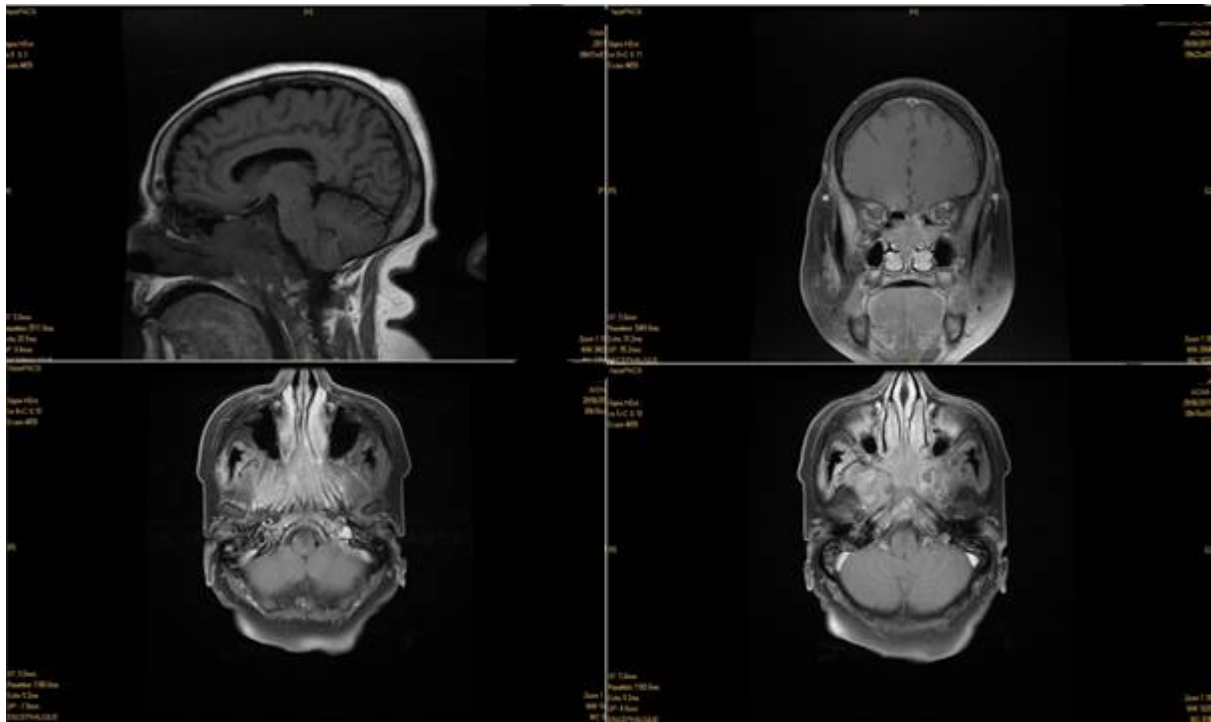


Figure 1 Magnetic resonance (MR) images showing nasopharyngeal mass with intracranial extension

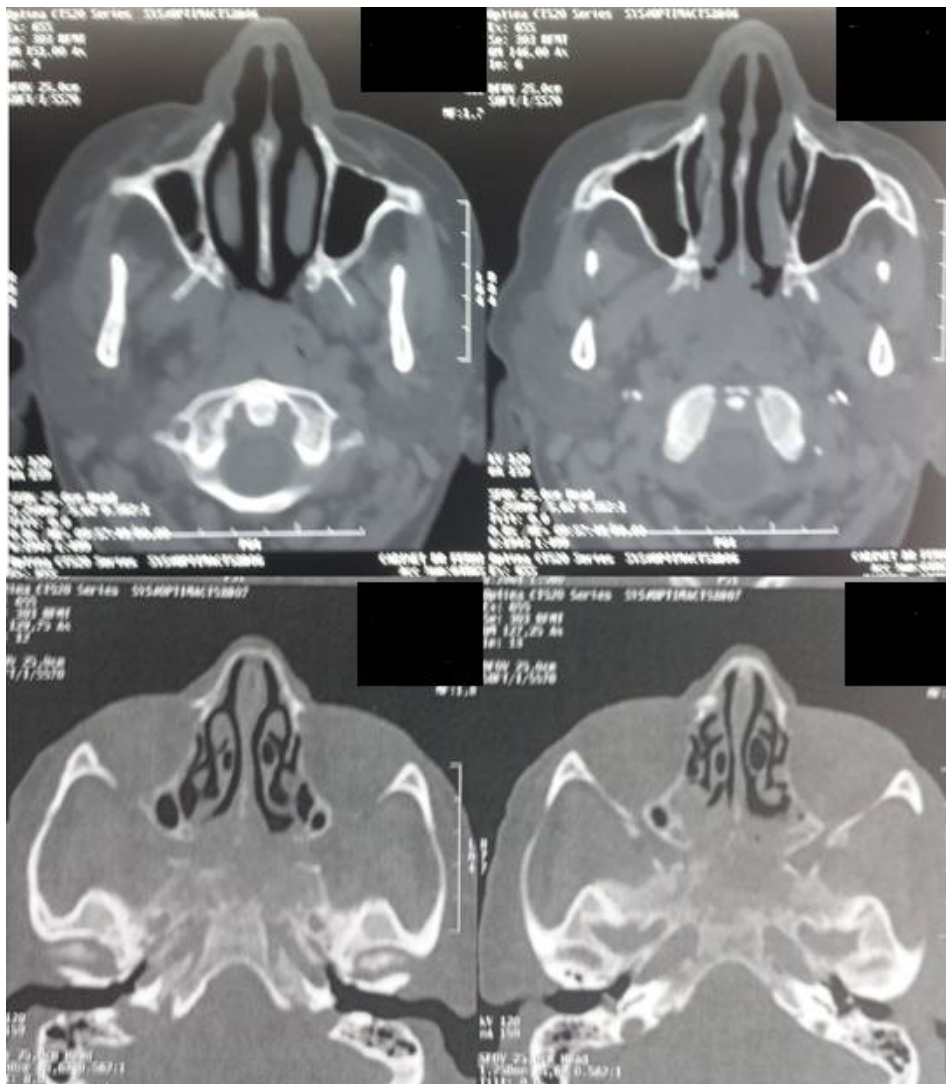


Figure 2 Head and neck CT scan shows nasopharynx NHL with extension into the sinus paranasalis, causing total obliteration nasopharyngeal cavity

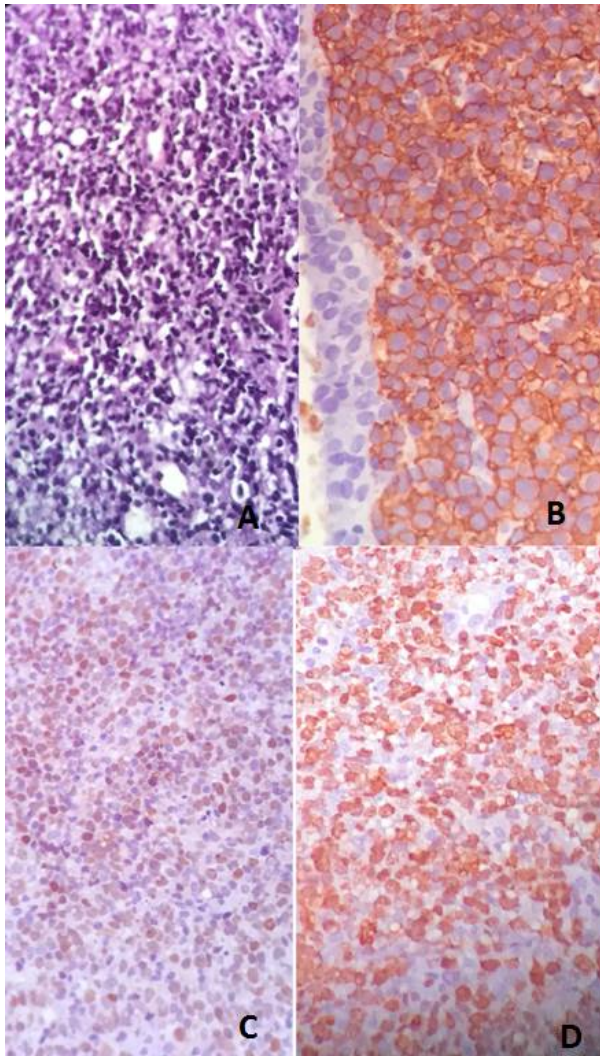


Figure 3 Histopathologic findings of nasopharyngeal mass: **A** HE staining demonstrating diffuse large B-cell non-Hodgkin lymphoma with lymphoid cells of medium to large size, most of the nucleolus located near the nuclear membrane ($\times 400$). Immunohistochemical analysis demonstrating strong positivity for CD20 **B**, BCL6 **C** and Ki67 expression **D** in tumor cells.

The diagnosis of diffuse large B cell lymphoma (DLBCL) was then retained. Based on the data, chemotherapy with regimen CHOP (cyclophosphamide, hydroxydaunorubicin, oncovin/vincristine, and prednisone) was given. However, we could not evaluate the treatment response because the patient sadly passed away after receiving the first cycle of chemotherapy.

DISCUSSION

NHL is a group of histologically and biologically heterogeneous clonal malignant diseases arising from the lymphoid system. Primary extranodal site is the form of presentation of approximately 25-40% of all NHL. After the gastrointestinal tract, extranodal NHL arising in the head, the neck (the Waldeyer's ring tonsils being the commonest affected site) follows, accounting for 10% of all NHL and 1% of head and neck tumors, the majority of them being squamous cell carcinomas or one of their variants [4]

Primary NHL of the nasopharynx corresponds to a small proportion (10–28%) of Waldeyer's lymphomas with an aggressive clinical course and poor outcome even in the early stages. Its incidence is higher in Asian patients than in

Western patients. Men are more often affected than women, and most patients present between the sixth and eighth decades of life. Patients with an underlying immunodeficiency disorder tend to present at a younger age [5].

NHL's symptomatology is generally not very specific. Clinical signs of most commonly encountered complaints are nasal obstruction, auditory disorders and cervical lymphadenopathy. Involvement of cranial nerves is extremely rare[6], which is not the case in our patient whose neurological examination showed right trigeminal nerve dysfunction, hypoesthesia of the right hemiface, and limited eye movement.

According to the literature, the LMNHs belong to the B or T-natural killer (T / NK) lines. They are part of the mature Lymphoproliferative syndromes. The B phenotype predominates for non-Hodgkin's lymphomas of the cavum (60%) whereas the T / NK type predominates in the nasal cavity [3]. Epstein-Barr virus was incriminated in the genesis of non-Hodgkin's lymphomas of the nasopharynx but only of type T, as immunohistochemistry or molecular biology reveals it almost constantly for T-lymphomas but not for B [6].

There is no standard treatment for non-Hodgkin lymphoma of the nasopharynx[4]. It often involves radiotherapy for localized and low-grade tumors, while more extensive and aggressive tumors are associated with therapeutic association. Exclusive irradiation may be the treatment of choice for low-grade localized lymphomas in some studies, the combination of chemotherapy (three to four courses of CHOP) and radiotherapy resulted in good outcomes in both overall survival and relapse rates [6]. The addition of Rituximab to CHOP-type chemotherapy is essential in the case of anti-CD-positive antibodies to immunohistochemistry. In the presence of unfavorable prognostic factors, the most intensive chemotherapies are indicated: ACVBP (doxorubicin, cyclophosphamide, vindesine, bleomycin and prednisone), CHOEP (CHOP and etoposide), mega-CHOEP or the like[7]. Based on the data, suggested that this patient suffered from NHL stage IV, we treated with supportive treatment and chemotherapy regimen CHOP. However, we could not evaluate the treatment response because the patient sadly passed away after receiving the first cycle of chemotherapy.

CONCLUSION

NHL nasopharynx is a rare incidence representing about 10% of all NHL and 1% of head and neck tumors. The diagnosis will always involve a biopsy to type the lymphoma and to determine certain prognostic factors related to the tumor cell. A good therapeutic outcome is dependent on early diagnosis and a multidisciplinary approach.

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