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Case Report

Olfactory Neuroblastoma: A Case Report

El Messaoudi Lina[1, 3*], Al rajab Sundos[1, 3], Dani Bouchra[1, 3], Bencheikh Razika[2, 3], Benbouzid Mohamed Anas[2, 3], Oujilal ABdelhamid[2, 3], Leila Essakalli Houssyni[2, 3]

[1]Resident Physician in Otorhinolaryngology, Department of Otorhinolaryngology, Head and Neck Surgery, Ibn Sina University Hospital, Rabat, Morocco
[2]Professor of Otorhinolaryngology, Department of Otorhinolaryngology, Head and Neck Surgery, Ibn Sina University Hospital, Rabat, Morocco
[3]Faculty of Medicine and Pharmacy of Rabat, Mohammed V University, Rabat, Morocco

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***Corresponding author:** El Messaoudi Lina

Abstract

Olfactory neuroblastoma is a rare sinonasal malignant tumor, characterised by clinical polymorphism and local agressivity. In spite of the surgery and the radiotherapy, the forecast remains reserved in the long term.

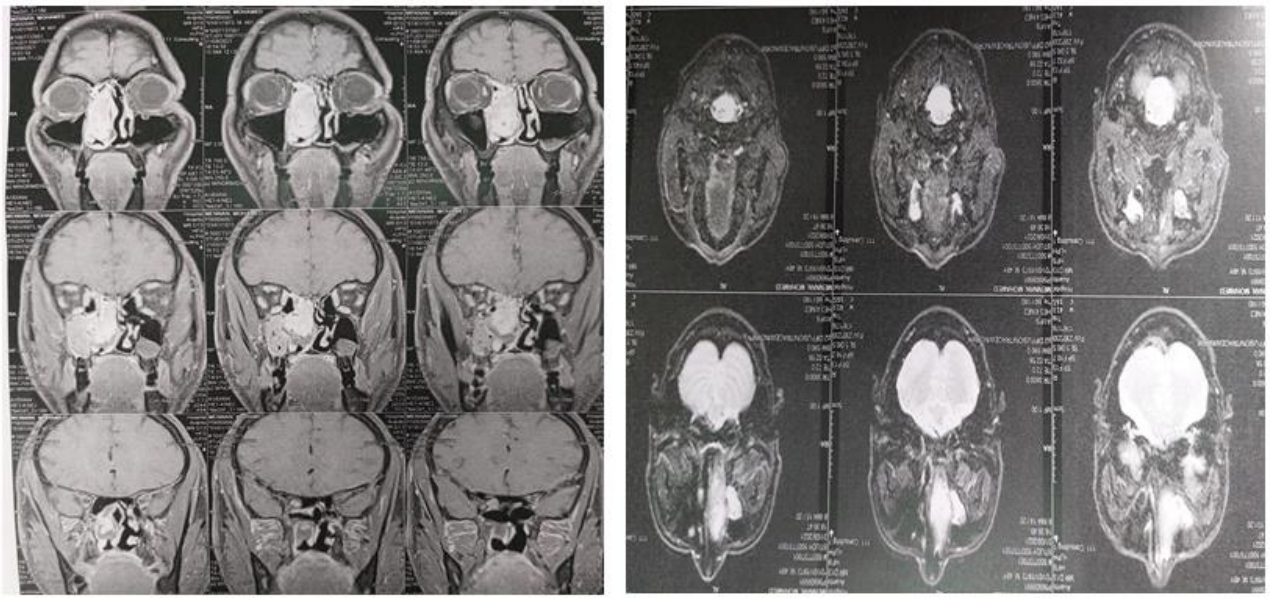
Keywords: Olfactory neuroblastoma: ON – Imagery – Immunohistochemestry – Surgery – Radiotherapy.

INTRODUCTION

Olfactory neuroblastoma (ONB) is a rare malignant tumor arising from the olfactory epithelium of the nasal cavity and the paranasal sinuses. The anatomic origin in the superior nasal cavity leads to nonspecific symptoms that make early diagnosis difficult. The most common symptoms of ON are unilateral nasal obstruction (70%), and epistaxis (50%). Other symptoms include headaches, pain, excessive lacrimation, rhinorrhea, anosmia. ON originates from olfactory epithelium; however ON rarely causes anosmia. The CT and MRI allow a precise locoregional assessment. The diagnosis is antomopathological and treatment is based on surgery and radiotherapy. This tumor poses problems of management being given the most often late diagnosis and significant extension that can hinder resection complete tumor; The present study reports the case of a patient with a mass in the nasal cavity who was treated by combined surgical excision and RT.

CASE REPORT

A 59-year-old male was admitted in the Department of Otorhinolaryngology, Head and Neck Surgery, Ibn Sina University Hospital, Rabat, Morocco in august 2020 for total left nasal obstruction progressively evolving for about one year, recurrent micro epistaxis, without associated ophthalmic signs; Patient history revealed no systemic diseases and no previous sinonasal surgery. The clinical endoscopic examination found a presence of a tissue process filling the entire right nasal cavity; deviating the septum to the left side causing destruction of the nasal dorsum; the cervical palpation found no palpable lymphadenopathy, a tumour biopsy was taken and a MRI of the nasal sinuses and skull was ordered. The biopsy revealed it to be a neuroblastoma tumour and the radiological exploration has objectified the presence of an aggressive nasal tumor process: filling the right nasal fossa and lysing the middle and inferior turbinates; it extends to the homolateral ethmoidal massif and bulges into the internal wall of the right maxillary sinus which is the site of fluid retention behind it extends to the choana. The therapeutic management consisted of excision by nasal endoscopic surgery. The definitive anatomopathological study of the piece was completed by immunohistochemistry which confirmed the diagnosis of neuroblastoma with a healthy margin of resection; then the patient was referred to the department of radiation oncology for further management.



Magnetic resonance imaging of the facial sinuses of our patient showing an expansive process of the right nasal cavity of tissue signal in discrete hypersignal T2;intermediate signal in diffusion b 1000; and intensely enhanced after injection of contrast product. It fills the right FN and lyses the upper and middle cones

DISCUSSION

Olfactory neuroblastoma was first described in 1924 by Berger *et al*., [2]. 1000 cases since have been described in the literature [3]. It is a rare tumor representing 1.2% of all malignant tumors nasal sinus and 3 to 6% of all tumors nasal; The increase in cases published these recent years correspond more to the improvement of diagnostic possibilities than to the increase in frequency disease [6]. In most series, this tumor also affects both sexes [7] although some authors report a slight female predominance [6]. She occurs at any age [8] but it is classically described 2 peaks frequency: between 10 and 20 years, and between 50 and 60 years [3, 5, 9]. No risk factor has been clearly identified in Literature. However, some works suggest a possible role of nitrosamines, wood dust as well as certain genetic anomalies (3p-); (17q +) [10]. Clinically, in 75% of cases, the tumor is revealed by rhino sinus signs [11], mainly like nasal obstruction and epistaxis, elsewhere it is anosmia, rhinorrhea and nasal sinus pain. It's done, the one-sidedness and the antiquity of the symptomatology of progressive worsening which should attract attention [4]. The achievement ophthalmologic with invasion of the orbit is observed in 20 to 30% (2.5) of cases leading to exophthalmos, reduced visual acuity or even ophthalmoplegia. The presence of eye signs in the foreground indicates a late stage of the disease. Pain syndrome, the presence of lymphadenopathy cervical or paraneoplastic syndrome are also rare [12]. On endoscopic examination, the tumor presents as a polypoid formation, varying in color from gray to dark red, friable, and readily haemorrhagic (2.5). The ophthalmologic examination is compulsory in front of the frequency of eye signs. The neurological examination should be systematic given the proximity of the tumor to the base of the skull and the frequent endocranial extension. Examination of lymph node areas cervical is also systematic especially as neuroblastoma is endowed with lymphophilia. CT with carrying out axial and coronal slices before and after injection of the contrast product, constitutes the examination of choice by putting in evidence a homogeneous full opacity which may contain intra tumoral calcifications, which are moderately enhanced after injection of the contrast product [5]. Specify the exact limits of intracranial extension and to screen invasion of the anterior floor (2.5); In terms of extension, Kadish proposed (in 1976) a classification clinical in 3 stages [1, 2, 14]:

- Stage A: Tumor limited to the nasal cavity
- Stage B: Tumor limited to the nasal cavity and sinuses
- Stage C: Tumor extended beyond the nasal cavities and sinuses.

This classification was modified by Morita in 1993 [12]. More recently Dulguerov [14, 15], proposed a more precise classification based on the classification TNM classification and using CT and MRI:

- T1 = Nasal and / or sinus tumor leaving a space air between the tumor and the riddled slide
- T2 = Tumor coming into contact with the riddled slide, or even the eroding.
- T3 = Extradural intracranial tumor and / or affected orbital.
- T4 = Intracranial tumor, intra dural.

The diagnosis of neuroblastoma evoked on imaging is histological [13], it is done on morphology and immunohistochemistry. This one is recommended

[13] for low-grade forms, essential [13] for high-grade forms where it eliminates certain differential diagnoses such as lymphoma, adenocarcinoma, plasmacytoma, melanoma, rhabdomyosarcoma, paraganglioma, and Ewing's sarcoma. There are no specific markers of olfactory esthesioneuroblastomas but an evocative profile highlighting the double differentiation of these lesions [16]. It shows the positivity of neuroendocrine markers, and of the S100 protein having histopronostic value, and the usual negativity epithelial markers. Treatment of neuroblastoma is considered depending on whether the tumor is operable or not. Thus, if the tumor is operable, surgery complete macroscopic and microscopic excision with safety margins followed by radiotherapy on the tumor bed and the first lymph node relays is the treatment standard with curative aim of neuroblastomas of sinus (Grade C recommendation) [13]. There is no first standard, however, any first path must meet two objectives [17]: On the one hand, the possibility of controlling all the anatomical boundaries of the tumor and riddled blade; On the other hand, the realization of a truly carcinological resection in one piece, avoiding if possible the tumor fragmentation or excision by fragmentation [17]. There are essentially three approaches: craniofacial,transfacial and nowadays more and more endoscopic endonasal surgery mainly in Kadish stages A and B [13]. The surgical approach is either transfacial performing a para-latero-nasal rhinotomy (for stages A and B of Kadish), or via the high route ub-frontal in the event of an attack on the base of the skull. If there is an orbital extension, the attitudes are extremely nuanced, but most surgical teams are currently conservative with regard to the eyeball; due to the lack of significant difference in exenteration on survival or recurrence rates. In the event of an inoperable tumor, treatment is multimodal, combining first chemotherapy, radiotherapy possibly followed by craniofacial surgery (professional consensus) [13]. Preoperative radiotherapy, although recommended by some centers, is not a standard [3, 8]. This radiotherapy targets the tumor bed as well as the lymph node areas, the irradiation dose can range from 45 to 60 Gy in the event of a large tumor volume. Olfactory neuroblastoma is a tumor with a very poor prognosis. Survival is approximately 50% at 5 years and 30% at 10 years [15]. Local and loco regional recurrences represent 60% [13], these recurrences can be early or late, thus justifying lifelong monitoring of these patients. The main prognostic factor is the clinical stage at the time of diagnosis. Age, sex and ethnicity were not found as prognostic factors. The histological grade of Hyams also appears to be a significant factor in the prognosis with 56% survival for low grades I and II versus 25% for high grades III and IV. The other factors are represented by the TNM stage and the treatment, in fact, survival in the event of a combination of surgery and radiotherapy is superior to radiotherapy alone or to surgery alone [13].

CONCLUSION

Olfactory neuroblastoma is a rare locally aggressive tumor of the nasal sinus cavities posing diagnostic difficulties and problems with therapeutic management. The treatment widely accepted in the literature remains anterior craniofacial resection followed by postoperative radiotherapy for localized stages. Endoscopic endonasal surgery seems to be increasingly indicated. Nevertheless, this tumor remains of poor prognosis and gladly recurrent.

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