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Esthesioneuroblastoma an Exceptional Malignant Tumor: A Case Report

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Abstract **Case Report**

Esthesioneuroblastoma is a rare malignant tumor developed at the expense of the olfactory neuroepithelium. It is one of tumors of the nasal cavities and has the particularity of being of slow evolution with frequent recurrences after more than 10 years. We report a case of A 24 year old patient who presented an esthesioneuroblastoma revealed by a recurrent unilateral right epistaxis, he was evaluated with a Craniocervical MRI. A biopsy was performed and the anatomopathological examination revealed an Esthesioneuroblastoma. The patient was operated in neurosurgery and he received Chemotherapy followed by concomitant radiotherapy chemotherapy. Pathological and radiological evaluation, treatement of this neoplasm are discussed.

Keywords: Esthesioneuroblastoma, malignant tumor, neuroepithelium, neurosurgery.

INTRODUCTION

Esthesioneuroblastoma is a rare malignant tumor developed at the expense of the olfactory neuroepithelium. It is one of the rarest tumors of the nasal cavity that represents less than 5% [1].

This type of tumor is often manifested by rhinological, ocular or neurological signs, that can be inaugural or secondary. The CT and MRI allow a precise locoregional assessment [2]. The diagnosis is anatomopathological and the treatment is still controversial, combining surgery, radiotherapy and chemotherapy. The choice of an optimal treatment is the main concern, in order to prevent dreadful complications and recurrences.

Esthesioneuroblastoma poses problems of management given the often late character of the diagnosis and the important extension that can hinder a complete removal of the tumor.

CASE REPORT

A 24 year old patient with no particular pathological history who presented an esthesioneuroblastoma revealed by a recurrent unilateral right epistaxis of low abundance associated with intermittent homolateral nasal obstruction, headaches, anosmia, rhinolalia, visual disturbance and right larmoiment complicated by an intracranial hypertension syndrome.

A cerebral CT scan revealed a right ethmoidonasal cerebral process with endocranial extension to the anterior stage; sphenoidal sinus, sieve blade and frontal lobe.

A craniocervical MRI revealed a large aggressive tumor process with an ethmoidal epicenter infiltrating the nasal cavities, right orbit with grade II exophthalmos with endocranial extension resulting in a compressive right frontal mass responsible of subfalcoral involvement and contralateral active hydrocephalus (esthesioneuroblastoma to be compared with anatomopathological data).

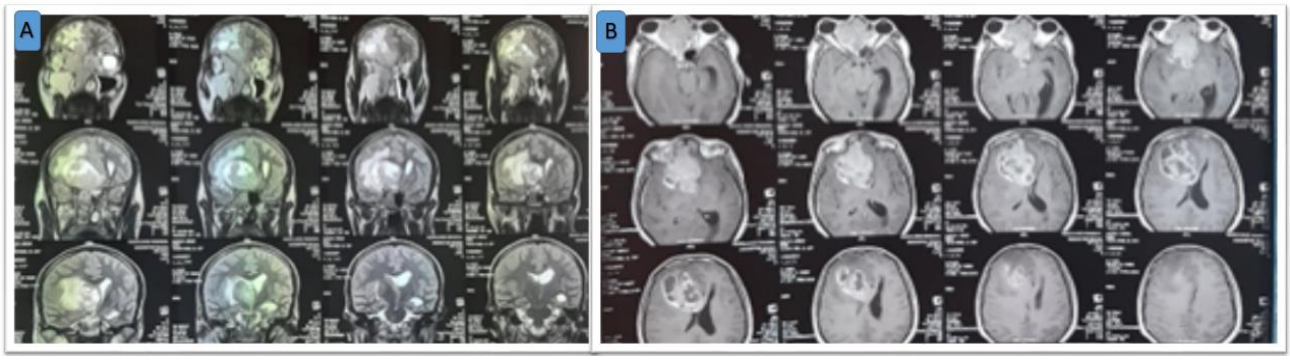

Figure 1: Craniocervical MRI; A) MRI frontal section T2 sequence, B) MRI axial section T1 sequence

The patient was operated in neurosurgery for a total macroscopic removal of the endocranial part of the tumor, and in the ORL department to remove the endonasal component of the tumor with posterior ethmoidectomy.

An exeresis was performed, which anatomopathological and Immunohistochemistry study revealed an Esthesioneuroblastoma.

A postoperative MRI revealed the persistence of the ethmoidofrontal process with orbital and endocranial extension measuring 5x6cm.

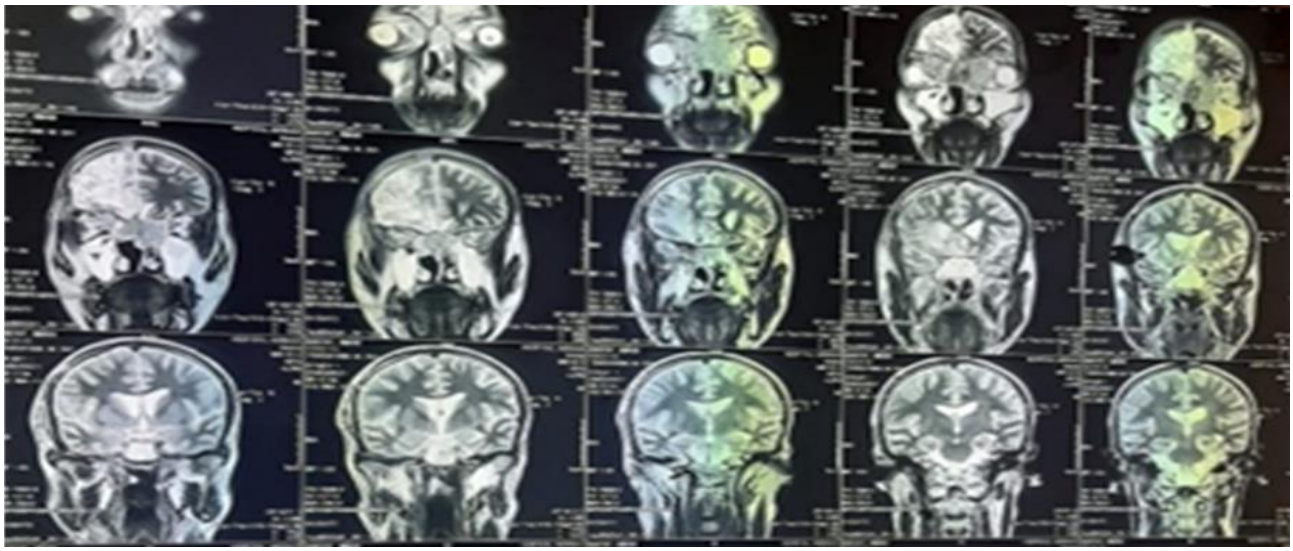

Figure 2: Postoperative craniocervical MRI frontal section T2 sequence

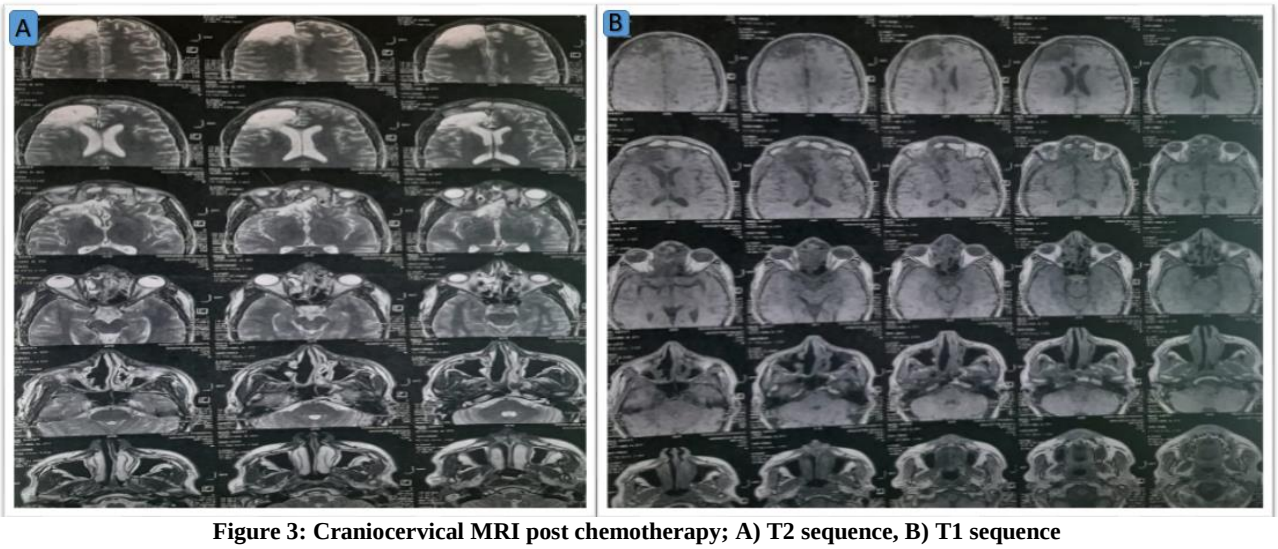

Figure 3: Craniocervical MRI post chemotherapy; A) T2 sequence, B) T1 sequence

The patient received Etoposide-Cisplatin chemotherapy (3 sessions) followed by concomitant radiotherapy chemotherapy (27 sessions of radiotherapy and 6 sessions of cisplatin chemotherapy).

An evaluation MRI revealed a right frontal cortico-cortical porencephalic cavity, surrounded by a gliosis responsible for a dilatation of the frontal horn of the homolateral lateral ventricle.

On the facial level: bilateral ethmoido-frontal lesion process measuring approximately 55x48x63mm. Hypertrophy of the inferior turbinates. Framed thickening of the maxillary sinuses.

On the complementary scan, a thickened, condensed and heterogeneous aspect of the bony cortex of the lateral masses of the ethmoid as well as the walls of the frontal sinuses with individualization of some bony sequestration probably related to a post-therapeutic bone remodelling.

The patient was referred to the ORL department for a biopsy of the fronto-ethmoidal lesion process.

DISCUSSION

ENB is a rare neuroepithelial tumor whose exact origin is still unknown [3]. The majority of studies suggest that they originate from basal cells of the olfactory neuroepithelium of the upper third of the nasal cavity [4, 5].

This tumoraffects both sexes equally [6], although some authors report a slight female predominance [7].

It occurs at any age [8] but 2 peaks of frequency are classically described: between 10 and 20 years of age, and between 50 and 60 years of age [11, 10, 9]. No risk factor has been clearly identified in the literature.

However, some studies suggest a possible role of nitrosamines, wood dust and certain genetic abnormalities (3p-); (17q+) [12].

Clinically, in 75% of cases, the tumor is revealed by rhino-sinus signs [13], essentially of the type nasal obstruction and epistaxis; elsewhere, it is anosmia,rhinorrhea and naso-sinusal pain. It is in fact,the unilaterality and the length of time of the progressively worsening symptomatology that should attract attention [14].

The diagnosis of ENB is often delayed due to a non-specific clinical presentation [5, 15]. Nasal obstruction (70%) and recurrent epistaxis (46%) are the most frequent reasons for consultation [1, 5, 15]. Orbital extension may be revealed by exophthalmos, decreased visual acuity, diplopia lacrimation or ptosis [5, 15]. Endo-sinusal extension may mimic an acute recurrent sinusitis picture [5]. Neurological signs are rare. On endoscopic examination, the tumor appears as a polypoid mass varying in color from gray to dark red. Lymph node involvement is not exceptional and is found in 20-25% of cases [3].

Imaging is essential for the diagnosis and postoperative monitoring of ENB. It allows the assessment of the tumor volume, its structure and extensions [15, 16]. The presence of calcifications, although rare, is specific [15].

On MRI, the tumor lesion is usually hyposignal T1, iso or hypersignal T2 with variable enhancement after injection of PDC [15].

Several prognostic classifications. Prognostic classifications have been proposed according to tumor extension. The most commonly accepted is that of Kadish proposed in 1976 [17].

A clinical classification in 3 stages:

- 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 cavity and sinuses.

The definitive diagnosis of ENB remains anatomopathological. It is far from being easy with the standard histological examination. The development of immunohistochemistry has been a turning point in the diagnosis of ENB.

Treatment is multimodal combining chemotherapy and radiotherapy, possibly followed by craniofacial surgery (professional consensus) [18].

Preoperative radiotherapy, although recommended by some recommended by some centers, is not a standard (10,8). This radiotherapy targets the tumor bed as well as the lymph node area, the dose of radiation can range from 45 to 60 Gy in case of large tumor volume [19].

The association of a postoperative radiotherapy is currently required [20]. It significantly reduces the risk of tumor recurrence and improves recurrence and improve long-term survival [21]. It is also indicated exclusively or in combination with chemotherapy for inoperable or locally extensive tumors [3].

For some authors, irradiation of the regional lymph nodes could be indicated immediately in cases of cervical immediately in cases of associated metastatic cervical cancer or prophylactically in the case of locally advanced tumors [22].

For others, in the absence of cervical adenopathy, cervical irradiation does not seem to modify the prognosis and may even hinder a possible lymph node curage in case of late lymph node metastasis [23].

Few studies have evaluated the role of chemotherapy in the treatment of esthesioneuroblastoma. It is reserved, alone or in combination with radiotherapy, for inoperable tumours, for those that are immediately metastatic and for tumour recurrences that are surgically inoperable [20, 21].

For some authors, neoadjuvant chemotherapy would be indicated in cases of extensive ENB, particularly intra-cranial or intra-orbital.

Despite treatment, ENB remains a malignant tumor with a poor prognosis. The mean 5-year survival ranged from 33% to 87% [21].

The prognostic factors reported in the literature are age, stage according to the Kadish classification differentiation grade according to Hyams, the presence of lymph node metastases or distant metastases, the limits of tumor removal, the tumor proliferation index, P53 overexpression and recently the overexpression and more recently the presence of a deletion at chromosome 11 [24, 25]. ENB is a recurrent and metastatic tumor. The recurrence rate has been estimated at 60% case [5].

CONCLUSION

Olfactory esthesioneuroblastoma is a rare malignant nasosinus tumor, characterized by its clinical polymorphism and local aggressiveness. Despite surgery and radiotherapy, the prognosis remains poor in the long term.

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