**Aggressive Esthesioneuroblastoma about A Case**

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**Abstract**

**Introduction:** Esthesioneuroblastomas (ENB) or olfactory neuroblastomas are rare tumors derived from the olfactory epithelium. These tumors have a propensity for local invasion into surrounding structures and distant metastasis, most commonly to the neck, lungs, and bones. We present a case of a 12-year-old child with olfactory neuroblastoma with lymph node metastasis with early recurrence. **Case Report:** It is a 12 year old child, without pathological history who presented an evolving right nasal obstruction for 8 months, permanent, associated with a rhinorrhea and intermittent epistaxis, associated with a mass of the ipsilatéral parotid and submandibular region. The clinical examination, showed a gray and pink mass occupying the entire right nasal and invaded the nasal septum and the contralateral nasal fossa, with a retro angulo maxillary mass extended to the parotid region and jugulo carotidien, hard, painful and fixed at the deep level (Figure 1).

A cervical and nasosinusian CT scan showed a tumor occupying the two nasal cavities the nasopharynx the ethmoid sinus and lysing the posterior internal wall of the right maxillary sinus and the sphenoid sinus body with invasion of the latter the two the scanner also showed the presence of cervical polyadenopathies on the right side (Figure 2).

The biopsy was performed under general anesthesia, the pathological and immunohistochemistry examination were in favor of an esthesio neuroblastoma , given the significant tumor extension, the decision was to do a chemotherapy at first, the post-chemotherapy scan showed tumor progression (Figure 3).

**Keywords:** Esthesioneuroblastoma, lymph node metastasis, surgery.

**INTRODUCTION**

Esthesioneuroblastoma (ENB), or olfactory neuroblastoma, is a rare malignant tumor arising from the olfactory sensory neuroepithelium, which accounts for 2%-6% of all primary sino-nasal malignancies [1, 2]. ENB was first described by Berger and Luc in 1924. No risk factor is currently recognized for this type of tumor, ENB has a bimodal distribution with no gender predilection and tends to occur around the second and sixth decade of life; however, sporadic cases have been reported in children younger than 10 years [3-5].

The natural history of ENB is varied and heterogenous, with some tumors demonstrating annindolent course and others exhibiting a highly aggressive behavior characterized by local recurrences and/or distant metastasis [3].

We present a case of a 12-year-old child with olfactory neuroblastoma with lymph node metastasis with early recurrence.

**CASE REPORT**

It is a 12 year old child, without pathological history who presented an evolving right nasal obstruction for 8 months, permanent, associated with a rhinorrhea and intermittent epistaxis, associated with a mass of the ipsilatéral parotid and submandibular region. The clinical examination, showed a gray and pink mass occupying the entire right nasal and invaded the nasal septum and the contralateral nasal fossa, with a retro angulo maxillary mass extended to the parotid region and jugulo carotidien, hard, painful and fixed at the deep level (Figure 1).

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**RESULTS**

The child then underwent transfacial surgery with parotid lymph node dissection and groups Ib, II, III et V right (Figure 4), he was then referred for radiotherapy. Three weeks after the operative procedure
the child presented a recurrence of tumor at the level of the operated lymph node sites and at the level of the contralateral side, the child died during the assessment.

**DISCUSSION**

ENB is a rare malignant tumor of the nasal sinus cavities. Its development is thought to originate from olfactory neuroepithelial cells located at the top of the olfactory slits.

These tumors usually present during the sixth and seventh decades, but cases have also been reported in children and in elderly subjects over the age of 80 years [5, 7]. The absence of significance prognostic of these factors on survival, as reported in the literature, is probably due to the very small sample sizes of published series. Similarly, no risk factor for the
pathogenesis of these tumours has ever been identified [8].

Its mode of disclosure is most often nasal obstruction (70%) or unilateral epistaxis (46%) [9]. In some cases, it can manifest as headache, rhinorrhea, anosmia, lacrimation or visual disturbances associated with sinus symptomatology (8-10% of cases) [9]. In our case, the initial symptomatology was the simultaneous appearance of nasal obstruction and a laterocervical mass.

ENB tends to be locally aggressive and metastasizes distally, resulting in decades-delayed recurrences [3, 4, 8, 10-13] with metastatic disease around 20% of the time but this statistic has been reported as high as 48% [7, 8, 14-20]. The typical sites of metastasis are cervical lymph nodes, lungs, and bones [14, 21, 22]. Less common sites are the liver, mediastinum, adrenal gland, ovary, spleen, parotid glands, central nervous system, and spinal epidural space [14]. Along with metastasis, patient may present with SIADH (syndrome of inappropriate antidiuretic hormone) with dilutional hyponatremia or excess ectopic adrenocorticotropic hormone production leading to Cushing syndrome, although these are rare [23]. In our patient there were parotid metastases then cervical lymph nodes united then bilateral, ENB staging can be evaluated according to two classifications: the Kadish classification modified by Morita and the Duguerov TNM classification [9, 22].

Computed tomography (CT) and magnetic resonance imaging (MRI) with and without contrast is the first line for evaluation of ENBs [3, 14, 24-27]. Before imaging, it is imperative to obtain a complete medical history, thorough physical examination, and a full record of any other comorbid medical conditions that may alter the treatment plan [14]. The first step is to obtain a full assessment by an otolaryngologist to determine the extent of the disease as well as provide a diagnostic biopsy for histologic grading [14]. A full neurologic and ophthalmologic examination before any extensive surgery can provide a baseline to determine any residual side effects from treatment [14]. Typically, imaging studies show one of the most characteristic findings: a dumbbell-shaped mass extending across the cribriform plate [23, 3, 25]. Where as the waist of the dumbbell is at the cribriform plate, the upper portion is in the anterior cranial fossa and the lower portion is in the nasal cavity [23, 3, 25, 28]. The presence of peritumoral cysts at the tumor-brain interface is another characteristic imaging feature of ENBs [23, 28]. The CT scan is a helpful to identify the lesion but more importantly, it can depict bony erosion of the cribriform plate, orbit, and air sinuses [3, 25, 26]. Typically, on CT, a homogenous mass with necrotic non enhancing areas is observed [3, 25, 26]. The MRI scan helps indicate the extent of soft tissue involvement [3]. It is ideal to obtain both scans before any surgical treatment because ENBs can have significant soft tissue involvement that cannot be properly identified on CT scan only. Contrast MRI can help differentiate the tumor from obstructed secretions in the paranasal sinuses, determine meningeal and extradural invasion, and identify the extent of perineural involvement [3, 25, 26].

Histological diagnosis is not difficult if the tumor is well differentiated. It is then composed of small homogeneous and uniform cells with round or oval nuclei, with rosette or pseudo-rosette formations. When the tumor is little or undifferentiated, composed of small hyperchromatic and anaplastic cells, the diagnosis requires immunohistochemical studies with the search for a positive labeling for synaptophysin (> 90%) [29], NSE (neuron specific enolase, > 90%) [29], chro-mogranin, CD56, and lack of expression of muscle markers (desmin, vimentin, actin, myogenin) and markers of epithelial cell differentiation (cyto-keratin) or common leukocytic antigen. The diagnosis sometimes requires the use of electron microscopy which reveals very dense neurosecretory granules in the cytoplasm and nervous processes [29]. Hyams et al., [30] classified ENB into four histopathological stages (I to IV) based on the preservation of lobular architecture, mitotic index, nuclear polymorphism, fibrillar matrix, rosettes and lanecrosis.

There is no agreed standard-of-care treatment algorithm for ENBs. Recent technological advances, improved surgical technique, and better understanding of the disease process has forced physicians to constantly develop new treatment plans, which has made it difficult to assess the effectiveness of each individual treatment [14]. Recently, the interest has shifted from subtotal and total resections to endoscopic transnasal/endonasal procedures with adjuvant radiotherapy or stereotactic radiosurgery. Adjuvant use of radiation and chemotherapy has also shown some benefits. Indeed, surgery remains the first-line treatment for these tumors, and appears in meta-analyses as an independent factor of progression-free survival and overall survival [32]. The surgical technique depends on the initial tumor stage. For localized tumors, the transfacial route is currently preferred [32]. However, the development of endoscopic surgery over the past decade has enabled comparable carcinological results to be obtained, limiting surgical morbidity and the duration of hospitalization, in particular for stage T1 or T2 tumors [33]. In meta-analyses, endoscopic resection seems to have better results in terms of overall survival than transfacial surgery. These results should be predicted with caution as studies using transfacial surgery are significantly older than those using endoscopic surgery. This fact may partly explain the more modest results [31].

In the event of a locally advanced tumor (invasion of the orbit or the anterior cerebral fossa), a
mixed surgical approach, by transfacial or endoscopic and neurosurgical route is preferable. Despite the absence of studies of a satisfactory level of evidence, adjuvant radiotherapy is currently considered as standard treatment after surgery, whether histologically complete or not [34, 7].

On the other hand, exclusive radiotherapy should not be used, except in the event of an absolute contraindication to surgery, due to the significantly lower local control and specific survival rate compared to the joint approach [34, 7].

Chemotherapy has shown promising results in patients with high-grade (Hyams grade III or IV) tumors compared with low-grade tumors [14, 36]. In patients with high-grade stage C ENB, adjuvant chemotherapy after complete resection has been beneficial [14, 35]. However, lack of a standardized treatment regimen and small sample sizes in previous studies have made it difficult to provide definitive recommendations for treatment [14]. Theoretically, the similar chemotherapy regimens used for neuroblastomas, small-cell lung carcinoma, and primitive neuroectodermal tumors have been used for ENBs because of their similar histologic and ultrastructural features [12, 21, 36].

CONCLUSION

ENB is a rare malignant tumor originating in the olfactory neuroepithelium that can present with intracranial involvement; strict cases require a multidisciplinary treatment plan. Given its malignant histopathology, metastases are a concern; the most common area for metastasis is the cervical lymph nodes. Thus, we recommend performing head and neck imaging at the very least for appropriate staging. Tumors are chemosensitive and radiosensitive and the literature supports both neoadjuvant and adjuvant therapies with surgery to reduce the tumor burden and provide symptomatic relief from compression of adjacent structures.

Declaration of links of interest

The authors declare that they have no links of interest.

REFERENCES


