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Trans-Sinus Approach for Total Resection of Esthesioneuroblastoma Recurrence: Case Report and Review of the Literature

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Abstract

Background: The Esthesioneuroblastoma (ENB) is a rare tumor of the nasal cavity derived from the olfactory neuroepithelium, represents only 3-6% of all cancers in the nasal cavity and paranasal sinuses. The rarity of ENB has made difficult to accurately assess survival and prognosis; the Hyams' histologic grading and Kadish staging system are the best-studied factors correlating with prognosis, and treatment planning. Metastases are found up to 30% of cases, being the leptomeningeal spreading extremely rare. A multidisciplinary approach of these patients is mandatory, with the craniofacial surgery with or without radiotherapy being the gold standard for the treatment.

Methods: We present a rare case of a patient with diagnosis of ENB with sagittal venous sinus metastasis during his follow-up; describe the surgical and multidisciplinary approach for this case.

Results: The patient underwent a central fronto-parietal craniotomy with complete trans-sinus resection of the lesion, the postoperative period was uneventful and his neurological examination remained normal.

Conclusions: We concluded that long-term follow-up, with aggressive treatment of the recurrent tumor or regional metastasis, are necessary to improve the quality of life of the patients.

Keywords: Esthesioneuroblastoma; Sagittal sinus metastases; Trans-sinus approach

Introduction

Esthesioneuroblastoma (ENB) or Olfactory Neuroblastoma (ONB) is a rare malignant tumor of the superior nasal cavity derived from olfactory neuroepithelium that was described for the first time in 1924 [1,2]. The incidence is 0.4/million/year. It is responsible for approximately 6% of cancer cases in the nasal cavity and paranasal sinuses [1,3].

ENB has often been misdiagnosed with other neoplasms, such as sinonasal undifferentiated carcinoma, amelanotic melanoma, nasopharyngeal carcinoma, paraganglioma, small cell neuroendocrine carcinoma, atypical carcinoid, embryonal rhabdomyosarcoma, Ewing's sarcoma, lymphoma, extracranial meningioma, chordoma, desmoplastic round-cell tumor, pituitary adenoma, and metastatic small cell lung cancer. Uncertainty about the precise histologic origin of ENB has led to the use of various names for this tumor, but the only two terms used in recent published literature is esthesioneuroblastoma and olfactory neuroblastoma [1,4].

The Hyams' histologic grading and Kadish staging system are the best studied factors correlating with prognosis, and thus useful for treatment planning [5,6].

Surgical resection of ENB is usually combined with postoperative radiotherapy due to the high risk of local recurrence. Novel approaches include Endonasal Endoscopy Resection (EER), allowing efficient local control and is associated with lower morbidity [1,7,8]. Adjuvant chemotherapy studies have proved been useful, especially in C stage of Kadish with high grade of ENB. Cisplatin-based chemotherapy, usually with Etoposide, is an accepted mode of treatment in advanced, recurrent, especially high-grade cases, but also non-platinum schemes (irinotecan, docetaxel, doxorubicin, ifosfamide, vincristine) were described to be effective [9-11].

ENB is locally aggressive and can metastasize by lymphatic and hematogenous routes. The cervical lymph nodes are the most common site of metastasis. It can spread via nasal mucosa in all directions, involving the paranasal sinuses, nasal cavities and cross the cribriform plate and involving even the brain [1,12-14].

We present a case of patient treated at the National Institute of Cancer (INCan), Mexico, with a rare metastatic ENB to middle third of the superior sagittal sinus, and describe the surgical approach and short-term outcome.

Case Report

A 51-year-old man admitted in the National Institute of Cancer (INCan), Mexico, in the neuroscience clinic, with history of progressive nasal obstruction, and intermittent epistaxis during the last 2 months. A painless polypoid mass of 2 × 3 centimeters of diameter was seen through both nostrils. His vision and eye movements were normal in both eyes. Paranasal Computed Tomography (CT) scan confirmed this mass filling the nasal cavity. Although this mass was extended to bilateral and ethmoid sinuses, frontal sinuses and invaded the antero-inferior surface of the sphenoid sinus, consider as Kadish C. Intranasal biopsy with local anesthesia was performed, and the histopathological examination revealed ENB. The patient was scheduled in the OR, and the tumor was totally removed through a subfrontal approach, frontal craniotomy and additional lateral rhinotomy.

Intensity Modulated Paranasal Radiation therapy was done, 70 Grays during 35 fractions, and concomitant weekly chemotherapy with 50mg/m2 CDDP (Cisplatin). Frontal sinuses and meningeal involvement recurrence was seen, five months after the first surgery. Second surgery included totally removed through the subfrontal approach with lateral rhinotomy and medial left maxillary osteotomy, without findings of brain involvement during the operation. Nine months later a left lateral neck mass was identified and a fine needle aspiration was performed, the histopathological diagnosis was same, and the patient was scheduled for an extensive neck dissection surgery. Whole cervical radiotherapy was done in 54 Grays during 18 fractions. The patient present 23 months later a left parotid growing tumor with same histological origin and a left radical parotidectomy was carried out. A new lesion was identified two months later, in the middle third of the SSS (superior sagittal sinus) (Figure 1), radio-oncologist consider it was not able to be treated, cranial angiotomography shows partial occlusion and collateral venous drainage, we decide under patient confirmation and good medical status, develop a central fronto-parietal craniotomy with complete trans-sinus resection of the lesion (Figure 2), the postoperative period was uneventful and his neurological examination remained normal. He was discharged to home on fifth day postoperatively. He was live without neurological deficit (Figure 3).

Discussion

ENB is a rare tumor of the nasal cavity that is derived from the olfactory neuroepithelium. Berger described it in 1924 [2]. This tumor has often been referred to as an olfactory neuroblastoma, although the exact cell of origin has not yet been proved [3]. The olfactory mucosa arises from a condensation of ectoderm in the embryo, and it normally matures to show supporting epithelium, sensory neurons and basal reserve cells, and is thought that ENB arise from the mitotically active basal cells that give rise to neuronal and sustentacular cells [4]. ENB represents only 3-6% of all cancers in the nasal cavity and paranasal sinuses [3].

Although ENB is uncommon, some consistent features of his disease have been established: it has approximately equal sex distribution, it affects a wide range of age groups, with a bimodal distribution, but recent SEER data supports an unimodal distribution ranging from 45 to 56 years of age; and, overtime, it can locally extend to involve the surrounding paranasal sinuses, cribiform plate and orbits [1,15].

ENB has no specific symptoms, related to the local extension, unilateral nasal obstruction (53-100%), epistaxis (10-52%), headache (10-20%), and hyposmia/anosmia (6-35%); with extension of disease outside the nasal cavity and paranasal sinuses, symptoms of orbital and cranial involvement can manifest (20%) [1,16].

The rarity of ENB has made accurate assessment of survival and prognostic strength of various staging systems difficult [3]. Kadish proposed a system in which for group A, the tumor is limited to the nasal cavity, group B, tumor is localized to the nasal cavity and paranasal sinuses; and for group C the tumor extends beyond the nasal cavity and paranasal sinuses, including cribiform plate, skull base, orbits and intracranial compartment [6]. Morita proposed a modification to the initial 3-tier Kadish staging system, that included an additional stage for patients who had distant metastases at the time of diagnosis (stage D) [10]. Nodal status has been reported to be one of the most important prognostic factors in survival, a finding that is consistent with the clear impact in disease specific survival [17]. A weakness cited in the Kadish system was that it did not allow for stratification of patients with pathologic adenopathy. Dulguerov proposed a TNM style of staging system for ENB that allowed for the inclusion of nodal status [18].

According to Hyams a distinction between low grade (stage 1 and 2) and high grade tumors (stage 3 and 4) is made. The graduation is based on defined factors such as lobular architecture, neurofibrillary background, rosettes, nuclear polymorphism, mitosis, necrosis and calcification [5]. Insufficient responses to initial therapy, local recurrence and higher distant metastasis rates are reported to occur more frequently in patients with high-grade tumors [5,8].

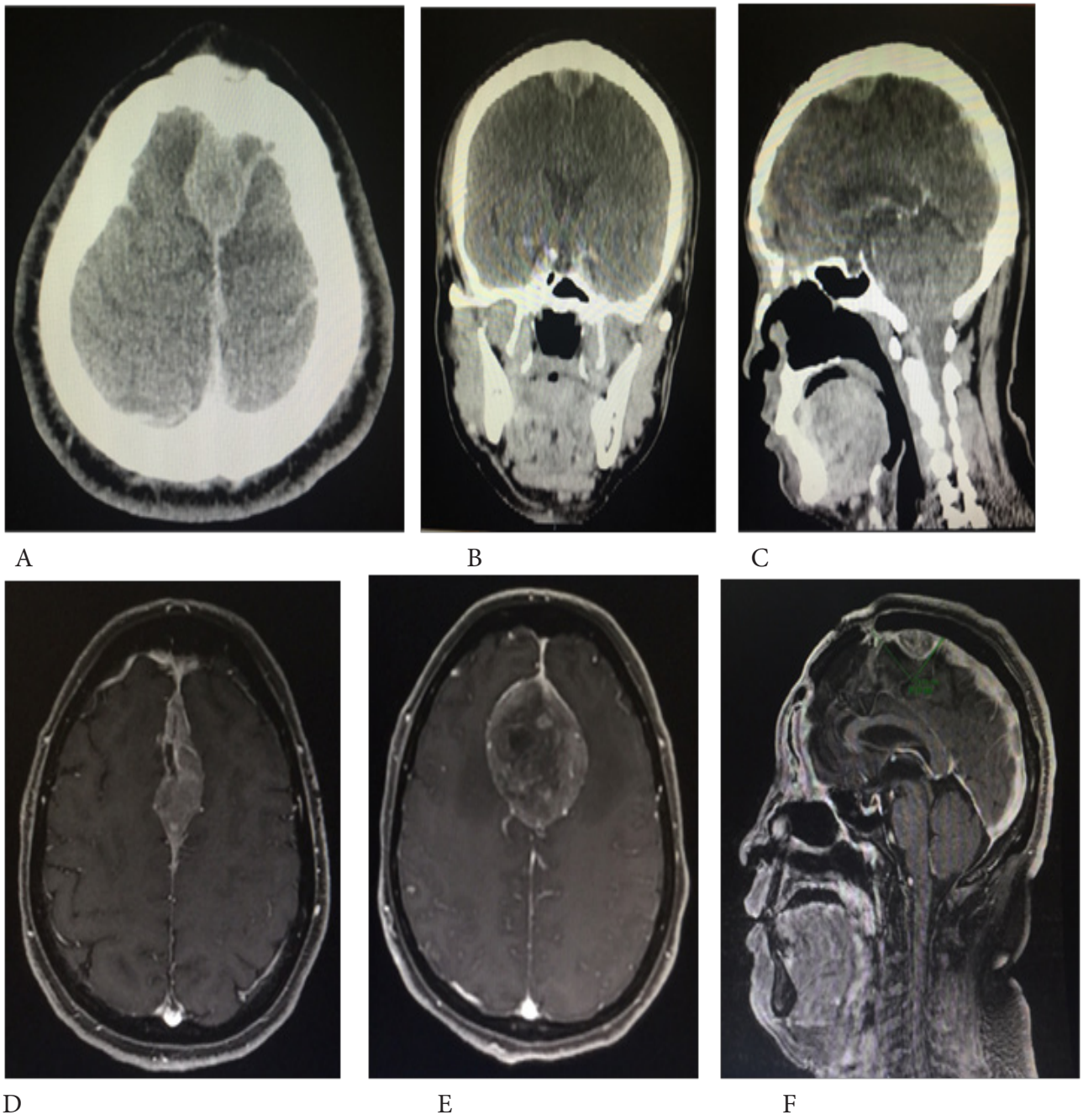
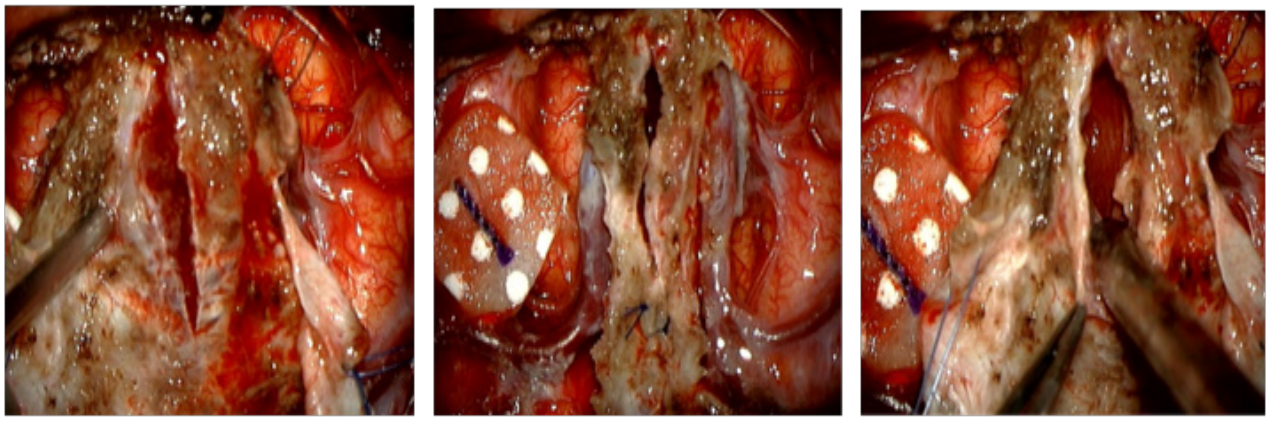
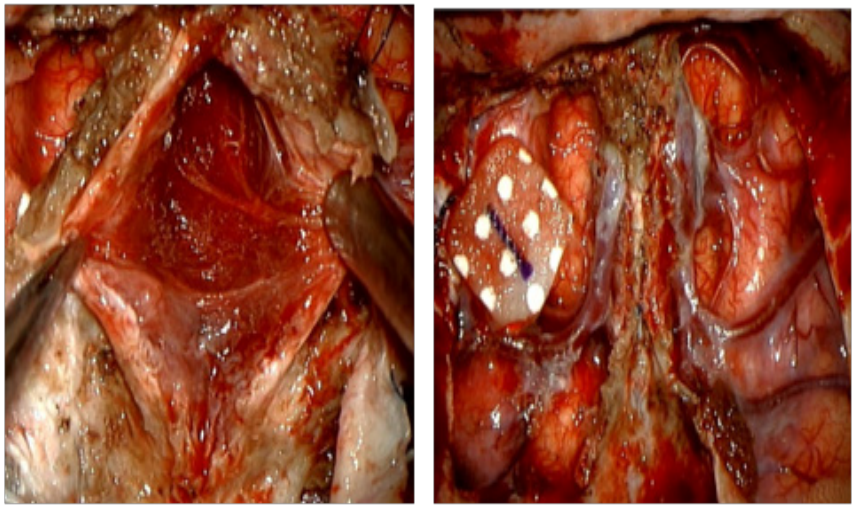


Figure 1: A-C Brain CT scan showing intrasinus metastatic ENB. D-F Contrast enhanced MRI showing metastatic ENB in the middle third of the SSS.

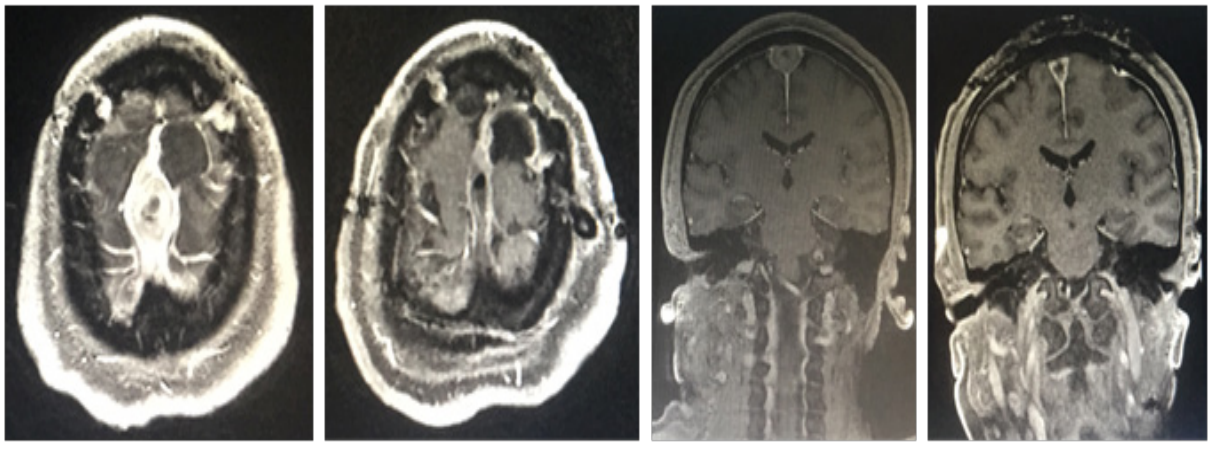


A B C



D E

Figure2: A-E, Intraoperative images showing the intrasinus metastases of the ENB resection



A B C D

Figure3: A-B PreOp and PosOp Contrast enhanced axial MRI comparison. C-D, PreOp and PosOp Contrast enhanced coronal MRI comparison.

49 patients with ENB were treated at the Mayo Clinic between 1951 and 1990. 5-year survival rate was 69%, tumor progression rate was 51% and metastasis was seen in 31%. The pathological grade was the most significant prognostic factor identified, 5-year survival rate was 80% for low grade tumors and 40% for high-grade tumors [10].

The ability to resect and accurately stage the disease by means of craniofacial resection, combined with radiotherapy and/or chemotherapy when appropriate, has resulted in significantly improved survival [9]. Several case reports have been published suggesting that endonasal endoscopic resection may be a viable alternative [8].

The combined treatment of ENB by craniofacial resection and radiotherapy has produced a substantial improvement in 5 and 10 years' survival, doubling the survival rates from an era before craniofacial surgery, providing the gold standard by which other approaches must be judge [1,9]. Chemotherapy should not be used as single-modality therapy for initial treatment but may provide additional benefit when used in combination with radiation and surgery, particularly in advanced-stage disease. Combined chemotherapy should be considered as initial therapy for unresectable tumors and metastatic disease, and as salvage therapy in disease recurrence, regimens with cisplatin and etoposide may be useful, but also non-platinum schemes (irinotecan, docetaxel, doxorubicin, ifosfamide, vincristine) were described to be effective [7,11]. Neoadjuvant chemotherapy has been advocated for locally invasive and advanced staged ENB, and has demonstrated the capacity to significantly decrease gross tumor volume before definitive surgery and/or radiation [1].

The use of an endoscope is a helpful adjuvant surgical tool, but given the difficulty in accurate diagnosis of tumor extent, surgeons must be prepared and able to proceed to craniofacial resection if the tumor extent is beyond that which can be encompassed endoscopically to obtain optimum results [9].

Radiosurgery is a recent innovation among treatment modalities, and endoscopic surgery represents a reasonable pre-radiosurgical alternative to conventional surgery. In selected cases provides excellent quality of life, less injury to the patient, and fewer side effects and long-term effect tan other treatment strategies [19]. Probable neither radiosurgery nor endoscopy alone would be sufficient for an effective treatment of ENB. However, the combination of both minimally invasive methods seems capable of yielding therapeutic results comparable with surgery followed by radiotherapy [8].

The most common site of metastases are the cervical lymph nodes (10-33%), while distant sites, including lungs, brain and bone, are less common [17]. Systemic metastases occurs by hematogenous and lymphatic spread, but distal central nervous system metastases from leptomeningeal dissemination is an extremely rare sequel, with a grave prognosis with an expected survival of less than 2 years [14]. The most accepted theory of distant dural invasion, is that tumor cells travel in the meningeal arteries or via retrograde flow using the valveless venous system and sinuses, resulting in tumor seeding [20]. The possibility of leptomeningeal dissemination should be kept in mind, when monitoring patients with ENB [12,13,21].

Tumor invasion to dural sinuses had been studied more widely in meningiomas. The anterior half of the SSS is narrower and has fewer associated venous lacunae, fewer pacchionian bodies, and smaller numbers of adjoining cortical veins entering the sinus than the posterior half, which facilitates surgical exploration. MRA provides accurate, noninvasive visualization of the arterial and venous anatomy, which facilitates analysis of sinus patency and invasion [22]. In tumors that completely obliterate the SSS, collateral anastomotic networks have usually developed over time and can prevent symptomatic venous hypertension. Resection of completely occluded sinuses should be considered a generally safe strategy [23,24].

Recurrence can occur years after the completion of treatment, even more than 10 years later. Therefore, long-term follow-up is needed, although the exact frequency of visits and the necessity for radiological studies have not been established [1,18]. Recurrent tumor and regional metastasis should be treated aggressivelybecause this approach has been shown to be worthwhile [10].

Conclusion

ENB is a rare and malignant tumor of the nasal cavity; his nonspecific symptoms entail in most of the cases a late diagnosis, with advanced stage of the disease. Metastasis has been shown to occur in many different locations, but intrasinus metastases are rare, with worst prognosis. Despite the advances of surgical techniques, microsurgical and endoscopical, and the use of adjuvant therapy with radiotherapy or chemotherapy, recurrences are common. This demands a long-term follow-up, with aggressive treatment of the recurrent tumor or regional metastases, to improve the quality of life of the patients. Further studies should be addressing the long-term outcomes of the treated recurrent disease.

Disclosure

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Conflict of Interests

Authors declared that they have no conflict of interest.

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