

Esthesioneuroblastoma Treated with Endoscopic Craniofacial Resection: A Report of a Case

Mitchell Ray Gore*

State University of New York Upstate Medical University, New York, USA

Abstract

Herein the author presents a case of esthesioneuroblastoma, a rare malignancy affecting the sinonasal cavity. This uncommon lesion is thought to originate from the olfactory neuroepithelium. The mass may invade the orbit and intracranial space, and may metastasize to the neck or to distant sites. This case was successfully treated with an endoscopic craniofacial resection, multilayer skull base reconstruction, and postoperative radiation. The differential diagnosis, radiographic findings, and surgical and adjuvant treatment of this rare tumor are discussed.

Keywords: Esthesioneuroblastoma; Craniofacial resection; Nasoseptal flap; Olfactory neuroblastoma

Introduction

Esthesioneuroblastoma (ENB) is an uncommon tumor occurring in the sinonasal region. It represents approximately 3% to 6% of primary malignant sinonasal tumors [1,2]. Esthesioneuroblastoma was first described in 1924 by Berger et al. [3] and likely arises from the cells of the olfactory epithelium. The first and most widely used staging system was described by Kadish et al. in 1976 [4] and was modified by Morita et al. in 1993 [5] to include stage A (disease limited to the nasal cavity), stage B (involving the paranasal sinuses) stage C (extension beyond the cribriform plate and paranasal sinuses into dura, brain, orbit, etc.), and stage D (involving cervical lymph nodes and/or distant metastases). Dulguerov et al. [6] proposed a TNM staging system that closely correlates with survival outcomes, and surgery (either endoscopic or open) combined with radiation therapy appears to offer the most favorable survival outcomes [7].

Case Report

An otherwise healthy 37-year-old female was referred to another Otolaryngologist for two months of left sided nasal obstruction and epistaxis. He performed endoscopic biopsy in the office and the pathology demonstrated nests of uniform small round blue cells with scant cytoplasm and round nuclei consistent with an esthesioneuroblastoma. She had an MRI (Figure 1) showing a left sided nasal mass with some extension into the left ethmoids that appeared to abut the left olfactory bulb but did not appear to invade the dura or anterior cranial fossa. A maxillofacial CT scan. Figure 2 demonstrated similar findings, with a left sided nasal/ethmoid mass but no orbital or intracranial invasion. A PET scan showed no cervical or distant metastasis, and she was staged as a Kadish B/T1N0M0. The patient was referred to the author for definitive surgical treatment. Two weeks after her initial office biopsy she underwent endoscopic craniofacial resection as typically described [8], with Draf III frontal sinusotomy, septectomy, removal of the middle



Figure 2: Coronal CT of the esthesioneuroblastoma.



Figure 3: Endoscopic view of the esthesioneuroblastoma.

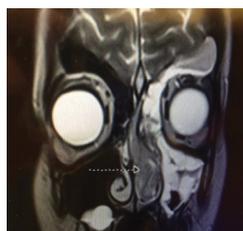


Figure 1: Coronal MRI of the esthesioneuroblastoma.

and inferior turbinates, and excision of the ethmoid roof, cribriform plate, and crista galli. The bilateral olfactory bulbs were excised and were negative for tumor on frozen section, and left and right dural margins from the anterior cranial fossa were also negative for tumor on frozen section. Figure 3 shows the left sided fleshy nasal mass in the left nasal cavity and olfactory cleft intraoperatively prior to excision. Figure 4 shows the Draf III frontal sinusotomy superiorly with the mass (inferior and posterior to the frontal sinuses) on the underside of the cribriform plate being reflected inferiorly off the anterior cranial fossa. Figure 5 shows the final defect with the Draf III frontal common cavity superiorly, sphenoid sinuses inferiorly, and the edges of the ethmoid roof and dural defect after negative margins were obtained. The skull base was reconstructed using an Alloderm (LifeCell Corporation/

*Corresponding author: Mitchell Ray Gore, State University of New York Upstate Medical University, New York, USA, Tel: +1 315-464-4570; E-mail: mgoremphd@gmail.com

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Kinetic Concepts, Inc., San Antonio, Texas) inlay graft on the dural side (Figure 6), and a right nasoseptal flap onlay [9] (Figure 7). The patient had intermittent clear rhinorrhea for approximately 1 week postoperatively that resolved with bedrest and sleeping with the head elevated. Six weeks after her initial negative-margin surgery and skull base reconstruction she underwent six weeks total of post-operative radiation (total 60 Gray) to the sinuses/anterior skull base and has shown no evidence of recurrence at endoscopic examination/debridement every 3 months for 3 years at the time of this article. Figure 8 shows her skull base reconstruction healed with no cerebrospinal fluid (CSF) leak at 6 months post-op. Figure 9 shows a hematoxylin and eosin (H&E) stain of the tumor showing a highly cellular tumor with scattered mitotic figures consistent with esthesioneuroblastoma. She never experienced any neck or distant spread.

Discussion

Esthesioneuroblastoma is a relatively uncommon tumor of the nasal

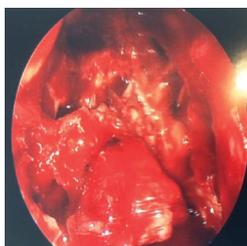


Figure 4: Intraoperative endoscopic view of the esthesioneuroblastoma during resection.



Figure 5: Final endoscopic craniofacial defect.

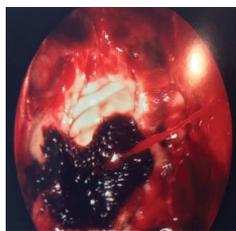


Figure 6: Alloderm inlay reconstruction of the skull base.



Figure 7: Nasoseptal flap reconstruction of the skull base.



Figure 8: Healed skull base reconstruction at six months after surgery.

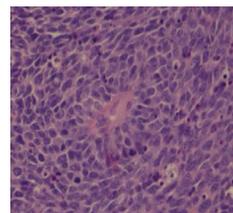


Figure 9: H&E stain of the esthesioneuroblastoma.

cavity and paranasal sinuses arising from olfactory neurosecretory cells near the cribriform plate and olfactory cleft. There is a bimodal age distribution with one peak in young adult patients (~2nd decade) and another peak in the 5th to 6th decades. Computed tomography (CT) typically shows a mass with soft tissue attenuation, and if contrasted the mass will typically show homogeneous enhancement. Focal calcifications may sometimes be seen. Bony destruction may be seen with larger tumors. On MRI esthesioneuroblastoma shows heterogeneous intermediate signal on T1 and T2 sequences, and shows moderate to intense enhancement with contrast. The differential includes meningioma, hemangiopericytoma, adenocarcinoma, sinonasal undifferentiated carcinoma, squamous cell carcinoma, sarcomas, metastatic masses such as melanoma (which may also be a primary sinonasal tumor) or renal cell carcinoma, lymphoma, nasopharyngeal carcinoma, chordoma, juvenile nasopharyngeal carcinoma (in pre-pubescent males), or large pituitary macroadenoma.

Several studies and case series have examined the recurrence rate and outcomes with various modalities. Surgical excision with negative margins is the mainstay of treatment, especially for smaller tumors, and complete resection can often be accomplished with endoscopic techniques, especially for smaller Kadish A and B tumors [10]. Endoscopic techniques may also decrease complication rates and may result in shorter post-operative recovery times. Larger tumors may also necessitate open craniotomy or open craniofacial approaches. Postoperative radiation is routinely administered and has been shown to decrease the risk of recurrence in conjunction with surgical resection [6]. Patients with negative margins and no evidence of cervical or distant metastases can achieve long-term survival with surgical resection and post-operative radiation [11]. Positive surgical margins, advanced Kadish or T stage, and cervical or distant metastases are correlated with decreased survival. Studies have shown that patients with cervical metastasis can benefit from salvage with neck dissection and post-operative neck irradiation [12]. In patients requiring skull base resection a multilayer repair of the skull base, preferably with a vascularized flap such as the nasoseptal flap, decreases the risk of post-operative CSF leak [13]. The literature has shown that patients with cervical adenopathy at the time of diagnosis, or with cervical metastases occurring after the presentation of the primary tumor, will benefit from treatment of the cervical metastases with a combination of surgery with neck dissection and post-operative neck irradiation. This combined

modality treatment of neck metastases demonstrated a statistically significant survival benefit over radiation alone or neck dissection alone [6,12,14]. Salvage of local recurrence has also been studied in patients with esthesioneuroblastoma, but a meta-analysis failed to demonstrate superiority of salvage surgery + radiation over surgery alone or radiation alone for local recurrence [15]. This disparity with the advantage shown with surgery + radiation for neck metastases may be due to the fact that many patients have already had adjuvant radiation to the primary site and may not be eligible for re-irradiation if they experience local recurrence. The higher survival rates and more favorable prognosis for esthesioneuroblastoma relative to more aggressive sinonasal lesions such as sinonasal melanoma or sinonasal undifferentiated carcinoma make it well-suited to the endoscopic craniofacial approach. A recent review [16] based on the Oxford Center for Evidence-based Medicine guidelines demonstrated that endoscopic approaches for esthesioneuroblastoma provide favorable survival outcomes, and that endoscopic approaches may actually provide better survival outcomes than traditional open approaches for esthesioneuroblastoma, especially for lower stage tumors. This patient had a favorable tumor confined to the nasal cavity and paranasal sinuses, and did well after endoscopic craniofacial resection with multilayer skull base repair and post-operative radiation, with no recurrence noted in the three years since her treatment.

Conclusion

We report a case of esthesioneuroblastoma involving the nasal cavity and ethmoid sinuses. Negative margins were obtained with an endoscopic resection, and the patient has remained tumor free after post-operative radiation. Although a rare tumor, esthesioneuroblastoma should be in the differential of any nasal mass. Prompt imaging and referral to an Otolaryngologist facilitates diagnosis and definitive treatment.

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