ORIGINAL ARTICLE

Giant Esthesioneuroblastoma: Is There Any Scope of Endoscopic Approach?

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Abstract

Esthesioneuroblastoma is a rare neoplasm of nose & paranasal sinuses and anterior skullbase. Management includes either radiotherapy or surgery or both. Surgical management can be done by external neurosurgical approach (i.e. craniofacial resection) or endoscopic approach. In the present case report we present our experience of the management of an extensive esthesioneuroblastoma kadish stage C treated by endoscopic excision and postoperative radiotherapy.

Keywords: Esthesioneuroblastoma, anterior skull base, kadish stage, cribriform plate.

Esthesioneuroblastoma is an uncommon malignant neoplasm of the anterior skull base. It accounts for approximately 6% of nasal cavity/paranasal sinus cancers.[1] This tumor typically presents with advanced disease at the time of diagnosis, which is attributed to the nonspecific symptoms associated with the disease. The most common symptoms at presentation include nasal obstruction, epistaxis, anosmia, and headache. The treatment of esthesioneuroblastoma is complicated by several factors: the rarity of the tumor, the lack of a universally accepted staging system, and a lack of consistency of treatment as a result of significant advances in this field over time. Treatment modalities include radiotherapy (RT) alone, surgery, combined RT and surgery and the use of chemotherapy for various stages of disease.[2] Endoscopic excision has been described for this tumor, but this is usually limited to early stage (Kadish stage A or B) tumors. Here, we describe a Kadish grade C tumor managed by means of endoscopic excision and postoperative radiotherapy.

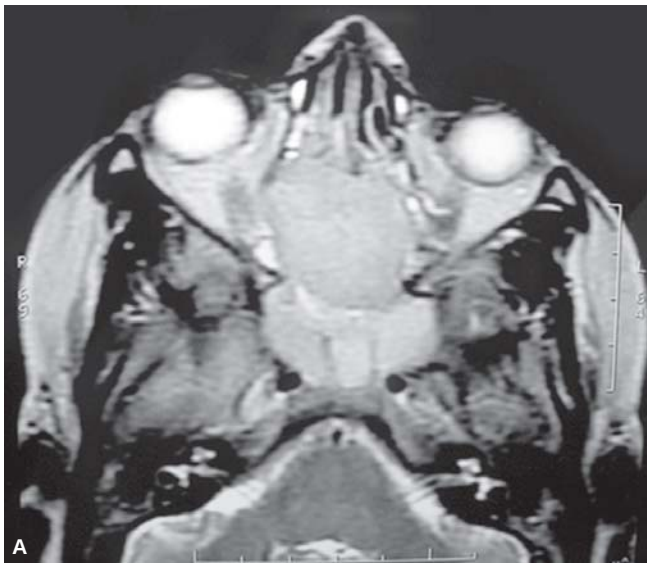
CASE REPORT

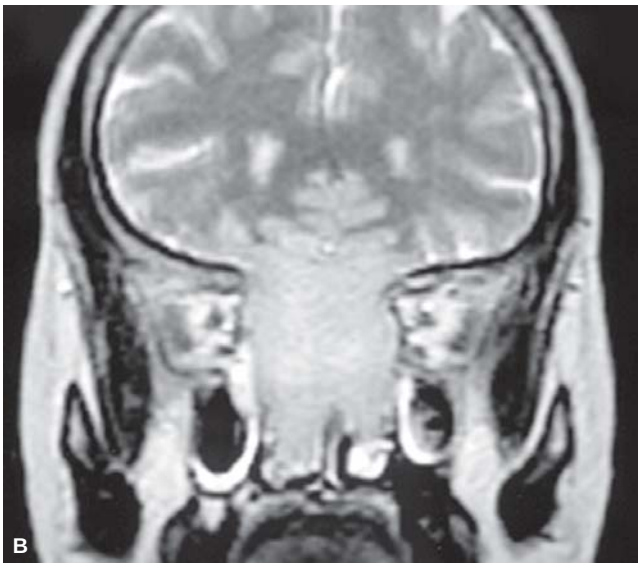
A 46 years old female patient presented to us with history of generalized headache and bilateral nasal obstruction since 1½ months. This was associated with blood stained nasal discharge and occasional episodes of frank epistaxis. She developed right eye vision loss for 25 days and left eye vision loss for 5 days. She also gave history of projectile vomiting. There was no history of loss of consciousness, seizures, proptosis or trauma. Anterior rhinoscopy and endoscopic examination revealed grayish mass with blood clots filling bilateral posterior nasal cavities and destruction of the posterior nasal septum. Visual acuity was negative for perception of light on both sides and pupils were mid-dilated (more dilated on right side). Extraocular movements were also restricted bilaterally. Fundus examination revealed disk pallor on both sides.

Magnetic Resonance Imaging of brain and paranasal sinuses revealed a nasoethmoid mass involving the sphenoid and eroding the anterior and middle skull base with intracranial extension (Figs 1A and B).

In view of short history of vision loss and fundus examination revealing disk pallor (and not atrophy) patient was immediately planned for endoscopic excision.

Middle turbinate was partially removed on right side and completely on left side to create space for proper exposure. Tumor was removed piecemeal with Blakseley forceps and microdebrider till skull base which was seen to be eroded from right fovea ethmoidalis to left fovea ethmoidalis including bilateral cribriform plates and partly the roof and





Figures 1A and B: Preoperative MRI photograph axial and coronal cuts showing enhancing soft tissue lesion in ethmoids and sphenoid with intracranial and intraorbital extension

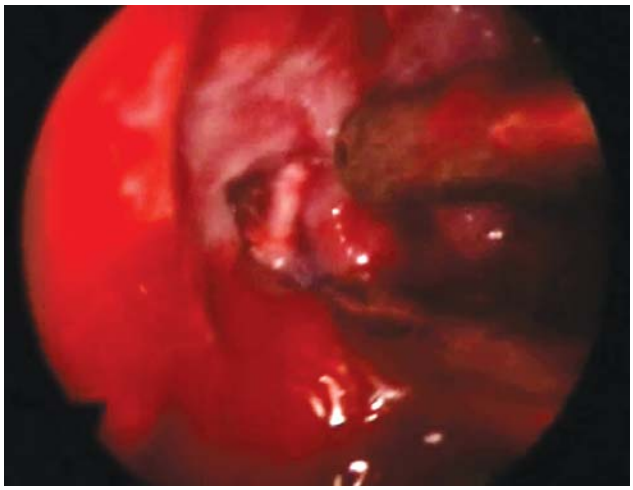


Figure 2: Intraoperative picture showing dehiscent optic nerve

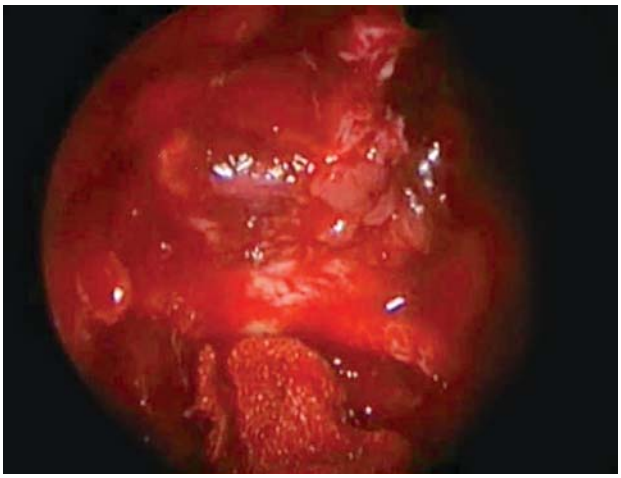
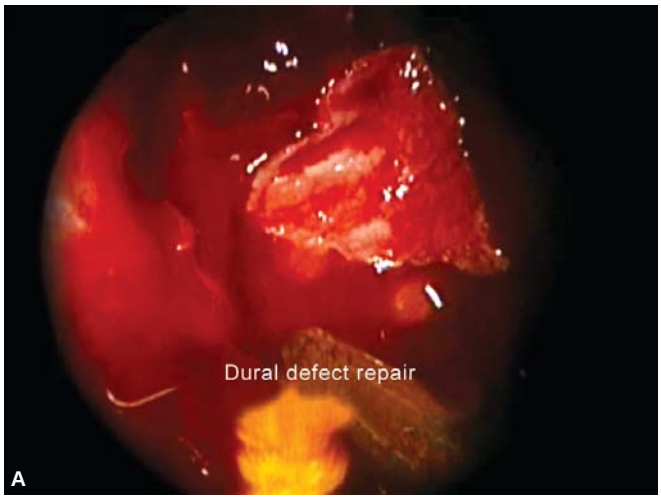


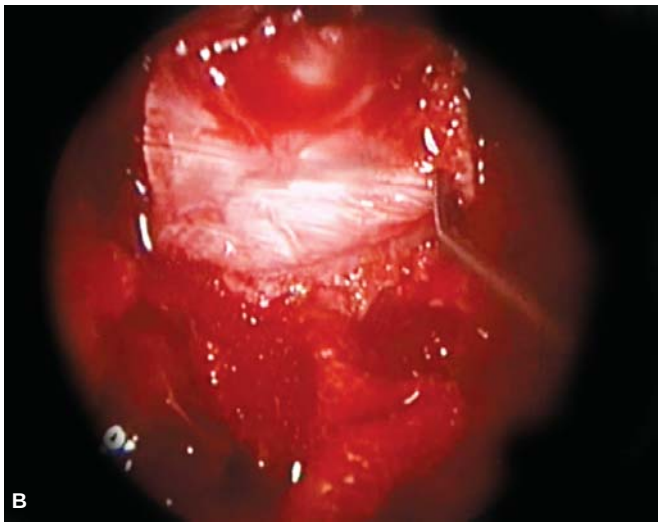
Figure 3: Intraoperative picture showing the exposed optic chiasma

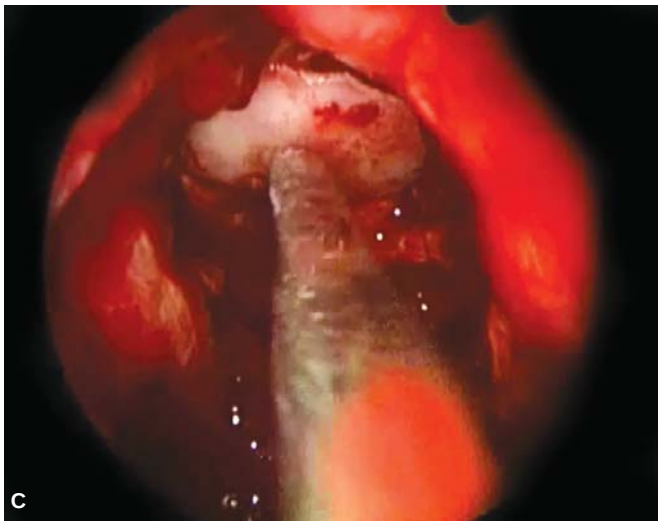
lateral walls of sphenoid sinus. Tumor was meticulously removed from these areas. Bilateral internal carotid arteries were identified. Bilateral orbital apex, annulus of Zinn and optic nerves were delineated up to the optic chiasma and optic nerve decompression was done (Figs 2 and 3). The tumor was seen to extend beyond the optic chiasma which was removed to visualize the optic tracts. Dural defect of approx. 2*2 cm was seen superior to optic chiasma with active CSF leak. Fascia lata and muscle from right anterolateral thigh were harvested. The dural defect was then repaired with the harvested fascia and muscle and preserved septal cartilage (Figs 4A to C). Region of skull base defect and optic chiasma were also supported with fascia. Bilateral anterior nasal packing was done. Postoperative period was uneventful. Patient was given radiotherapy after 3 weeks. The left eye vision improved to finger counting close to face at 8 months follow-up. The right eye vision however, remained PL negative. There was no locoregional recurrence (Fig. 5).

DISCUSSION

Esthesioneuroblastoma is believed to arise from the olfactory neuroepithelium located in the upper third of the nasal cavity including the nasal septum, cribriform plate, and the middle and superior turbinates.[3] The tumor presents as a smooth surfaced polyp with an intact respiratory mucosa. The







Figures 4A to C: Intraoperative pictures showing the repair of skull base defect in multiple layers using fascia lata and cartilage

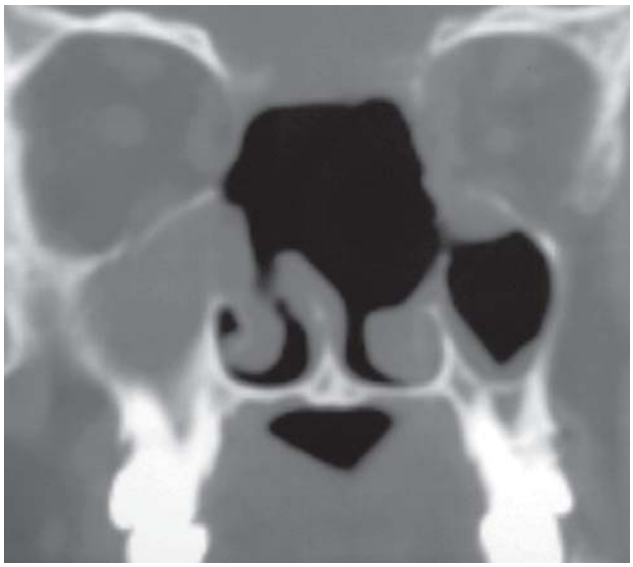


Figure 5: Postoperative CT photograph showing complete disease clearance

malignant cells are localized in nests in the submucosa and stroma. The red appearance is due to the rich vascularized stroma of this tumor.

Patients with esthesioneuroblastomas usually complain of unilateral nasal obstruction and epistaxis. Less commonly, patients experience anosmia, headache, and ocular disturbances. Diagnosis is frequently delayed, in some studies up to 6 months, because nasal obstruction, the most common symptom, is more often associated with less severe sinonasal problems.[4]

Esthesioneuroblastomas are graded histologically and staged clinically and both systems correlate with prognosis. In the Hyam's classification, tumors are graded 1 through 4 with those showing less differentiation receiving higher grades.[4] The Kadish system is based on the spread of the tumor.[3] Under this system, stage A tumors are confined to the nasal cavity, stage B tumors involve the sinuses, and stage C lesions invade the anterior cranial fossa or the orbit.

Treatment for esthesioneuroblastoma traditionally involves craniofacial resection (CFR), in the form of bifrontal craniotomy and lateral rhinotomy, followed by radiotherapy. However, this approach involves great degree of morbidity, facial disfigurement and mortality. Supporters of CFR maintain that, despite significant morbidity, traditional CFR allows for a true *en bloc* oncological resection. Although theoretically, piecemeal removal may increase the rate of local recurrence, this has not been demonstrated clinically.[5] Also, a true *en bloc* resection is rarely possible with traditional CFR due to the limited access to certain key anatomical areas, including the sphenoid sinus, frontal

recess, nasion, and orbital apex. Furthermore, piecemeal resection does not adversely affect survival.[5,6] It was because of this, that people started attempting endoscopic approach for this tumor. Initially it started as bifrontal craniotomy combined with endoscopic excision from below.[7-9] Authors (Unger et al, 2001) then reported combination of endoscopic surgery and transnasal stereotactic surgery.[10] However, this could never become popular because of technical difficulties.

Transnasal endoscopic resection has various advantages, besides lower morbidity and mortality. Also, lumbar drains and tracheostomies can be avoided. One of the important advantages is that there is no need for frontal lobe retraction. The operative time and hospital stay is also reduced. Endoscopic resection also allows preservation of important uninvolved structures.[11] Perhaps the most important goal is complete excision and avoidance of recurrence. With the advent of angled endoscopes which allow for better visualization of the field, complete tumor removal can be ensured and any CSF leak can also be repaired at the same time.[5]

Postoperative radiotherapy also plays an important role in this and the preferred treatment of esthesioneuroblastoma is combined modality of surgery and radiotherapy. However, radiotherapy has its own side-effects. It has been suggested that postoperative radiotherapy may be necessary only for tumors that demonstrate high-grade pathology, positive margins, large volume of disease, or extensive local invasion.[12] In fact, few authors have reported higher incidence of regional and distant failures after combined surgery and RT compared with surgery alone. Though these studies deal with cases operated by CFR, it can be extrapolated to endoscopic resection and in selected early stage cases, radiotherapy may be avoided and patients kept on close follow-up.[13]

Importantly, patients should be selected carefully for this approach with help of imaging. Informed written consent should be taken for an external approach, however, small the tumor might be. This may be required for oncological reasons. Though the case we operated upon was Kadish stage C, patients with smaller and early stage tumors can be better candidates for endoscopic resection.

Postoperative radiotherapy also plays an important role and combined modality treatment including both surgery and radiotherapy is the optimum and preferred management of esthesioneuroblastoma.

REFERENCES

1. Svane-Knudsen V, Jorgensen KE, Hansen O, et al. Cancer of the nasal cavity and paranasal sinuses: A series of 115 patients. Rhinology 1998;36 (1):12-14.
2. Simon JH, Zhen W, McCulloch TM, et al. Esthesioneuroblastoma: The University of Iowa experience 1978-1998. Laryngoscope 2001;111 (3):488-93.
3. Broich G, Pagliari A, Ottaviani F. Esthesioneuroblastoma: A general review of the cases published since the discovery of the tumour in 1924, Anticancer Res 1997;17:2683-706.
4. Dulgerov P, Allal AS, Calceterra TC. Esthesioneuroblastoma: A meta-analysis and review. Lancet Oncol 2001;2:683-90.
5. Batra PS, Citardi MJ, Worley S, et al. Resection of anterior skull base tumors: Comparison of combined traditional and endoscopic techniques. Am J Rhinol 2005;19:521-28.
6. McCutcheon IE, Blacklock JB, Weber RS, et al. Anterior transcranial (craniofacial) resection of tumors of the paranasal sinuses: Surgical technique and results. Neurosurg 1996;38: 471-79 Discussion 79-80.
7. Yuen AP, Fan YW, Fung CF, et al. Endoscopic-assisted cranionasal resection of olfactory neuroblastoma. Head-Neck 2005;27:488-93.
8. Yuen AP, Fung CF, Hung KN. Endoscopic cranionasal resection of anterior skull base tumor. Am J Otolaryngol 1997;18: 431-33.
9. Thaler ER, Kotapka M, Lanza DC, et al. Endoscopically assisted anterior cranial skull base resection of sinonasal tumors. Am J Rhinol 1999;13:303-10.
10. Unger F, Walch C, Stammberger H, et al. Olfactory neuroblastoma (esthesioneuroblastoma): Report of six cases treated by a novel combination of endoscopic surgery and radiosurgery. Minim Invasive Neurosurg 2001;44:79-84.
11. Har-El G, Casiano RR. Endoscopic management of anterior skull base tumors. Otolaryngol Clin North Am 2005;38:133-44.
12. Bentz BG, Bilsky MH, Shah JP, et al. Anterior skull base surgery for malignant tumors: A multivariate analysis of 27 years of experience. Head-Neck 2003;25:515-20.
13. Biller HF, Lawson W, Sachdev VP, et al. Esthesioneuroblastoma: Surgical treatment without radiation. Laryngoscope 1990;100 (11):1199-201.