Nasal Endoscopic and Anterior Craniotomy Resection for Esthesioneuroblastoma: How We Did It

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Abstract The objective of this study was to report and present an approach to combine functional and esthetic technique of endoscopic nasal tumor resection with anterior cranial fossa extension resection in a case of Esthesioneuroblastoma (ENB), a rare slow growing malignant neoplasm of the olfactory mucosa. For this purpose, the mass was excised by nasal endoscopic approach with microdebrider and anterior craniotomy with bi-coronal incision. The cribriform plate defect was reconstructed with temporalis fascia and nasal defect with fascia and rotated middle turbinate. Radiotherapy was given. At 4 years follow up no evidence of tumor locally or systemic metastasis were seen. In conclusion endoscopic excision complemented with anterior craniotomy for the anterior cranial fossa extension of resectable ENB with post-op radiotherapy is an effective treatment modality satisfying both oncosurgical as well as esthetic requirements.

Keywords Anterior craniotomy · Esthesioneuroblastoma · Endoscopic nasal surgery · Olfactory neuroblastoma

Introduction

Esthesioneuroblastoma (ENB) or olfactory neuroblastoma is a rare slow growing malignant neoplasm of neuroectodermal origin arising from neuroepithelial cells of olfactory

membrane of nose. It was first described by Berger et al. [1]. Less than 1,000 cases have been reported since 1924 [2]. There are various opinions regarding its origin, diagnosis and management protocols. It shows a varying biologic activity from an indolent growth to highly aggressive growth. Treatment plans range from a minimally invasive approach to craniofacial resection combined with radiotherapy [3]. In recent years development of skull base surgical teams have made combined resections of these tumors possible by allowing nasal, sinus, cribriform plate and anterior cranial fossa components to be excised concomitantly. However, they involve external facial incisions resulting in cosmetic defect. With development of endoscopic techniques it is possible to completely excise nasal and sinus tumors without requiring any external facial incisions [4]. We present a case of ENB which was successfully removed by combining the functional and esthetic technique of endoscopic power assisted nasal resection with cribriform plate and anterior cranial fossa extension resection using anterior craniotomy.

Case Report

A 52 year male farmer from Latur, a rural area of Maharashtra, India in March 2007 had presented with left sided nasal block, epiphora, headache, and epistaxis. On anterior rhinoscopy an irregular fleshy mass was seen arising from left nostril which bled on touch. Eye movements and vision were normal. There were no palpable neck nodes and rest of otorhinolaryngeal examination was normal.

High resolution CT scan (3 mm cuts) of paranasal sinuses was done to delineate extent of the mass. A minimally enhancing homogenous soft tissue mass of size 35 mm × 25 mm × 30 mm was seen in left nasal cavity,

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extending into the ethmoid sinus, and the cranial cavity with destruction of cribriform plate (Fig. 1). The orbital walls were normal. MRI was not available, hence not done.

Nasal endoscopy was done with biopsy under local anesthesia. On histopathological analysis, the tumor was identified as ENB. It showed homogenous small cells with uniform round to oval nuclei with rosette and pseudorosette formation and eosinophilic fibrillary intercellular background (Fig. 2). It was confirmed with immunohistochemistry.

The tumor was staged as Type C by Kadish classification [5], T2 as per Biller et al. [6], and T3 by Dulguerov's classification [5]. This staging helped plan the management in the form of surgical resection followed by radiotherapy.

The first author was the chief operating surgeon assisted by neurosurgeon who performed the craniotomy. The patient's head was prepared for anterior craniotomy. General anesthesia was given with patient in supine position and head elevated by 15°. Draping was done to allow movement of head by the otolaryngologist and the neurosurgeon adequately. Eyes were kept visible. A 0° nasal endoscope attached to camera and monitor was introduced. The septum was cauterized with suction diathermy 1 cm below the lower extent of tumor. A few pieces of tumor were taken with for histopathology. The tumor was then excised using the microdebrider under endoscopic guidance from the nose and paranasal sinuses. The sphenoid sinus was opened. The tumor was resected up to its origin at the cribriform plate and bleeding controlled with suction and bipolar diathermy. The orbital walls were preserved. After the intranasal resection was done, the neurosurgeon performed the anterior craniotomy. A bi-coronal incision was made taking care to preserve the vascular pericranial

Fig. 1 HRCT (coronal) of paranasal sinuses showing left sided mass extending into the ethmoid sinus and the cranial cavity with destruction of cribriform plate

Fig. 2 Histopathology: homogenous small cells with uniform round to oval nuclei with rosette and pseudorosette formation and eosinophilic fibrillary intercellular background (H and E $\times 100$)

fascia flap. Craniotomy was performed and the frontal bone removed. The frontal lobe was retracted extradurally, the cribriform plate was identified and intracranial extent of tumor noted. The ENT surgeon simultaneously placed the endoscope in the nose; the light from endoscope inferiorly helped the neurosurgeon working from above to delineate the lateral margins at the roof of ethmoid sinus. The cribriform plate and anterior cranial fossa extension of the tumor were removed completely using bone chisels.

The cribriform plate defect reconstruction was done with temporalis fascia, bony defect of cranium closed and osteoplastic flap repositioned and sutured. The nasal defect was reconstructed with temporalis fascia and rotated middle turbinate. Intranasal pack was placed.

Patient was kept in surgical ICU for 2 days. CSF was drained by lumbar puncture to decrease intracranial pressure. Intravenous antibiotics and mannitol were administered. Post-operative stay was uneventful. Histopathologic examination confirmed the diagnosis of ENB. After 3 weeks patient was given 6000 cGY radiotherapy in 30 fractions.

Examination after 1 month showed good healing and no CSF leak or tumor. Repeated endoscopy and CT scan done at 3 years follow up showed no evidence of any tumor (Figs. 3 and 4). There is no evidence of any locoregional recurrence or systemic metastasis even after 4 years of follow up in January 2011. He has been advised regular follow up.

Discussion

Esthesioneuroblastoma constitutes 3% of all intranasal neoplasms in all ages, peak being in the 2nd and 6th decade with equal distribution in both sexes [5]. The common symptoms are nasal obstruction, recurrent epistaxis, anosmia, and headache. In extensive cases, orbital complaints

Fig. 3 Nasal endoscopy at 3 years follow up showing no evidence of tumor

Fig. 4 HRCT (coronal) of paranasal sinuses at 3 years follow up showing no evidence of tumor recurrence

like proptosis, diplopia, and epiphora may be present [2]. It is highly vascular; hence the reddish pink intranasal mass bleeds easily. ENB presents as a homogenous soft tissue mass on CT scan showing moderate enhancement. MRI helps in delineating sinonasal, intraorbital or an intracranial extension. Nasal endoscopy and biopsy help in diagnosis.

Typical histopathological findings of ENB are homogenous small cells with uniform round to oval nuclei with rosette (Flexner–Wintersteiner) or pseudorosette (Homer–Wright) formation and eosinophilic fibrillary intercellular background [2]. Immunohistochemistry gives a definitive diagnosis it being positive for neuroendocrine markers like S-100 protein, chromograinin and synaptophysin.

Various staging systems have been proposed over years and have been improvised to assist in guiding treatment and prognosis [2].

Literature considers surgical resection combined with post-operative radiotherapy as the gold standard in management of ENB. Adjuvant chemotherapy has been also used. Craniofacial resection with post op radiotherapy provides better 5 years disease free survival rate (86%) compared to other treatment modalities [5]. These

resections however not only involve anterior craniotomy but also external facial incisions allowing good surgical exposure but poor cosmetic defects.

The development of endoscopic techniques has provided us technology to excise nasal and sinus tumors without external incisions. Devaiah et al. [4] in their retrospective study of seven patients of ENB who were endoscopically operated along with anterior craniotomy, concluded that this treatment option was effective. They found that visualization with this technique was exceptional. We had similar experience. The resection of cribriform plate even in small tumors is best done safely and completely by anterior craniotomy, rather than endoscopic technique alone. Hence, a combined technique involving an experienced endoscopic surgeon and a neurosurgeon is more preferable. The use of suction microdebriders further enhances the visualization providing good operative field and shortening the operative time. They allow rapid excision of main bulk of tumors so that residual tumor can be removed from recesses found throughout the anterior skull base [7]. We used this technique in our case.

The reconstruction of dural and nasal defects presents a challenge. The dural defect can be closed by fascia lata. The pericranial flap over the anterior craniotomy is sutured to the dura to gain a watertight seal [4]. The nasal defect can be closed by skin graft with fat or nasal septal mucoperichondrium [4]. In our patient the cribriform plate defect was reconstructed with temporalis fascia and nasal defect with fascia and rotated middle turbinate.

Conclusions/Key Message

Endoscopic excision complimented with anterior craniotomy for the anterior cranial fossa extension of resectable ENB with post-op radiotherapy is an effective treatment modality satisfying both oncosurgical as well as esthetic requirements. Advantages are: (1) avoidance of deforming facial incisions, (2) enhanced tumor visualization with endoscope and precise microscopic excision by microdebrider, (3) complete removal of extensive lesions with closure of anterior cranial defects, (4) allows wide drainage of sinuses near resections, (5) early healing allowing early management by radiotherapy.

Summary

- Case of ENB, Type C by Kadish is reported.
- Combined endoscopic resection with anterior craniotomy is described.
- Advantages of the surgical procedure are elaborated with review of literature.

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