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Hyperostotic Esthesioneuroblastoma

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Abstract

Esthesioneuroblastomas are rare, soft-tissue tumors that can often extend from the sinonasal cavity into the intracranial and orbital space. Prognosis depends upon the histological grade and location/extent of the tumor. Treatment often consists of maximum surgical resection followed by adjuvant chemoradiation therapy.

We present a case of a patient with esthesioneuroblastoma accompanied by an extensive osteoblastic reaction leading to significant hyperostosis along the skull base. His presenting symptoms included diplopia, and imaging revealed invasion of the orbital and intracranial spaces. Although a gross total resection of the soft tissue component of the tumor was achieved, a complete removal of the involved hyperostotic skull base could not be performed despite endoscopic endonasal and bifrontal craniotomy approaches in the same operative setting. Symptomatically, the patient improved and went on to receive chemoradiation therapy; he remains clinically and radiographically stable at 12 months. Investigation into the genetics and immunohistochemistry of this rare, hyperostotic variant of esthesioneuroblastoma may provide details regarding its aggressive nature.

Keywords

Esthesioneuroblastoma; hyperostosis

Introduction

Esthesioneuroblastoma is an uncommon malignant neoplasm that arises from olfactory neuroepithelium and is largely considered a soft tissue tumor. Despite a sinonasal origin, these tumors have the potential for bony erosion and can extend through the anterior fossa

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Financial Disclosure/Conflict of Interest Statement

Drs. Ramanathan and Germanwala have no financial disclosure or conflict of interest pertaining to any of the drugs, materials, and/or devices discussed in this report.

Authorship Justification

Dr. Ramanathan and Dr. Germanwala were surgically and clinically involved in the described case and assisted in writing all portions of the manuscript.

floor and invade the intracranial cavity. In very rare instances, this soft tissue neoplasm can produce an osteoblastic change in these areas of bony erosion. Although these hyperostotic changes are classically seen with anterior skull base meningiomas, we report a case of esthesioneuroblastoma with extensive hyperostosis of the skull base, sphenoethmoid region, middle turbinate, and orbit.

Case Presentation

A 42 year-old man presented with a one-month history of sinonasal fullness and a one-week history of diplopia. Physical examination revealed a complete left abducens palsy and diagnostic nasal endoscopy revealed a large mass medial to the left middle turbinate extending to the sphenoethmoid recess (Fig. 1a). Imaging by CT and MRI revealed a large left sided skull base mass centered in the left ethmoid and cribriform plate. The mass eroded the left lamina papyracea and anterior fossa floor and extended into the orbit and intracranial cavity (Fig. 2a). Extensive osteoblastic change was noted along the remaining surrounding bone of the skull base, orbit, and sphenoethmoid regions (Fig. 2b). An endoscopic endonasal biopsy of the lesion was performed and revealed a Hyams grade 3 (Kadish stage C) esthesioneuroblastoma. Given the large intracranial component with extensive surrounding osteoblastosis, the patient underwent a two-step procedure. A bifrontal craniotomy was performed to remove the osteoblastic skull base and intracranial/sinonasal tumor. In the same operative setting, an endoscopic endonasal approach was performed in conjunction with neuro-ophthalmology to remove the osteoblastic lamina papyracea, decompress the orbit, and explore for additional tumor. A gross total resection of the soft tissue tumor was performed at the conclusion of the procedures. Margins were obtained from the left periorbita, nasal septum, and subfrontal dura, which were all negative. The large skull base defect was reconstructed using a vascularized pericranial flap in addition to a vascularized pedicled right-sided nasoseptal flap (Fig. 1b). Postoperatively, the patient had complete resolution of his diplopia and abducens palsy after several days. As the extensive hyperostotic skull base could not entirely be resected, the patient underwent chemotherapy followed by radiation therapy to the nasal cavity, paranasal sinuses, and anterior skull base. In follow up, the patient remains symptomatically free and repeat imaging at 12 months continues to reveal no evidence of residual tumor (Fig. 2c). The patient did not develop any signs of CSF leak or meningitis.

Discussion

Esthesioneuroblastoma is a malignant, slow growing, soft neurogenic tumor that originates in the olfactory mucosa of the upper nasal cavity. Esthesioneuroblastoma associated with bony hyperostosis is extremely rare. A survey of the literature demonstrates only one prior report of two cases that showed much less hyperostosis than this case without an intracranial component[1]. Immunohistochemistry and genetic studies may provide information that explains the more aggressive behavior of this variant.

In general, hyperostosis in the setting of a skull base mass is highly suggestive of meningioma[2]. These regions are often due to actual invasion of the bone with tumor, and aggressive surgical removal, when possible, is recommended[3]. Management of such

lesions where the osteoblastic skull base may not be resectable from a purely endoscopic endonasal resection should include consideration for a bifrontal craniotomy to achieve higher rates of bony removal; although a gross total resection of the bony component could not be achieved in this case, maximum bony resection was performed without neurovascular injury through this approach.

Another interesting aspect of this case is the large skull base defect that was created as a result of removing portions of the hyperostotic skull base. Previous studies have demonstrated the limitations of using the pericranial flap alone to reconstruct large skull base defects[4]. This case utilized a double flap technique, combining both vascularized pedicled pericranial (anterior) and nasoseptal (posterior) flaps to close the large defect[5]. Given the likely need for postoperative radiation, skull base reconstruction with generous, robust tissue was a priority.

Conclusion

As demonstrated in this case, the presence of an osteoblastic reaction in the setting of a soft tissue mass in the nasal and intracranial cavities is rare; however, esthesioneuroblastoma should be considered in the differential diagnosis. A tissue proven biopsy as an initial step may help guide the best overall management for these patients. Given the rarity of these cases, overall prognosis of the hyperostotic variant of esthesioneuroblastoma remains to be determined; however, given the extensive surrounding bony changes, it is hypothesized that these tumors are more aggressive. Treatment will likely include a multidisciplinary approach towards surgical resection and postoperative chemotherapy and radiation with close follow up. Investigation into the genetics of these rare tumors may provide additional details regarding their pathophysiology.

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Figure 1.

1a: Diagnostic left nasal endoscopy revealed a large mass medial to the middle turbinate extending to the sphenoid recess.

1b: Intraoperative endoscopic image depicting skull base reconstruction following tumor resection. LP= Lamina Papyracea, NSF= Nasoseptal Flap, PCF= Pericranial Flap.

Figure 2.

2a: Preoperative coronal MRI T1 with contrast revealing left sinonasal mass extending into the orbit and intracranial space.

2b: Preoperative sagittal CT demonstrating extensive hyperostosis of the skull base.

2c: Postoperative coronal MRI T1 with contrast demonstrating gross total resection of the tumor.