ABSTRACT

Esthesioneuroblastoma is an uncommon malignant neuroectodermal tumor arising from olfactory epithelium that was first described in 1924. There are only about 100 reported cases in the pediatric population and patients usually present with nasal obstruction, pain, hyposmia and epistaxis [1]. Surgical treatment for anterior skull base malignancies, such as esthesioneuroblastomas, has traditionally been performed by craniofacial resection with a transfacial transantral approach. Typically, a head and neck surgeon resects the intranasal tumor via a lateral rhinotomy, while the neurosurgeon performs a frontal craniotomy. In children, this approach has a reported postoperative complication risk of 25%, which includes CSF leaks, wound infections, and death [2]. Furthermore, the open surgery can lead to facial asymmetry, especially in children, by disrupting craniofacial growth centers. Other side effects from the craniotomy and resection of the frontal bone include seizures, mental status changes, frontal lobe abscesses, meningitis, hemorrhage, and pnuemocephalus [3].

With the improvement in endoscopic surgical techniques and instruments designed for endonasal skull base surgery, endoscopic approaches have been showing significant promise as new treatment modality for select sinonasal and skull base tumors [4]. Due to the rarity of anterior skull base malignancies in children, there is a need to report on outcomes of this less invasive and promising modality in pediatric pts.

INTRODUCTION

Esthesioneuroblastoma is a rare tumor with only about 100 reported cases. Standard surgical treatment is the open craniofacial resection. Pre-treatment imaging is critical for planning, and surgery remains the primary treatment modality followed by adjuvant therapy. We report a 12-year-old female with an endoscopic assisted craniofacial resection of an extensively calcified Kadish C and Dulguerov-Calceterra T3N0M0 esthesioneuroblastoma. Following radiation therapy, she is disease-free 18 months post-treatment. Although there are few reported cases of pediatric pts., our case supports the use of endonasal endoscopic approaches for select pediatric skull base malignancies.

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DISCUSSION

Although open craniofacial resection has been successful, this approach has potential for significant morbidity. In a recent review including 84 pediatric patients, from 17 institutions, undergoing craniofacial resections for anterior skull base malignancies from 1950 to 2000, the postoperative complication rate was 25% (range 10-50%). Postoperative complications included wound complications (13%), postoperative death (4.8%), orbital complications (3.6%), CNS complications including meningitis or cerebrospinal fluid leak (2.4%), and systemic complications (1.2%) [3]. We cannot compare the complication rate to the endoscopic approach in pediatric cases because there are very few reported cases. In adults, the largest series of endoscopic resected skull base tumors included 62 patients, and the incidence of similar complications was 15% [4].

CONCLUSIONS

In the recognition of the invasiveness, recent reports demonstrate the effectiveness of endoscopic approaches in the oncologic outcomes when compared to craniofacial approaches for a wide variety of skull base malignancies, such as esthesioneuroblastoma, but these series do not include the pediatric population [7]. A recent meta-analysis actually showed a significantly better survival curve in patients with esthesioneuroblastomas treated with an endonasal approach compared to craniofacial approach to a open approach [6]. However, this benefit is slightly misleading since the open surgical approach cohort had more patients with higher tumor stage. Nevertheless, a recent systematic review revealed similar disease and survival outcomes among adult patients having either endoscopic or open craniofacial resection for various sinonasal malignancies [7].

REFERENCES


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Case Presentation

A 12-year-old female presented with a two-year history of increasing right nasal airway obstruction, of recurrent right-sided epistaxis and a four-month history of increasing right nasal airway obstruction, pain, hyposmia and epistaxis.

CT scan and MRI revealed a large right-sided nasal tumor (Figure 1). There was erosion of the lamina papyracea with subsequent mass effect on the orbital contents. There was also tumor spread through the cribiform plate, but without dural enhancement. Due to the extensive calcifications seen on CT, the initial diagnosis was suggestive of a fibrousosus lesion such as a fibrosarcoma.

Figure 1: (A) Coronal and (B) axial CT scans revealing coarsely calcified esthesioneuroblastoma with orbital invasion and (C) MRI showing no intranasal spread.

Surgical Treatment

Our patient underwent endonasal-assisted craniofacial resection (CFR) followed by radiation therapy. No transfacial or biconcave incisions were used. The intranasal tumor was resected completely via an endoscopic craniofacial CFR, which included resection of the ipsilateral anterior skull base. The tumour was first approached using a transperiorbital approach. The tumor was then excised via an endoscopic transnasal transcaruncular approach. The intracranial extension of the tumor was excised via a transsphenoidal approach, and the tumor in the orbit, including the orbital periosteum, was resected completely via an endonasal endoscopic endoscopic transsphenoidal approach with a total of 54 Gy and is disease-free 18 months post-treatment.

Figure 2: (A) Well-formed lobules of closely packed neoplastic cells separated by highly vascularized stroma. (B) Tumor cells with fairly uniform round to ovoid nuclei set in a finely fibrillar stroma.

Figure 3: MRI T2 coronal image 13 months post-treatment showing no evidence of recurrence.

DISCUSSION cont.

Our case of a successful endonasal assisted craniofacial resection of a Kadish C and Dulguerov-Calcaterra T3N0M0 esthesioneuroblastoma highlights the use of this approach for select pediatric anterior skull base malignancies. More experience is required to identify the ideal patients for this minimally invasive approach, and to determine long-term treatment outcomes.

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Our case of a successful endonasal assisted craniofacial resection of a Kadish C and Dulguerov-Calcaterra T3N0M0 esthesioneuroblastoma highlights the use of this approach for select pediatric anterior skull base malignancies. More experience is required to identify the ideal patients for this minimally invasive approach, and to determine long-term treatment outcomes.

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