The Pediatric Anterior Skull Base: An Otolaryngologist’s Perspective

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ABSTRACT
Anterior skull base tumors have traditionally posed a therapeutic challenge. However, the advancement of skull base and endoscopic surgery has allowed for more of these lesions to be amenable to surgical resection. Though common in the adult population, surgical approaches in the pediatric population is not widely described. This chapter discusses the presentation and treatment for various pediatric anterior skull base lesions. Surgical approaches, complications, and the role of the otolaryngologist is also discussed.

Keywords: Anterior skull base tumors; Children; Pediatric skull base surgery.

INTRODUCTION
Skull base tumors have traditionally posed a therapeutic challenge to the otolaryngologist and neurosurgeon. Prior to the advent of skull base surgery, malignant tumors involving the skull base were considered inoperable and almost universally fatal. However, with the advancement of skull base surgery and endoscopic techniques, previously unresectable lesions can now be managed surgically, and the role of the otolaryngologist is increasing, especially for those lesions involving the anterior cranial fossa. Diagnosis and management of skull base tumors are well described in adults, but only a few reports exist pertaining to the pediatric population, primarily because skull base tumors are generally uncommon but even rarer in children. In adults, meningiomas and sinonasal malignancies are the most common anterior skull base lesions, whereas fibrous dysplasia, esthesioneuroblastomas, and encephaloceles predominate in children. In addition, juvenile nasopharyngeal angiofibromas (JNAs) originate from the sinonasal tract but can extend to involve the skull base. Children pose a therapeutic challenge because of their developing skull, face, and sinuses and the desire to avoid any intervention that may lead to developmental complications. This article will focus on the presentation and treatment of pediatric anterior skull base lesions. Common surgical approaches, complications, outcomes, and, specifically, the role of the otolaryngologist in the treatment team will be addressed.

ANATOMY
The anterior cranial fossa is composed of the orbital portion of the frontal bone anteriorly, cribriform plate of the ethmoid bone centrally, and the lesser wing and body of the sphenoid bone posteriorly. It is demarcated from the middle cranial fossa by the anterior clinoid processes and the lesser sphenoid wings, which mark its posterior extent (Fig. 1). Anteriorly, the cribriform plate transmits the olfactory nerves from the superior nasal mucosa, and posteriorly, it articulates with the sphenoid body, which is the site, from anterior to posterior, of the planum sphenoidale, limbus sphenoidale, chiasmatic sulcus, tuberculum sellae, pituitary fossa, and dorsum sellae. The planum sphenoidale forms the roof of the posterior ethmoid sinus and the anterior part of the sphenoid sinus and borders on the optic canals posterolaterally.

When accessing the anterior skull base “from below,” it is important to note that the floor of the anterior cranial fossa is uneven. The orbital roofs slope downward medially to join the ethmoid sinuses, and this downward slope becomes more exaggerated heading to the cribriform area (Fig. 2). Thus, the midline is usually the lowest point of the skull base, and an axial plane of dissection, i.e., safe at the area of the ethmoid sinuses may risk injury to the brain if extended medially at the same level.

HISTORY AND PHYSICAL EXAM
Clinical findings in pediatric patients with anterior skull base lesions are determined by the location and extent of the tumor and do not differ significantly from those seen
in the adult population. In a retrospective chart review of children undergoing resection of skull base lesions, Hanbali et al found that the most common complaints were visual, nasal, and facial deformity. Blindness or diplopia may predict orbital involvement, while anosmia can be seen in advanced tumors involving the cribriform plate. Other clinical manifestations noted in the literature include recurrent sinus infection, headache, and heaviness of the head.

Physical exam includes a thorough cranial nerve exam as well as an endoscopic exam. Hanbali et al found that the most common clinical findings in children with skull base lesions requiring resection included decreased facial sensation, restricted ocular motility, decreased visual acuity, and proptosis. Examination with a 2.7 mm flexible scope or a rigid telescope—0°, 30°, and 70°—is feasible with appropriate topical anesthesia and parental cooperation with nurse assistance. If an adequate exam cannot be performed, general anesthesia may be necessary for nasal endoscopy.

Imaging via computed tomography (CT) and magnetic resonance imaging (MRI) is essential and should be performed with contrast in axial, coronal, and sagittal planes. Fine-cut CT (submillimeter slices) provides critical anatomic information important for surgery, including the presence and extent of erosion of the skull base or orbital wall, position of vessels and nerves, and presence of intersinus septa. Magnetic resonance imaging can help differentiate between neoplastic or inflammatory tissue and assists in confirming certain diagnoses, such as meningoceles, nasal gliomas, and retained secretions. Computed tomography or MR angiography may be considered for lesions extending to the middle cranial fossa involving the internal carotid artery (ICA), verteobasilar system, or cavernous sinus. Conventional angiography may be considered in certain situations: evaluation of the circle of Willis, extent of tumor involvement on the ICA, or distinguishing tumor from aneurysm.

DIFFERENTIAL DIAGNOSIS

Pediatric skull base tumors are rare, usually benign, and have a male predominance. Although tumor types vary tremendously, most tumors are of mesenchymal origin. Whereas the most common anterior skull base tumors in adults are nasal or sinonasal malignancies and meningiomas, these are rare in children, who are more likely to have encephaloceles, fibrous dysplasia, esthesioneuroblastomas, and nerve sheath tumors.

Encephaloceles are extensions of intracranial structures outside the skull and have an incidence of about 0.2 per 1,000 live births and fetal deaths. Interestingly, in North America, these usually occur occipitally, but in South America, they are usually found in the anterior cranial fossa.

Fibrous dysplasia is an anomaly of the precursors of bone, in which the transformation of woven bone to lamellar bone does not occur, leading to an overgrowth of well-vascularized fibrous stroma surrounding distorted osseous trabeculae. When it involves the sphenoid wing, it
can encroach upon the optic nerve, leading to progressive visual loss.\(^2\) Although there is a 25% risk of recurrence with partial removal of the lesion, radiation is not recommended in cases of incomplete resection as there is risk of malignant transformation.\(^3\) Unfortunately, there are also reports of spontaneous malignant transformation in the absence of radiation exposure.\(^4\)

Esthesioneuroblastomas, or olfactory neuroblastomas, are tumors of neuroectodermal origin believed to arise from the mitotically active basal layer of the olfactory epithelium.\(^5\) Surgery alone is advocated in cases of low-grade tumors if complete resection with margins can be achieved, with the addition of radiation for close margins or residual or recurrent disease. Chemotherapy is recommended for high-grade tumors.\(^2\)

Though rare, sinonasal pathology, such as a JNA, can extend to involve the skull base. These benign, vascular, locally aggressive tumors are predominantly found in adolescent males and are thought to originate from a persistent vascular plexus after involution of the first branchial arch\(^6\) at the sphenopalatine foramen. It can grow toward the pterygomaxillary fossa, infratemporal fossa, and the inferior orbital fissure with extension into the anterior and middle cranial fossae (Fig. 3). There are many classification systems including Fisch, Chandler, Andrews, and most recently Radkowski, which help define the tumor based on extent and skull base erosion. The tumors are preoperatively embolized, and surgical resection may involve an open or, more commonly, an endoscopic technique. The presence of sinonasal pathology with potential skull base extension highlights the importance that otolaryngologists, especially the rhinologist or pediatric otolaryngologist who manages these patients, have a strong understanding of skull base pathology and anatomy.

Other anterior skull base lesions described in children include ossifying fibromas, sarcomas, pleomorphic neurofibromas, adenocarcinomas, adenoid cystic carcinomas, and neuroblastomas.\(^1,3\) When malignant, sarcomas are the most common skull base malignancy in children, sixfold more common than nonsarcoma malignancies, and predominate at the anterior, compared with the middle or posterior, skull base in a two-to-one ratio.\(^5\) Some sarcomas, including osteosarcoma, malignant fibrous histiocytoma, fibrosarcoma, and spindle cell sarcoma, have been reported in children who had previously undergone radiation, most commonly for retinoblastoma.\(^1\)

**MANAGEMENT**

The management of anterior skull base tumors in children is similar to that in adults and depends on the tumor type and location, surgical resectability, and the availability and effectiveness of adjuvant therapies. However, management strategies must take into account the patient's age and developmental stage, as many therapies are known to have detrimental effects on growth and function.\(^4\)

**Medical**

Chemotherapy has not been useful for solid tumors of the skull base in children, other than for rhabdomyosarcoma.\(^5\)

Crani al radiotherapy, be it external beam or brachytherapy, is frequently used as adjuvant therapy in adults with solid tumors of the skull base. However, it has a more limited role in children because of concerns for generalized growth retardation, pituitary insufficiency and its associated hormonal disturbances, and irreversible visual consequences due to proximity of the orbit.\(^4\) In regard to stereotactic radiosurgery, long-term effects in children have not been adequately assessed, and its role in the treatment of benign skull base tumors in children is unclear.\(^7\) Thus, for pediatric patients harboring benign tumors or low-grade malignant lesions, achieving complete resection is more critical than it is in adults, because supplementation with adjunctive radiotherapy is not desirable. In fact, even subtotally resected tumors are generally observed, with radiation being reserved only for progressive, multifocal, or unresectable disease.\(^5\)

**Surgical**

Prior to proceeding with surgery, the operability of the lesion must first be assessed. Invasion of the brain stem, internal carotid arteries, cavernous sinuses, spinal cord, or any portion of the brain that, if removed, will give a poor quality of life are all contraindications to surgery.\(^8,10\) Invasion of the optic chiasm is a relative contraindication,
because blindness is not an acceptable outcome of surgery. Additionally, evidence of distant metastases or poor physical state of the child would also serve as contraindications.

There are various craniofacial approaches to skull base tumors, and the decision primarily depends on the extent of the lesion. In young children, the anterior cranial fossa is relatively shallow and the frontal sinuses are not well developed, making access to this area easier than in adults. The most commonly used approaches combine frontal craniotomy with some form of transfacial exposure, including transoral or transpalatal approach, lateral rhinotomy, Le Fort I osteotomy, and midfacial degloving. Although these approaches are commonly utilized in adults, they must frequently be modified in children to avoid complications specific to this population.

In 1910, an endonasal transseptal, transsphenoidal approach was first described to remove pituitary tumors. The development of endoscopic technique and image-guided computer-assisted surgery has revolutionized the surgical management of skull base tumors, and endonasal surgery has become the procedure of choice, primarily for midline lesions. With improved technology, angled scopes, image guidance systems, and high-speed drills, it is now possible to access the midline skull base from the frontal sinus to the second cervical vertebra and from the sella to the jugular foramen endoscopically (Fig. 4). The endoscope can also be used as an adjunct with open approaches to eliminate the need for certain facial incisions as well as to look in areas hidden from the field of view of a microscope. The endoscopic approach should not be used if there is involvement of the orbit, far lateral extent of the maxillary sinus, involvement of dura lateral to the maximum convexity of the orbit, internal carotid or cavernous sinus extension, or any instance where it will not be possible to remove all tumor safely with a margin of healthy tissue.

There are various endoscopic endonasal approaches that can be used, and the decision depends on the location, nature, and malignancy of the lesion or tumor. These approaches can be classified as transnasal, transethmoid, and transseptal. Extended transnasal approaches include transplanum, transphenoidal, transpterygoid, transclival, and transpharyngeal. A combination of these approaches may be needed if the lesion extends posteriorly to the middle cranial fossa. The transethmoid approach is appropriate for lesions involving the ethmoid sinus, medial orbital wall, and sphenoid sinus. The transseptal and transnasal approaches are ideal for lesions involving the central skull base, clivus, sella, and parasellar regions, such as pituitary adenomas, and avoid the lateral wall of the sphenoid sinus, carotid artery, and optic nerve (Fig. 5).

Skull base defects were initially closed with full-thickness skin grafts when this surgery was pioneered in the 1950s and 1960s. The 1970s brought a shift to using galeal and pericranial flaps and now more commonly the nasoseptal flap is used as a pedicled vascularized flap in the closure of the defects (Figs 6 and 7).

Robotic endoscopic skull base surgery and transoral robotic surgery theoretically provide the advantage of improved visualization, access, and precision, but more clinical investigation is needed before these techniques are widely accepted.

**COMPLICATIONS**

Potential complications of skull base surgery include cerebrospinal fluid leak and the increased risk of...
meningitis associated with it, visual disturbances, intracranial complications from direct injury to the brain, carotid artery, cranial nerves, venous sinuses, and metabolic complications involving the pituitary gland such as diabetes insipidus. Cerebrospinal fluid leak is the most common complication, usually manifesting as rhinorrhea, and can occur in up to 20% of resections. Some can be managed conservatively, usually with a lumbar drain, but postoperative leaks often require surgical closure. Most orbital complications are due to direct injury to the optic nerve or extraocular muscles or from bleeding into the orbit, causing diplopia, decreased visual acuity, or even blindness. Pneumocephalus can occur suddenly in the postoperative period if a patient attempts to blow his or her nose, and tension pneumocephalus can manifest as confusion, obtundation, and neurologic deterioration due to the intracranial mass effect. Though bleeding is a risk in any procedure, there are a relatively large number of important vessels susceptible to injury during skull base surgery including the anterior and posterior ethmoid arteries, sphenopalatine artery, maxillary artery, and ICA and its branches. In a relatively large series of 26 pediatric patients who underwent skull base approaches for tumor resection, Teo et al found an overall complication rate of 57%. Most were due to worsening of preexisting cranial nerve palsies with 37% having permanent sequelae. Another study found a 30% minor complication rate without any major complications or deaths. One complication that is surprisingly absent from reports is retardation of growth as a result of craniofacial resection. This is thought to be because osteotomies do not normally pass through the growth centers of the facial skeleton. In their report on 26 pediatric patients, Teo et al found no long-term disturbances of facial growth patterns after surgical disruption of the skull base.

OUTCOMES
A higher percentage of initial complete resections of anterior skull base tumors has been noted in pediatric patients when compared with their adult counterparts, and is thought to be in part due to better defined tissue planes in children. The higher rate of complete resection, in combination with the high incidence of benign tumors, explains why children have better prognosis with skull base lesions (81–83% 3-year survival compared with 45–80% in adults). Castelnuovo et al found that radical endoscopic endonasal resection led to either complete or partial recovery of quality of life within the first year of surgery. McCoul et al also noted improvement in quality of life in a prospective study.

CONCLUSION
Tumors of the skull base are rare in the pediatric population. Resection is the optimal treatment of skull base benign lesions and for most low-grade malignancies. The surgical approach needs to be individualized based on the age of the patient, tumor location, and its pathological features. As an otolaryngologist who sees many of these highly complex skull base children, it is imperative to have a strong foundation in skull base anatomy, understand surgical options, be facile with the endoscope, and communicate well with the neurosurgeon, ophthalmologist, radiologist, and oncologist.

REFERENCES