Transsphenoidal Surgery for Craniopharyngiomas

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Abstract: Craniopharyngiomas are rare epithelial tumors that arise along the path of the craniopharyngeal duct, account for 2-5% of all primary intracranial neoplasms, and have a bimodal distribution with peak incidence rates in children 5-14 years of age and adults 50-74 years of age. They are WHO grade I tumors, with adamantinomatous and papillary subtypes. The transsphenoidal corridor offers the most versatile approach to the skull base, and the development of expanded endoscopic endonasal transsphenoidal (EEET) approaches has led to the ability to address significantly more craniopharyngiomas via the transsphenoidal route. The approach and extent of the skull base exposure is tailored to the type and location of the tumor.

Location: Craniopharyngiomas most often extend from a suprasellar location in association with the infundibulum, and the vast majority have a suprasellar component. Only small minority of these tumors will be purely intrasellar, and most have suprasellar and intrasellar components. They often abut or adhere to important neurovascular structures located in and surrounding the suprasellar space including the optic apparatus, infundibulum, pituitary gland, floor of the third ventricle, hypothalamus, and vessels of the circle of Willis.

Presentation: The most common presenting symptoms are headache and visual disturbance, usually bitemporal hemianopsia. Hypothalamic-pituitary axis dysfunction, obesity, diabetes insipidus, and growth impairment in children are common at presentation. Hydrocephalus is present in some cases and is a more common in children. Other symptoms include cognitive dysfunction, lethargy, gait disturbance, and focal neurologic deficits.

Evaluation: The evaluation begins with a detailed history and neurologic exam. Any patient with visual complaints, visual deficits identified on exam, or evidence of optic chiasm compression on imaging should undergo a formal neuroophthalmologic examination. All patients should undergo a thorough endocrine evaluation.

MRI: MRI classically demonstrates calcification, cyst formation, and contrast enhancement of the solid portion of the tumor. Tumors with higher protein content tend to be more hypointense on T1WI and with those with lower protein content tend to be more hypointense. They are generally hypointense on T2WI.

Classification: Type I tumors are preinfundibular, type II tumors are transinfundibular, type III tumors are retroinfundibular, and type IV are isolated third ventricular tumors, which are not amenable to transsphenoidal resection. Craniopharyngiomas demonstrate diverse growth patterns, and are often not restricted to a single compartment. Since this classification was developed for suprasellar craniopharyngiomas, primarily sellar subdiaphragmatic lesions are not classified within this scheme.

Type II Craniopharyngioma

CT: Pre-operative CT imaging is helpful for assessing the pneumatization of and position of septa within the sphenoid sinus. A highly pneumatized sphenoid sinus can distort landmarks and place the ICA and ON at risk. A poorly pneumatized sphenoid sinus can make the approach more difficult, but modern instrumentation and neuronavigation make traversing even minimally pneumatized sinuses possible. This is particularly important in the pediatric population, as pneumatization of the sphenoid sinus is a progressive process.

Preparation and Positioning: The patient is positioned supine with the in a headrest with the head extended. Neuronavigation is required for EEET approaches, and a lumbar drain is placed for post-operative CSF diversion with wide skull base exposures. The operative sites are prepared, an arterial line is placed, the nasal cavity decongested, antibiotics and stress dose methylprednisolone are administered.

Nasal and Sinus Exposure: In the case of an EEET approach, the procedure begins with the elevation of a nasoseptal flap. With primarily sellar and infradiaphragmatic tumors, a nasoseptal flap is usually not needed. The bilateral middle and inferior turbinates are lateralized, and the flap is elevated. The bilateral sphenoid ostia are excised and the posterior nasal septum removed. Wide bilateral sphenoidotomies are then made, beginning at the ostia, such that a large single working cavity is created. If an extended approach is being undertaken, bilateral posterior ethmoidectomies are then performed. These expose the planum sphenoidale. Once complete, the exposure extends from the ethmoid sinuses anteriorly to the clival recess posteriorly, and gives a panoramic view of the planum sphenoidale, tuberculum sellae, and sella.

Skull base exposure: The extent of the skull base exposure is determined by the tumor size and location. Primarily sellar infradiaphragmatic tumors can be accessed by a standard endoscopic approach to the sella and removal of the anterior sellar wall, whereas suprasellar lesions require more extensive exposure. The exposure for a suprasellar tumor begins with removal of the tuberculum and upper half of the sella with exposure of the anterior intercavernous sinus. Removal of the middle clinoinds widens the exposure to allow early identification of the ON and paracarotid ICA. Removal of the planum may extend anteriorly about 1.5-2 cm if needed, but should not extend past the rostrum of the sphenoid sinus to avoid damaging the cribiform plate, olfactory nerve fibers, and posterior ethmoidal arteries.

Type I tumors: Removal of the upper sella, tuberculum, and planum is sufficient to access type I tumors. The superior intercavernous sinus is ligated and the dura opened in cruciate fashion. The tumor visible as soon as the dura is opened, and we first incise the capsule and drain any cystic contents. Debunking is then performed, followed by extracapsular dissection.

Type II tumors: In addition to the exposure described for type I lesions, type II lesions require removal of bone over the entire anterior sella and downward retraction of the pituitary gland. This facilitates the more anterior angle needed to access a lesion infiltrating along the infundibulum and extending into the floor of the third ventricle. Transection of the infundibulum may be required to achieve gross total resection.

Type III tumors: Type III lesions are centered behind the chiasm and stalk in the pre-peduncular cistern. Additional inferior exposure is required, extending into the sellar floor, to visualize the inferior intercavernous sinus. The superior and inferior intercavernous sinuses are split, and the diaphragm released, to increase access to the dorsum sella. The bone of the dorsum and posterior clinoinds is then removed, the dura along the cistus opened, and the tumor accessed in the prepuducular cistern.

Reconstruction: For larger EEET exposures, we combine a gasket seal method with fascia lata and a nasal septal flap. A MEDPOR buttress is placed over a fascia latta graft and countersunk into the defect. After placing the gasket seal component, a nasoseptal flap is placed. A fat graft may be used to obliterate the sphenoid sinus prior to placement of the nasoseptal flap. The nasoseptal is secured in place with DuraSeal (Covidien) or fibrin glue. This layer is then buttressed with a Foley catheter balloon, nasal tampons, or other device.

Postoperative Care: Patients with complex closure of large skull base defects typically require lumbar drainage for 24 hours or longer to eliminate the risk of CSF leak and allow for settling in of the skull base repair. If a post-operative CSF leak persists, it should be managed conservatively with continued temporary CSF diversion, head of bed elevation, and a bowel regimen. Permanent CSF diversion may be a reasonable next step in some cases that persist despite conservative management. Diabetes insipidus should be expected and its management anticipated, but will usually resolve over about a month. Anterior pituitary dysfunction will be present in 38-57% of patients. Stress dose methylprednisolone should be continued in the immediate post-operative period. Careful endocrine monitoring and collaboration with an endocrinology team is important; we typically continue steroid replacement for 2 weeks and then reassess with morning cortisol and cosyntropin stimulation testing. The need for thyroid replacement is typically assessed 10-14 days after surgery. Further replacement of other anterior pituitary hormones is determined on a case-by-case basis.

References