Introduction

Cranioopharyngioma represent one of the most challenging brain tumors and pose unique surgical dilemmas when primarily arising from within the third ventricle. Cranioopharyngiomas that reside mostly within the third ventricle rather than the suprasellar cistern are often considered for a midline approach such as endoscopic endonasal, subfrontal trans-laminaerinalis, or interhemispheric transcallosal, to gain optimal access and minimize hypothalamic disruption. Where appropriate, transcortical transforminal endoscopic approaches may also be used for smaller anterior third ventricular tumors. We describe the evolution of our decision-making algorithm (Figure 1) for approach selection to large cranioopharyngiomas of the third ventricle via illustrative cases, demonstrating the importance of combined approaches in order to manage these large and complex tumors with minimal or no morbidity.

Example Case 1

A 41-year-old man presented to neurosurgical evaluation after several months of headache and memory loss. Magnetic resonance imaging revealed hydrocephalus due to a 3-cm mostly solid cranioopharyngioma arising from the anterior third ventricle, without expansion of the pre-infundibular recess (Figure 2A). The patient underwent a stage I interhemispheric transcalfosal approach for resection of the majority of the lesion, relieving the obstructive hydrocephalus. A small residual anteriorly and inferiorly could not be reached safely due to poor visualization of the optic apparatus and anterior circulation (Figure 2B), and so the patient several weeks later underwent stage II endoscopic endonasal transphenoidal resection of the residual, with gross-total resection and resolution of his symptoms (Figure 2C). He is neurologically intact in follow-up.

Example Case 2

A 78-year-old woman with multiple medical co-morbidities presented with blurry vision, memory loss, and polyuria. MRI revealed a mostly cystic cranioopharyngioma with expansion of the pre-infundibular recess of the third ventricle (Figure 3A). The patient underwent a stage I endoscopic endonasal transphenooidal approach with excellent results. A near total resection was achieved with minimal capsule left adherent to the hypothalamus. One year later, she presented with mild recurrent symptoms and a recurrent cyst inside the third ventricle (Figure 3B). She underwent a stage II endoscopic transventricular-transforaminal approach to access the lesion through a virgin, minimally invasive approach due to her medical co-morbidities (Figure 3C). After stage II she returned to her baseline (neurologically intact).

Example Case 3

A 46-year-old man presented with fatigue and hypogonadism. Brain MRI revealed a large 3.5-cm primarily solid cranioopharyngioma occupying the suprasellar space and third ventricle (Figure 4A). He underwent stage I endoscopic endonasal resection, which was quite limited by preservation of the pituitary gland and stalk, preventing a good visualization of the lateral extension of the tumor and the tumor anterior and above the chiasm. Based on our modern algorithm, an interhemispheric transcallosal open approach may have been better suited as the stage I procedure. Expected residual tumor was left within the third ventricle both near the optics and posteriorly (Figure 4B). He was taken back to the OR for a planned stage II subfrontal trans-laminaerinalis approach. The procedure achieved decompression of the optic apparatus. The main surgical limitation of this stage was the superior trajectory, as the anterior communicating artery (ACoA) was found inferior with the perforators to the hypothalamus running above it obscuring the superior-most trajectory. The window of exposure between the chiasm and the ACoA was limited. The patient suffered early progression of minimal residual disease on follow-up MRI (Figure 4C) and he was referred back for additional surgery prior to radiation given the aggressive clinical course of the tumor. A stage III transcortical-transventricular transchoroidal approach was performed with port access for resection with minimal residual on MRI in the hypothalamus (Figure 4D). The patient went on to proton beam therapy with no progression of disease and neurologically intact after 18-months follow-up.

Discussion and Conclusions

Resecting a large third ventricular cranioopharyngioma often is safest for the patient when staged, as no one approach adequately accesses the entire third ventricle. When choosing what approach to start with for a planned staged procedure, we often are guided by the patient’s presenting symptoms as well as the tumor characteristics. Those patients with vision loss and optic chiasm compression should undergo a stage I that addresses this compression such as EEA. Residual asymptomatic tumor, for example within the posterior third ventricle, could then be addressed via a transcallosal route later once recovered from the first surgery. If the predominant symptom is hydrocephalus, we first debulk the ventricular component obstructing CSF flow such as via the transcallosal route. If the floor of the third ventricle is not expanded by the lesion, stalk-sparing EEA techniques are very difficult and an open approach may be preferred. Given the critical and fragile nature of the surrounding structures, and the tendency for cranioopharyngioma to adhere to these structures, we prefer staging the resection to utilize multiple surgical corridors when a tumor is difficult to resect from a single trajectory. Lastly, although we attempt total resection whenever possible, we prefer to leave a small amount of tumor for radiation therapy if removal would lead to injury to the hypothalamus.

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