Endoscopic Endonasal Management of Craniopharyngioma

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KEYWORDS

• Craniopharyngioma • Endonasal • Endoscopic • Transsphenoidal • Suprasellar • Transtuberculum • Transplanum

KEY POINTS

- Gross total resection of craniopharyngioma reduces the likelihood of recurrence.
- Planned subtotal resection +/− adjuvant radiation therapy may be a reasonable treatment approach in selected cases.
- The transsphenoidal corridor and endoscopic, endonasal technique are used to resect not only sellar and sellar-suprasellar but also purely suprasellar intraventricular craniopharyngioma.
- A key aspect of this technique is the expansion of the operative field by incorporation of the transtuberculum and transplanum approaches.
- The primary limitation of the transsphenoidal approach is its lateral reach, and as a general rule, this approach should not be applied to tumors extending more than a centimeter beyond the lateral limits of the exposure.

A video of endoscopic endonasal resection of a supra sellar craniopharyngioma accompanies this article at www.oto.theclinics.com/

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INTRODUCTION

Craniopharynigoma are rare, benign tumors of the central nervous system. Thought to arise from remnants of Rathke pouch in the sellar region, they represent less than 1% of all primary central nervous system tumors. Despite their benign histology, these tumors have posed a significant treatment challenge. Their central location in the sellar and suprasellar region and frequent invasion into critical neurovascular structures, such as the pituitary gland, hypothalamus, and optic apparatus, makes gross total resection (GTR) challenging. Although tumors may still recur after GTR the likelihood increases dramatically with subtotal resection.[1–6] Nevertheless, significant controversy exists regarding the optimal treatment strategy, with some groups pursuing conservative subtotal resection with either upfront or salvage radiotherapy or radiosurgery to lessen the risks of aggressive surgical resection.[1–18]

As these treatment paradigms have evolved, so too have the surgical techniques and approaches to craniopharyngioma, with the goal of maximizing resection and minimizing collateral damage. Traditional transcranial approaches require some degree of brain retraction and manipulation of cerebrovascular structures that lay between the surgeon and the pathology.[8,19] The transsphenoidal approach offers a more direct route to the sellar/suprasellar region and if performed with the operating microscope provides only a limited field of view, which often precludes safe and complete resection of these lesions.[19–23] The use of the transsphenoidal corridor was broadened with the development of expanded approaches, which offered a safe alternative method for reaching suprasellar craniopharyngioma.[24–32] The incorporation of the endoscope was the next step on the evolutionary ladder. By virtue of an improved field of view and superb illumination, this allowed for further expansion of the endonasal corridor. Finally, the development of a variety of complementary multilayer techniques to repair the skull base minimized the risk of postoperative cerebrospinal fluid (CSF) leak.[8,19] Several groups have now published their series incorporating these innovations into a fully endoscopic, endonasal, extended transsphenoidal approach for sellar and suprasellar craniopharyngioma.[2,8,33–42]

Epidemiology and Clinical Characteristics

Craniopharyngioma is a histologically benign, but locally aggressive sellar/suprasellar tumor characterized by a propensity to involve surrounding neurovascular structures. The overall incidence rate is between 1.3 and 1.7 cases per 1,000,000 person-years.[1,5,6,9,13] As is often noted in the literature, there is a bimodal age distribution with an incidence of 1.9 cases per 1,000,000 person-years in children ages 0 to 19 and an incidence of 2.1 cases per 1,000,000 person-years in adults aged 40 to 79.[1,7,10,12,14] The population-adjusted incidence rates suggest that craniopharyngioma has a slightly higher incidence in black persons. There is an even distribution between genders.[1,8,9]

Most craniopharyngiomas are located in the parasellar region. Most involve the suprasellar cistern to some degree. Occasionally the tumor can extend into neighboring cranial fossae and rarely is found in ectopic locations.[19,43] The most common presenting symptoms are related to increased intracranial pressure secondary to obstructive hydrocephalus (eg, headaches, nausea, and vomiting); visual dysfunction from direct compression of the optic chiasm, nerve, or tract; hormonal imbalance from pituitary stalk infiltration; and behavioral/developmental abnormality from hypothalamic injury (occurring primarily in children).[19,43]