Craniopharyngioma is the most common intracranial nonglial tumor that is often associated with cystic components.\textsuperscript{1–3} Although there are instances of craniopharyngioma occurring in adults, these tumors are predominantly found in children.\textsuperscript{1,2} Many MR imaging and computerized tomography studies have demonstrated that most cases involve primarily a solitary cyst, whereas in some cases there is a small tumor with one or more cysts, and a few cases involve purely solid components.\textsuperscript{6} The tumors usually grow on the cisternal surface of the hypothalamic regions; they can also grow from the infundibulum or tuber cinereum on the floor of the third ventricle.\textsuperscript{9} Some tumors even exhibit fingerlike attachments to critical structures such as the hypothalamus, optic nerves, pituitary stalk, and basal cerebrovasculature.\textsuperscript{1} Complete microsurgical resection of the tumor appears to offer the most favorable patient outcome.\textsuperscript{2–4,9,10} Nevertheless, with highly invasive tumors, a neurosurgeon must consider the risk of hypothalamic or major vessel damage caused by aggressive surgery, and may need to consider various other therapeutic options.

Because most cases of craniopharyngioma have a cystic element, reduction of the cyst volume is often desirable. This can be achieved through various methods of aspiration, shunt systems, and introduction of chemotherapeutic elements. An Ommaya reservoir system is commonly used for drainage of the cystic elements; it is placed during open surgery or by stereotactic accesses, which typically involve the use of a neuroendoscope, x-ray films, or intraoperative MR imaging.\textsuperscript{3,11}

Other modalities of treatment for craniopharyngioma, which are often used in conjunction with surgical techniques and/or Ommaya reservoirs, are radiotherapy and radiosurgery.\textsuperscript{1,3,4,6–8,10} It has been reported that good long-term tumor control can be achieved using stereotactic intracavity irradiation.\textsuperscript{1,9,10} A combination of subtotal tumor removal and radiotherapy has been recommended in some studies as having 10-year recurrence-free survival rates comparable to total resection.\textsuperscript{4,10} Gamma knife surgery is currently the preferred method of radiosurgery. The basic concept of radiosurgery is to deliver precisely defined high dosages of radiation and rapid falloff in dose outside the target area, with the hope of reducing treatment-induced morbidity due to radiation-related damage and complications.

A multimodality approach to craniopharyngioma, including minimally invasive stereotactic techniques, microsurgery, conventional radiotherapy, and radiosurgery has been recommended to tackle craniopharyngioma aggressively while minimizing harm to the patient. With all approaches, there are varying levels of risk for endocrinological morbidity, vascular complications, neuropsychological and behavioral disorders, neurocognitive disorders, and learning disabilities. Although many treatment options are available, total tumor resection remains the most commonly performed procedure for treatment of craniopharyngioma, and it is still believed to give the patient the greatest chance of having an independent and productive life with low risk of recurrences. The authors prefer the subfrontal transbasal approach for resection of these tumors, and they describe this approach and illustrate it with the accompanying figures.
Although many treatment options are available, total tumor resection remains the most commonly performed procedure for treatment of craniopharyngioma, and it is still believed to give the patient the greatest chance of having an independent and productive life with low risk of recurrences. The authors prefer the subfrontal transbasal approach for resection of these tumors.

**OPERATIVE PROCEDURE**

Following appropriate and complete consent, the patient is brought into the operating room and placed on the operating table, where general endotracheal anesthesia is induced and intubation is performed. Intravenous and arterial lines are placed as well as a Foley catheter. The patient’s head is affixed in a Mayfield pin headrest in a supine position. A bicoronal incision is marked out from both zygomatic arches and runs behind the hairline when possible (Fig. 1). Minimal shaving of the hair just along the proposed path of the incision is performed. The patient is then prepared and draped in the usual sterile fashion.

A bicoronal scalp incision is made down to the pericranium by using a No. 10 blade. The flap is freed and reflected forward with a moistened sponge underneath the folded scalp and over the top of the reflected flap and held in place with fishhooks. The pericranium is then incised, freed, and reflected forward in a separate layer. A single burr hole is placed in the midline and a large frontal basal bone flap is created using the high-speed pneumatic craniotome and footplate attachments. In some cases, depending on the individual anatomical variation as well as the precise tumor size and location, an additional craniotomy removing the frontal–orbital rim may be performed (Fig. 2).

A No. 15 blade is used to incise the dura mater in a low, transverse incision, which is then extended laterally with Metzenbaum scissors and pick-ups. The anterior aspect of the sagittal sinus is occluded with several interrupted 4-0 Neurolon sutures. Two Fukushima retractors are placed from posterior to anterior and gently retract the frontal lobes posteriorly. We must note here that we prefer bone removal in lieu of brain retraction. Therefore, depending on the individual anatomy, additional bone removal may be performed to minimize brain retraction. Specifically, individual bone structure in relation to the optic apparatus is critical in terms of having a pre- compared with a postfixed chiasm. A prefixed chiasm may require more bone work for adequate exposure, whereas a postfixed chiasm should grant easier access.

At this point bilateral optic nerves are visualized and freed from surrounding arachnoid mater. The tumor may now be identified between the optic nerves, in either the optical–carotid triangles or lateral and deep to the carotid artery (Fig. 3). Careful dissection must take place here with surgical microscissors, various microdissectors, and a No. 11 blade for some sharp dissection. The tumor may extend around the pituitary stalk, to the sella turcica, and posteriorly into the third ventricle. Numerous structures must be identified and preserved, including the optic nerves and chiasm, the pituitary stalk and gland, and the carotid, basilar, anterior cerebral, middle cerebral, posterior communicating, and anterior choroidal arteries. When possible, we attempt to preserve the olfactory nerves as well; however, it is debatable whether this results in useful preservation of olfaction. The tumor is resected as completely yet safely as possible.

The surgical bed and all structures are carefully inspected while hemostasis is achieved. The wound is irrigated with copious amounts of lactated Ringer solution. The dura mater is closely reapproximated with running, locked 4-0 Neurolon sutures, and the vascularized pericranial graft is then attached to the dura with several interrupted 4-0 Neurolon sutures. The bone flap is replaced and secured to its normal anatomical position. The galea is reapproximated with several interrupted, inverted 3-0 Vicryl sutures, and the wound edges are closely reapproximated with skin staples. In lieu of staples in pediatric cases, we will often run a 4-0 Monocryl suture in a subcuticular fashion, followed by tissue glue for skin closure.
Subfrontal transbasal resection of craniopharyngioma

After the operation is complete, the patient’s head is removed from the Mayfield pin headrest. The general anesthesia is reversed and extubation is performed. The patient then proceeds to the postanesthesia care unit or directly to the intensive care unit.

DISCUSSION

Craniopharyngioma is the most common intracranial nonglial tumor found in children, but it rarely occurs in adults. The histological appearance of these lesions may be that of benign epithelial tumors, but they can cause serious neurological deterioration and eventually death. Many MR imaging and computerized tomography studies have demonstrated that approximately 90% of craniopharyngioma cases involve primarily a solitary cyst, whereas in approximately 30% of the cases a small tumor with one or more cysts is found, and approximately 10% of the cases involve purely solid components. The tumors usually grow on the cisternal surface of the hypothalamic regions; these lesions can also grow from the infundibulum or tuber cinereum on the floor of the third ventricle by developing a protrusion from the sellar–suprasellar region to the third ventricle and basal cistern. Many tumors exhibit vast, protrusive attachments to many critical structures such as the hypothalamus, optic nerves, pituitary stalk, and basal cerebral vasculature, which provide ample challenges to the neurosurgeons involved.

It is commonly accepted that complete microsurgical resection of the tumor appears to offer the most favorable patient outcome, and this modality is widely used as the primary approach to treating craniopharyngioma. Some of the surgical techniques that have been used to remove the tumors include interhemispheric, perional, subtemporal, transcallosal, transcortical, and translamina terminalis approaches. Many of these approaches have been well described in the literature. Craniopharyngiomas that extend deep into the third ventricle and basal or preptonee cistern present special surgical and clinical problems because of their deep localization as well as their relationship with hypothalamic structures and perforating vessels.

Because almost 90% of craniopharyngiomas have some form of cystic element, reduction of the cyst volume is often desirable. Various methods of aspiration, shunt systems, and introduction of chemotherapeutic elements have been used to reduce the size of the cysts before further procedures can be performed to remove or eliminate entire tumors. One of the most commonly used methods of cyst drainage is the Ommaya reservoir system. This device is placed during open surgery or by stereotactic surgery, which typically involves the use of a neuroendoscope, x-ray films, or intraoperative MR images. Ommaya reservoirs allow for the aspiration of the cyst and the introduction of alternative therapies for recurrent craniopharyngioma instead of multiple intrusive surgeries. One of the main concerns after any initial treatment has been the rate of recurrence. Ommaya reservoir systems allow for the treatment of recurrent tumors while sparing the patient from open surgery.

Radiotherapy and radiosurgery have been widely used for initial as well as adjuvant treatments; they are also commonly used in conjunction with total and subtotal resections and/or Ommaya reservoirs. There have been numerous studies indicating that good long-term tumor control can be achieved by the use of stereotactic intracavitary irradiation. A combination of subtotal tumor removal and radiotherapy has been suggested in some studies as having 10-year recurrence-free survival rates that are comparable to total resection. Nevertheless, as good as the outcome of radiotherapy may be, it is not without significant risks. There have been reports of radionecrosis, optic neuritis, malignant lesions, cognitive disturbances, and hypothalamic endocrine disorders caused by collateral radiation damage.

Radiosurgery can be considered somewhat as a modified form of radiotherapy. The basic intent of radiosurgery is to deliver a well-defined, high dosage of radiation with high precision and rapid falloff in dosage outside the target area, with the hope of reducing treatment-induced morbidity caused by radiation-related damage and complications. The predominant form of radiosurgery currently in use is gamma knife surgery, in which finely concentrated beams of radiation are directed at the site of the lesion. It has been found to be quite effective for many patients, especially those with smaller tumors that have refractory components or those with recurring tumors.

CONCLUSIONS

Although a surgical approach is widely espoused as the primary tool in the treatment of craniopharyngioma, multimodality approaches including minimally invasive stereotactic techniques, microsurgery, conventional radiotherapy, and radiosurgery have been considered to tackle craniopharyngioma aggressively while minimizing harm to the patient. Various approaches contain varying levels of risk for neuropsychological and behavioral disorders, neurocognitive disorders, learning disabilities, endocrinological morbidity, and vascular complications. The neurosurgeon must always carefully analyze the risks and benefits for
both the short- and the long-term before deciding on the approach that will be used for the excision or elimination of the tumor. Many advanced treatment options are available, and progress is being made continuously on various technologies and techniques. Nonetheless, total tumor resection remains the most commonly performed procedure for treatment of craniopharyngioma, and various modern microsurgical techniques have made it a relatively safe and effective way to remove the tumor and prevent recurrences. Total resection is still believed to give patients the greatest chance of having an independent and productive life, free from recurrences.

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References


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