ABSTRACT

The surgical treatment of craniopharyngiomas is challenging. An optimal surgical approach is extremely important to achieve complete removal of the tumor, which is often the goal of treatment. Conventionally, the endoscopic transsphenoidal approach is used for resection of craniopharyngiomas that are essentially confined to the sellar cavity, or have smaller suprasellar extension. However, the tumors located in the retrochiasmatic space are difficult to remove surgically due to a poor access. Traditionally, various transcranial microsurgical routes have been employed with limited success for resection of retrochiasmatic craniopharyngiomas. The transcranial approaches generally do not provide adequate exposure of the tumors originating in the space under the optic chiasm and nerves. Recently, the extended endonasal endoscopic surgical route, obtained by removal of the tuberculum sellae and planum sphenoidale, has been used with great success in the surgical management of tumors lying ventral to the optic chiasm, including craniopharyngiomas. It offers a direct midline access to the retrochiasmatic space and provides excellent visualization of the undersurface of the optic chiasm. It also allows extracapsular dissection using binostil-bimanual technique and facilitates complete removal of these formidable tumors. In this report, we describe step-by-step, the technical details of the endonasal endoscopic transplanum transtuberculum approach with emphasis on the operative nuances for removal of retrochiasmatic craniopharyngiomas.

Key words: Endoscopic endonasal approach; extended transsphenoidal approach; retrochiasmatic craniopharyngioma; transplanum transtuberculum

Introduction

Retrochiasmatic craniopharyngiomas represent approximately 11-46% of all craniopharyngiomas and pose a difficult surgical problem because of their critical location behind the optic chiasm.[1-5] Tumors in this location grow slowly and tend to extend into the third ventricle superiorly and into the interpeduncular cistern and retrosellar region inferiorly.[4,6,7] Gross-total resection with preservation of neurological and endocrinological functions is the ultimate goal in the treatment of craniopharyngiomas.[8-11] Due to their close proximity to critical neurovascular structures, the surgical removal of retrochiasmatic craniopharyngiomas presents a formidable challenge to neurosurgeons.

Traditionally, transcranial[4,12-22] and transsphenoidal microsurgical[23-25] approaches have been employed to resect these tumors, albeit with considerable limitations. The conventional anterior skull base transcranial approaches usually provide a restricted exposure of retrochiasmatic craniopharyngiomas, which are often hidden behind the anteriorly displaced or prefixed chiasm. The intracranial
Sankhla, et al.: Endoscopic removal of retrochiasmatic craniopharyngioma

Surgical Technique

Patient positioning

After induction of general anesthesia, the endotracheal tube is secured to the left side of face and throat packing is done to prevent collection of blood, cerebrospinal fluid (CSF), and irrigation fluid in the pharynx. A lumbar drain is placed for postoperative controlled drainage of CSF, if necessary. The patient is positioned supine on the operating table with the head either placed on a horse-shoe head rest or fixed in a three-point Mayfield head holder. The head is flexed toward the left shoulder by 15°, and rotated by 15° to the right side to achieve an appropriate trajectory and a comfortable access to the nasal cavity. The head is also extended by 5°–10° to enhance accessibility to the anterior skull base. The head end of the operating table is elevated slightly above the heart level to improve venous return. Frameless stereotactic neuronavigation is registered using preoperative computed tomography or magnetic resonance imaging data. Intraoperative image guidance provides anatomical orientation and is extremely helpful during bone removal, dural opening, and tumor removal. The nostrils are prepared externally with betadine, and the nasal cavity is packed with adrenaline-soaked pledgets for 7–10 min. The thigh is also prepared to harvest autologous fascia lata and fat for closure and reconstruction of the sellar floor. Intravenous antibiotics and dexamethasone are administered.

Endoscopic Endonasal Transsphenoidal Exposure

At our institution, the surgery is performed by a team comprised of a neurosurgeon and an otolaryngologist, both experienced and well-versed with the endoscopic skull base surgery. Both surgeons are positioned on the right side of the patient’s head and use the binostril method. The endoscope is placed in the upper part of the right nostril (12 O’ clock position) by one surgeon, and the other surgeon uses the lower half of the right nostril for suction (6 O’ clock position) and the entire left nasal cavity for instruments required for tumor dissection. We use the bimanual technique, which includes, one surgeon holding the scope in his left hand and saline irrigation in the other, and the other surgeon performing dissection using both hands under the endoscopic guidance provided by the first surgeon. The dynamic visualization provided by manual maneuvering of the scope facilitates a three-dimensional perception of the surgical target.

Using a 0° endoscope with 4-mm diameter and 18-cm length, the otolaryngologist performs the initial part of the approach, which consists of a right middle turbinectomy,
Sankhla, et al.: Endoscopic removal of retrochiasmatic craniopharyngioma

Neurology India / May 2015 / Volume 63 / Issue 3

Figure 1: Endoscopic endonasal approach to retrochiasmatic craniopharyngioma. Note the widened exposure obtained by removal of bone of the anterior wall of sella turcica, tuberculum sellae, and posterior planum sphenoidale. The extended endonasal approach also provides a straight trajectory and increased working angle along the vertical axis of the subchiasmatic-retrochiasmatic tumors

Figure 2: Preoperative T1-weighted contrast enhancing magnetic resonance (MR) images, coronal (a) and sagittal (b) showing a predominantly solid and partly calcified retrochiasmatic craniopharyngioma in a 13-year old boy who presented with headaches, vomiting, and visual impairment. Postoperative coronal (c) and sagittal (d) post-Gd MR study showing gross total resection of the tumor

Figure 3: A plain CT sagittal reconstruction image demonstrating an optimal removal of bone from the planum sphenoidale, tuberculum sellae, and the anterior wall and floor of the sella in endoscopic endonasal tranplanum transtuberculum approach

Figure 4: Surgical exposure: (a) Endoscopic view of the skull base showing important bony landmarks in the roof of the sphenoid sinus including, the planum sphenoidale (PS); tuberculum sellae (TS); floor of the sella turcica (SF); optic protuberance (OP); carotid protuberance (CP); medial optocarotid recess (M); lateral optocarotid recess (L); and the clivus (C). (b) Endoscopic view showing dura under the planum sphenoidale (PD); the superior intercavernous sinus (SIS), which is deep to the tuberculum sellae

The right middle turbinate is removed by making sharp cuts at its roots, that is, at its attachment to the skull base and the lateral nasal wall. Care is taken to leave a small tail at the attachment of the root of the middle turbinate to the skull base. This helps as a landmark, and also prevents iatrogenic CSF leak, which could happen if the turbinate is cut flush with the skull base. A posterior ethmoidectomy is performed on the right side to create enough space for placement of the endoscope and free movement of instruments in the operative field. If needed, a complete ethmoidectomy is performed on the right side to gain a straight access to the lateral wall of the sphenoid cavity. The inferior half of the superior turbinate is also excised bilaterally to expose the sphenoid ostia on both sides.

The vomer and perpendicular plate of ethmoid are excised as the next step, followed by resection of the sphenoid rostrum.
This maneuver provides a straight view into the sphenoid sinus cavity. The sphenoid ostia are then widened laterally and inferiorly. All the bony overhangs limiting visualization of the sellar floor, tuberculum and planum, and the lateral sphenoid wall should be drilled away. This maximal exposure helps to achieve control of the internal carotid artery (ICA) laterally and allows a straight trajectory to the sellar and suprasellar regions after dural opening. Complete removal of the sphenoid mucosa is essential to identify bony landmarks. It also prevents the formation of a postoperative mucocele and provides a necessary bony base for placement of the nasoseptal flap. All septations in the sphenoid cavity are removed with a fine diamond drill.

The medial opticocarotid recess is a very important landmark [Figure 4a-b]. It represents the ventral aspect of a pneumatized middle clinoid process and marks the medial aspect of the parasellar carotid canal and the cavernous sinus, the lateral edge of the sella, and the inferomedial aspect of the optic nerve. It also forms the most lateral extent of the tuberculum sellae. Removal of bone over the medial opticocarotid recess widens the intradural exposure and provides better anatomical orientation by early identification of the optic nerves and paraclinoid ICA, thereby, facilitating easy identification and protection of the superior hypophyseal artery and perforating arteries to the optic chiasm.[38,43,46]

When the medial opticocarotid recess is not clearly visible, the lateral opticocarotid recess and the bony prominences over the carotid artery can be used to localize the medial opticocarotid recess. The sella, which is often normal in configuration, is identified between the carotid prominences on both sides, and the clival recess is visible inferior to the...
sella. In the event of a non-pneumatized sphenoid sinus, as in young children, identification of these landmarks is extremely difficult. Careful drilling of the bone under image guidance is essential until the landmarks become distinguishable.

Transplanum Transtuberculum Exposure

The primary steps of the extended endoscopic endonasal technique that we used in these cases have been described in detail in other publications.[34,38,40,41,44,47] The bone of the sellar floor, tuberculum sellae, and planum sphenoidale (PD) is first thinned down with a high-speed diamond drill and is then removed carefully using a Kerrison's rongeur [Figure 4a–b]. The optic canals mark the lateral limits, whereas the posterior ethmoidal arteries mark the anterior limit of the bony resection. Resection of the skull base anterior to these arteries risks injury to the olfactory fibers and epithelium. Copious irrigation is used during drilling over the medial opticocarotid recess (which is the most critical anatomical landmark) to prevent thermal injury to the optic nerve. Venous bleeding, occasionally vigorous, is usually controlled with gelfoam or surgicel packing, or other local hemostatic agents.

The optimal exposure usually includes bone removal from the carotid artery on one side to the carotid artery on the other side in the horizontal plane [Figure 4a]. The extent of bony exposure in the sagittal plane is determined by the size and location of the tumor and can be further evaluated during surgery using navigation. Craniopharyngiomas confined to the sella require removal of the anterior sellar wall only. In cases of preinfundibular tumors, more bone removal over the tuberculum sellae and PD, rather than the anterior sellar wall, is necessary. It is important to expose the areas above, below, and over the superior intercavernous sinus (SIS) to achieve vascular control and to open the diaphragma sellae. Transinfundibular craniopharyngiomas require a steeper working angle to reach the superior ventricular extent of the tumors, and, therefore, additional bone removal from the anterior sella is often helpful. Extensive bone removal from the sellar floor along the inferior intercavernous sinus, and sometimes the posterior clinoid processes and dorsum sella, may be required to expose the retroinfundibular craniopharyngiomas extending from the infundibulum into the prepontine and interpeduncular cisterns.

Dural Opening

We open the dura in a transdiaphragmatic fashion with two incisions in the dura, parallel to each other, immediately superior and inferior to the SIS [Figure 5a-b]. The SIS is coagulated with bipolar forceps and is divided under direct vision to obtain access to the suprasellar cistern[31] [Figure 5b]. The sellar dura is incised in the midline from the lower horizontal incision and can be extended down to the lower end of the sella, if necessary. The dural edges are coagulated and shrunk to increase the dural opening [Figure 5c]. The dura may also be excised if further enlargement of the opening is required. Opening the dura in the planum and sellar regions in this manner provides an advantage of simultaneously exposing the suprasellar cistern containing the optic chiasm and anterior cerebral arterial complex, the diaphragma sellae, the supradiaphragmatic or subchiasmatic space containing the infundibulum, superior hypophyseal artery and perforating vessels, and the infradiaphragmatic space containing the pituitary gland. It also provides two distinct surgical corridors for tumor dissection, the superior or intra-arachnoidal or suprasellar route, and the inferior or extra-arachnoidal or endosellar route.

Tumor Removal

Our technique of endoscopic craniopharyngioma removal follows the same steps as that of the standard transcranial microsurgery, which includes identification of the tumor, internal tumor debulking, extracapsular dissection in the arachnoid-capsular plane, and protection of the neurovascular structures [Figure 6a-h]. Selection of an appropriate intradural corridor for tumor dissection and an intraoperative decision-making regarding the extent of tumor removal and the associated neurological and endocrinological morbidity are some of the crucial issues that influence the overall outcome.
The classification of craniopharyngiomas according to their location in relation to the infundibulum provided by Kassam et al.\[38\] is simple and useful because it facilitates surgical planning and intraoperative decision-making. Type I or preinfundibular tumors are located anterior to the pituitary stalk, tend to displace the optic chiasm posteriorly and superiorly and are visible as soon as the dura is opened. Tumor removal, thus, can be performed under direct vision through the superior part of the transtuberculum transplanum corridor. Type II or transinfundibular craniopharyngiomas involve the pituitary stalk and usually grow along its axis. These tumors extend superiorly into the third ventricle, and, therefore, need a steeper working angle through the lower part of the bony opening. A gross total resection of these tumors often requires sectioning of the pituitary stalk. Type III or retroinfundibular tumors are located posterior to the stalk and tend to grow either superiorly into the third ventricle (subtype IIIa) or posteriorly and inferiorly into the interpeduncular or prepontine cisterns (subtype IIIb). Multiple surgical corridors, including superior subchiasmatic, straight supradiaphragmatic, and inferior routes through dorsum sellae, either single or in combination, can be used for this group of tumors. Occasionally, a lateral transposition of the pituitary gland or an “above and below” approach is needed for a complete tumor exposure. Type IV craniopharyngiomas are purely intraventricular tumors and should be treated through the transcranial microscopic/endoscopic approaches.

The thin arachnoid layer of the suprasellar cistern over the tumor should be opened sharply, and a plane of dissection between the overlying arachnoid and the tumor capsule must be identified [Figure 6a]. The arachnoid covering is usually in two layers, and it is, thus, important to identify and distinguish the tumor arachnoid plane from the cisternal arachnoid plane. The optimal plane of safe dissection is between the tumor capsule and the tumor arachnoid. An attempt must be made to identify the pituitary stalk and the superior hypophyseal arteries as early as possible to ensure their protection from inadvertent injury [Figure 6a]. The capsule is incised (after confirming its relationship with the optic nerves and chiasm), allowing drainage of the cyst or internal decompression of the solid mass [Figure 6b]. Occasionally, firm consistency of the tumor mass due to the presence of calcification makes tumor decompression slow and difficult.

Once the tumor cyst has been drained, or the mass has been debulked adequately, mobilization of the redundant capsule is carried out using a gentle traction technique similar to one used in microscopic resection [Figure 6c-e]. Likewise, sharp dissection is used to divide arachnoid adhesions from the undersurface of the optic nerves and chiasm while meticulously preserving the perforators supplying the chiasm. Injury to the perforators is the main cause of visual deterioration after craniopharyngioma surgery. The process of capsule mobilization followed by more tumor debulking is continued and repeated until complete relaxation of the tumor capsule is achieved. Care must be exercised to avoid amputation of the tumor capsule prematurely and to ensure that enough tumor capsule is available to be grasped with tumor forceps for providing counter traction during the extracapsular dissection.

The arachnoid dissection is continued on each side of the tumor laterally until the opticocarotid cistern is entered, following which the ICA is traced superiorly preserving the superior hypophyseal arteries and other small perforating vessels. Extra care should be taken in larger tumors that are frequently in contact with the A1-A2 junction, anterior communicating artery, the recurrent arteries of Huebner, and the A2 segments of the anterior cerebral arteries. We recommend the use of a 30° endoscope for capsular dissection at this stage because it allows a direct “looking-up” view of the retrochiasmatic space. We also change the position of the scope from 12 o’clock to 6 o’clock in the right nostril to obtain a superior trajectory and an optimal upward visualization of the area.

As dissection continues posteriorly, great care is required to separate the tumor capsule from the floor of the third ventricle. When the floor of the third ventricle is involved, the tumor dissection is continued from the lateral and superior walls until most of the tumor has been removed. One can inspect the third ventricular cavity with a 30° endoscope for the presence of any residual tumor, which if present, can be removed carefully. When the tumor has significant third ventricular extension and a clear plane of dissection is difficult to find, it is safer to leave some tumor behind to prevent traction injury to the hypothalamic nuclei.

Next, attention should be paid to the inferior pole of the tumor, which is separated from the diaphragma sellae and pituitary gland. The diaphragma is transected if the tumor is extending into the sella. Management of the pituitary stalk is crucial and depends on several factors, including the patient’s preoperative endocrinological status, and probabilities of safe gross total tumor resection based on the intraoperative findings, and the surgeon’s assessment. Ideally, every attempt should be made to preserve the pituitary stalk [Figure 6a-c]. However, if the surgeon’s impression during surgery is strongly in favor of the possibility of a gross total resection, the stalk should be transacted, and a complete tumor removal should be attempted. A poor
preoperative hormonal status may also justify the decision to section the pituitary stalk.[41,44,48] For the same reason, in cases of Type II or transinfundibular craniopharyngiomas, it is preferable to perform a low stalk section to achieve a more aggressive removal of the tumor and plan a postoperative hormonal replacement therapy.[43,44,48] In the event in which the preoperative endocrinological functions are normal and/or when complete resection appears unlikely on inspection during surgery, it is appropriate to leave behind a small portion of the tumor in order to avoid any risk of stalk injury during an attempt at tumor removal.

In most cases, the Liliequist membrane is intact and serves as a protective barrier for the basilar artery, posterior cerebral arteries, and P1 perforating vessels.[49] However, the residual tumor may be occasionally found tethered posteriorly to the mamillary bodies or the basilar artery and its branches in the interpeduncular fossa. Holding the tumor capsule by tumor forceps in one hand, a gentle sharp dissection is performed in the arachnoid covering the mammary bodies, optic tracts, posterior cerebral arteries, posterior communicating arteries and thalamoperforators, and the pituitary stalk, to remove the tumor completely [Figure 6f-h]. The resection cavity is then inspected carefully with an angled endoscope for any residual tumor or active bleeding and is irrigated thoroughly with copious saline as the tumor contents can cause local irritation and may incite chemical meningitis.

Closure

A meticulous closure of the cranial base defect after tumor removal is of critical importance to prevent a postoperative CSF leak, pneumocephalus, and their potential complications. We perform a multi-layered reconstruction to achieve a water-tight closure[47] [Figure 7a-d]. As the first step, we prefer to place autologous fat in the sellar cavity to cover the arachnoid defect [Figure 7a]. Care must be taken to avoid a very tight packing or a very deep placement of the graft. A fascia lata graft harvested from the thigh is then placed intradurally as an underlay graft, carefully tucked underneath the dural edges [Figure 7b]. Wherever possible, a small piece of thin bone or cartilage obtained from the nasal septum, is placed snugly over the bony defect in the skull base [Figure 7c]. The final layer of a previously prepared vascularized pedicled nasoseptal flap is then rotated to cover the entire defect [Figure 7d]. We apply a thin layer of fibrin glue (Tisseel, Baxter Healthcare Corp) over the flap edges to prevent its displacement or migration. The flap is supported by the nasal Merocel (Medtronic Xomed) pack which is left in place for 3–4 days. A controlled drainage of CSF using a lumbar catheter is continued postoperatively for 5 days. We have found this method of closure very effective in preventing postoperative CSF leaks. Recent literature review suggests this technique to be very useful with rates of CSF rhinorrhea as low as <5% in most studies.[50,52]

Complications

Surgical complications of endonasal endoscopic craniopharyngioma removal can be encountered at the nasal level, in the sphenoid sinus, or during the intradural dissection.[52] Extrasellar complications usually include relatively minor problems like nasal bleeding, encrustation, and sphenoid sinus mucocele, or serious events like injury to the venous sinuses, carotid artery or optic nerve. Cautious use of bipolar coagulation, bone drill, and Kerrison’s rongeurs, along with copious saline irrigation can minimize some of these complications. Serious neurological deficits may occur during the intradural dissection, as a result of either direct injury to the neural tissue, or vascular compromise. Pituitary hormonal dysfunction due to injury to the pituitary stalk or hypothalamus, and cognitive and psychological abnormalities as a result of injury to the frontal/temporal lobes or the hypothalamus may also occur occasionally.

Cerebrospinal fluid leakage and sinonasal problems are the most common complications related to the endoscopic approach. The sinonasal sequelae are usually temporary, and most patients return to their normal baseline status over a period of time.[53-55] The postoperative CSF rhinorrhea has generally been a major complication of the extended endonasal approach for craniopharyngiomas and rates as high as 20–58% were reported in the past.[31,56,57] However, with the use of more effective closure techniques like a vascularised nasoseptal flap reconstruction, it has now become possible to reduce CSF leak rates to 0–10% in most series.[33,51,55,57] The overall incidence of CSF leak in our 15 cases of craniopharyngiomas removed through the endonasal endoscopic transplanum transtuberculum approach was 20%, which has, however, reduced significantly in last few years following the improved closure technique, as described in this article. Surgical procedure to close the dural defect was required in 1 patient (6.7%) in the early part of our experience. One other patient (6.7%) developed meningitis and required treatment with intravenous antibiotics.

The most common intracranial postoperative complication is diabetes insipidus, observed in 42–64% of patients.[33,56,57] The postoperative diabetes insipidus is transient in most cases and requires careful monitoring of fluids and electrolytes. However, few patients require hormonal replacement therapy for a long time. Anterior pituitary hormonal dysfunction is experienced in 28–46% of patients.[33,56,57] Postoperative
visual worsening can occur in about 2-7% of patients.[33,56,57] Hyperphagia as a consequence of hypothalamic injury causing morbid obesity, is a well-recognized complication following craniopharyngioma surgery.[51,57] Four patients (26.7%) in our series developed postoperative diabetes insipidus requiring treatment with desmopressin, and 1 patient suffered from panhypopituitarism. Postoperative transient hypothalamic disturbances, visual deterioration, and third nerve paresis were encountered in 1 patient each, with all 3 having recovered completely in a few weeks. None of the patients suffered from a serious vascular injury. Minor nasal bleeding, crusting, and congestions were observed in few patients and all of them were provided treatment based on their symptoms. There were no complications related to the lumbar drain.

Conclusions

The endoscopic endonasal transplanum transtuberculum approach offers a reasonable option for resection of the retrochiasmatic craniopharyngiomas. It has a definite advantage over the traditional microsurgical transcranial or transsphenoidal approaches with its ability to provide better visualization of the undersurface of the optic chiasm, hypothalamus, anterior third ventricle, and the interpeduncular and prepontine cisterns to facilitate a complete tumor removal. The results with respect to the extent of tumor removal and postoperative visual improvement are superior, and the approach-related complications are progressively reducing with increasing experience. With an increasing tendency to accept endoscopic techniques by most neurosurgeons in recent years, we hope that this technical report will be helpful to provide more confidence and guidance in the surgical management of patients with formidable tumors like retrochiasmatic craniopharyngiomas.

References

Sankhla, et al.: Endoscopic removal of retrochiasmatic craniopharyngioma


