CASE REPORT

Interhemispheric Transcallosal Transforaminal Approach and Microscopic Third Ventriculostomy for Intraventricular Cranio- pharyngioma Associated with Asymmetric Hydrocephalus: Case Report and Literature Review

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INTRODUCTION

Craniopharyngiomas account for 2% to 4% of intracranial neoplasms. However, the purely intraventricular variant is rare.¹ Craniopharyngioma usually grow on the cisternal surface of the hypothalamic region. It can also grow from the infundibulum or tuber cinereum on the floor of the third ventricle, developing exclusively into the third ventricle.² These lesions are difficult to treat and the management options are individualized for each patient.³

CASE REPORT

We report a case of a 43-year-old male admitted to our department with complaints of intermittent severe headache and progressive visual loss over the past two months. Physical examination revealed obesity. Visual field testing and perimetry demonstrated tendency to bitemporal visual field loss, predominantly for the right eye. Otherwise, his neurological examination was normal.

Magnetic resonance imaging (MRI) showed large tumor mass, located mainly within the third ventricle that had expanded bilaterally, predominantly to the right. The suprasellar cisterns were obliterated. The lamina terminalis was displaced anteriorly. The lesion obstructed the right foramen of Monroe that caused asymmetric hydrocephalus. It was hypointense on T1 and demonstrated vivid
honeycomb enhancement after gadolinium administration with numerous small cysts dispersed amid the solid tumor architecture (Fig. 1A-C).

The neuroimaging studies provided inconclusive data regarding the pure intraventricular localization of the tumor. For this reason, the patient was initially operated via bifrontal interhemispheric translamina terminalis approach. Intraoperatively, we confirmed that the tumor was entirely intraventricular. The anatomical limitations caused by the extremely short anterior communicating artery (AComA) restricted the surgical corridor and resulted in partial resection. Another obstacle was the solid consistency and large size of the tumor that was not amenable to safe dissection and manipulation. Six weeks later on a second stage, we performed right precoronal interhemispheric...

Figure 1. Preoperative T1 weighted MRI with gadolinium enhancement: A) axial image and C) coronal image – thick white arrow points at the tumor protruding through the right foramen of Monro leading to asymmetric obstructive hydrocephalus; B) sagittal image shows tumor mass located within the third ventricle with obliterated suprasellar cisterns – thin white arrow indicates pituitary gland and stalk.

Figure 2. Intraoperative images: A) tumor mass protruding through the right foramen of Monro (F – fornix; TSV – thalamostriate vein; T- tumor; SV – septal vein; ChPl – choroid plexus); B) microscopic third ventriculostomy following gross total tumor removal (FL – floor of third ventricle; P1 – P1 segment of the posterior cerebral artery).

Figure 3. Photomicrograph (H&E, x100) of the tumor specimen presented as poorly circumscribed nests and trabeculae of epithelium in fibrocollagenous stroma with peripheral cells showing nuclear palisading (white arrows). There is an area of abundant “wet” keratin (*) and calcification (CA). This morphological appearance is consistent with adamantinomatous variant of craniopharynigioma.

transcallosal transforaminal approach by modifying the existing bicoronal skin incision. The tumor was visible through the enlarged foramen of Monro (Fig. 2A). We used meticulous microsurgical dissection to debulk the tumor mass, followed by extracapsular dissection. The tumor was removed in a piecemeal fashion and gross total resection was achieved. The lesion was most adherent to the region of tuber cinereum. At the end of the procedure, we also conducted microscopic third ventriculostomy to shunt the CSF flow to the interpeduncular cistern (Fig. 2A-B). The histological...

Figure 4. Postoperative 4-month follow-up T1 weighted MRI with gadolinium enhancement demonstrating gross total tumor removal: A) axial, B) sagittal and C) coronal images.
examination was consistent with adamantinomatous variant of craniopharyngioma (Fig. 3).

Postoperatively, the patient developed diabetes insipidus and hypocortisolemia. Morning free cortisol levels dropped from 352 nmol/l preop to 33.32 nmol/l postop (range 185-624). Endocrine disturbances were effectively treated with hormone substitution therapy. The MRI at the fourth postoperative month confirmed gross total tumor removal (Fig. 4). At month 10 of follow-up, visual field perimetry indicated complete recovery of vision. The endocrine disturbances were still present. Morning free cortisol levels were 99.19 nmol/l dropping to 38.72 nmol/l in the evening.

DISCUSSION

The craniopharyngioma originates from squamous epithelial remnants of the Rathke’s pouch.1 The localization of the tumor is determined by the embryological development of the suprasellar region.2 The pars tuberalis, while developing from the ventral portion of the Rathke’s pouch, rotates to come in contact with the neuroectodermal layer of the ventral cerebral vesicle from which later develops the infundibulum and the third ventricular floor. The pial membrane, originating from the mesoderm, normally intervenes between the stomodeum and the cerebral vesicles by the fifth week of gestation before the rotation of pars tuberalis. Thus, it excludes the Rathke’s pouch cells from the subpial space. A craniopharyngioma evolving from these cells remains extra-pial. Depending upon the extent of rotation of the pars tuberalis, it can be located in the vicinity of the hypothalamic floor, the pituitary stalk or the posterior pituitary lobe respectively. If the development of the pial membrane is delayed, the Rathke’s pouch cells come directly in contact with the neuroectodermal layer of the ventral cerebral vesicle. A tumor arising from cells in this location, therefore, develops into a purely intraventricular craniopharyngioma.1,4 Our case supports this theory because the tumor was most adherent to the region of tuber cinereum, a fact also observed by other authors.1,5,6 As in our patient, the intra-ventricular craniopharyngiomas usually present at an older age.7

Analysis of neuroimaging data is of great importance for appropriate preoperative planning. The typical MRI features of intraventricular craniopharyngioma include intact third ventricular floor, patent suprasellar cistern, normal pituitary stalk and absence of sellar abnormality. Calcification observed in 50% to 80% of the suprasellar craniopharyngiomas is rare in the intraventricular variety.1,8 As also noted by us, solid tumors may show a more heterogeneous intensity pattern.1 Despite excellent overall survival rates, craniopharyngioma survivors have substantially reduced quality of life because of significant long-term sequelae, notably severe obesity in about 50% of patients and higher rate of cardiovascular mortality.9 Gain of weight was also observed in our case. The patient received substitution therapy for diabetes insipidus and hypocortisolemia that were still present 10 months postoperatively. Sughrue et al. reported that patients with gross-total tumor removal had over 2.5 times the rate of developing at least one endocrinopathy compared to patients receiving subtotal tumor removal.10

The two main surgical approaches for pure intraventricular craniopharyngiomas are the translamina terminalis and the transventricular surgical corridors. The translamina terminalis corridor using subfrontal, pterional, or basal interhemispheric approach has been used to treat suprasellar or intraventricular craniopharyngiomas.11-14 Although this route is appropriate for accessing tumors in the inferior part of the third ventricle, it is not suitable for removal of large lesions.14,15 Initially, we employed bifrontal interhemispheric translamina terminalis approach but due to large tumor size, anatomical limitations and fixed microsurgical angle that provided narrow and deep surgical corridor only partial tumor debulking was achieved. The interhemispheric approach is an ideal procedure to access tumors located in the suprasellar region, the anterior third ventricle and the basal cisterns. Some authors have proposed the splitting of the AComA in cases with large lesions to maximize tumor exposure16-18, but it has only rarely been practiced19. We did not divide the AComA because in our case the artery was too short to accommodate the placement of mini clips. Moreover, intentional AComA splitting may cause damage to perforating branches which can act as the main feeders of the infundibulum, optic chiasm and anterior hypothalamus.19-21 In addition, the utilization of the translamina terminalis approach places the optic pathway, columns of fornix, supraoptic nuclei, organ vasculosum, and tuber cinereum at risk because of retraction injury or perforator damage.1,2,22 However, Maira et al. consider that translamina terminalis route is a valid choice for the removal of purely intraventricular craniopharyngiomas without significant surgical-
related sequelae. On a second stage, we performed right interhemispheric transcallosal transfornaminal approach that allowed access to the third ventricle with minimal brain retraction. This route to the third ventricle is well tolerated and relatively easy to perform, but the operative distance is greater than in the basal approaches. Damage to the columns of fornix may cause memory deficits, and injury to the veins of the superficial or deep venous systems may give rise to venous infarction. We have chosen the right-sided surgical corridor because the right foramen of Monro was considerably enlarged by the tumor itself. (Fig. 1A, 1C; Fig. 2A). This approach provided us with excellent multi-angled microsurgical control of the dissection plane between the tumor and the ependymal surface during removal. In this way, we avoided the main disadvantage of the translamina terminalis route, which provides access to the third ventricle through a small opening and a very deep surgical corridor despite it offers early control to the tuber cinereum area. This approach was previously employed by Fukushima et al. who performed the same staged procedure. In case the tumor is located predominantly in the body of the third ventricle, the transcallosal approach may be combined with the subchoroidal or the interfornicial approach to access the third ventricle with high efficacy and safety. Yano et al. combined interhemispheric transcallosal approach assisted by flexible endoscope for successful tumor removal. They pointed out that this procedure is a safe option for removing third ventricular tumors, especially in cases with hydrocephalus. We consider that microscopic or endoscopic third ventriculostomy following total tumor removal could provide avoidance of shunt placement for cases with associated hydrocephalus as presented here.

CONCLUSION

Transcallosal transfornaminal approach combined with microscopic third ventriculostomy can be successfully employed for the removal of large purely intraventricular craniopharyngiomas associated with obstructive hydrocephalus. We recommend that large tumor size and anticipated solid consistency in combination with short anterior communicating artery should favor the selection of transventricular surgical approach. Thorough preoperative assessment of data from neuroimaging studies, including cerebral angiography, is crucial in such cases.

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Транскаллозальный трансформационный подход к интравентрикулярной краниофарингиоме

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Мы сообщаем о случае солидного адамантиноматозного варианта краниофарингиомы, полностью расположенного внутри третьего желудочка, вызывающий асимметричную обструктивную гидроцефалию у 43-летнего пациента. Пациент жалуется на периодические сильные боли и прогрессирующую билатеральную потерю зрения. Первичным поражением было достигнуто в бифронтальном межполушарном транслюминальном доступе, но полное исключение не представлялось возможным из-за короткой передней артерии, которая ограничивала доступ. На втором этапе мы использовали правосторонний межполушарный транскаллозальный трансфораминальный доступ и добились полного удаления опухоли с последующей микроскопической вентрикулостомией третьего желудочка. В этой статье обсуждается выбор подходящего хирургического подхода на основе углублённого обзора литературы, который обеспечивает благоприятный хирургический подход к лечению этих редких поражений.