Original Article

Trans-Cerebellomedullary Fissure Approach for Fourth Ventricular Tumors, Outcome Analysis in Thirty Consecutive Cases

Ashraf Mohamed Farid and Sherif Elsayed Elkheshin*
Department of Neurosurgery, Tanta University, Egypt

ABSTRACT
Background: The use of anatomical clefs in the cerebellomedullary fissure to approach the fourth ventricle eliminates violation of neural tissue. The median inferior sub-occipital approach along the “tonsilouveal” sulcus to treat fourth ventricle lesions was described. The approach through the cerebellomedullary fissure to the tela choroidea and inferior medullary velum provides an additional means to access the fourth ventricle and has been used by an increasing number of neurosurgeons. Objective: Evaluation of the efficacy of using the natural clefs in the posterior fossa through understanding the microsurgical anatomy of this region instead of violating functioning neural tissues and preservation of vascular structures. Planned total removal of the tumors is our aim, and if not possible, the maximum amount of tumor tissue to be removed with minimal morbidity. Patients and Methods: The study included thirty patients. All of them were subjected to history taking, complete neurological examination and imaging. Total tumor removal was the main surgical strategy. The uvulotonsillar and medullotonsillar spaces on both sides were opened along with the lateral and deep dissection around the surfaces of the tonsil and the biventral lobule without dividing any neural tissue, thus, overcoming the drawbacks of trans-vermian approach. Results: All of our patients suffered from pre-operative hydrocephalus and large tumor volume sufficient to obstruct the cerebrospinal fluid (CSF) pathways. We resorted to perform CSF diversion procedure in a total of twenty three patients. We were in need for upper cervical laminectomy in 14/30 (47%) due to both tonsillar herniation and tumor descent. Gross total resection could be achieved in 23/30 (77%). Transient post-operative mutism occurred in 4/30 (13%), while transient oral-pharyngeal apraxia occurred only in one patient (3%). The outcome according to Karnofsky scale was 100 in 20/30 (66.7%), 90 in 4/30 (13.3%), 80 in 2/30 (6.6%), 70 in 2/30 (6.6%) and 10 in 2/30 (6.6%). Conclusion: Pre-operative CSF diversion is safe. The telovelar approach provides adequate surgical exposure of the floor of the fourth ventricle and additional access to the superolateral recess with only a limited decrease in working angles when approaching the rostral portion of the ventricle. Preservation of the posterior inferior cerebellar artery (PICA) is so important. Full exposure could be achieved without removal or incision through the neural tissue of the cerebellum and good quality of life and complete cure became possible.

INTRODUCTION

The development of posterior fossa surgery is a hallmark contribution of Harvey Cushing’s to pediatric neurosurgery. Before Cushing, posterior fossa lesions were not considered for surgical resection and only osseous decompression was offered. The aim of tumor resection is to remove as much tumor as possible while preserving neurological function. Complete tumor resection, especially in malignant tumors, offers better conditions for adjuvant therapy options and in some cases, cure is possible. Splitting the vermis, although is a common surgical option to remove giant midline tumors; it results in various neurological deficits.

Matsushima et al. explained the methods to open the cerebellomedullary fissure to approach the fourth ventricle into three types: (1) Extensive (aqueduct type) for large sized extensive lesions that are located deep in the interior. In such tumor location, the uvulotonsillar and medullotonsillar spaces on both sides are dissected and the tonsils and/or uvula should be retracted superolaterally to achieve good visualization. (2) The lateral wall type, which is located apart from the midline. In this type of opening method, the tonsil and the inferolateral surface of the uvula on the lesion side are retracted to obtain appropriate visualization. Therefore, the uvulotonsillar and medullotonsillar spaces should be completely dissected on the targeted side. (3) The lateral recess opening method is used when a lesion is located in the most lateral portion of the fourth ventricle. Here, a lateral & deep dissection around the tonsils and the biventral lobule is required, with retraction of both structures.

PATIENTS & METHODS

Study population: This study included thirty patients; nineteen pediatric (63%) and eleven adults (37%). Twenty-one males & nine females were operated in our department in the period between November, 2011 and March, 2015. History was taken and neurological examination was carried out. Magnetic resonance imaging; axial, sagittal and coronal T2 and...
T1 with & without intravenous contrast agent was the neuroimaging modality.

**Imaging interpretation:** Tumor size (measured as the maximum tumor diameter) located in the posterior fossa mid-line was evaluated and graded as; Grade I (<0.5cm), Grade II (0.5-1.5cm) Grade III (1.6-3cm) while Grade IV (more than 3cm). Preoperative supratentorial hydrocephalus was taken in consideration for planning a possibility of CSF diversion procedure.

**Surgical procedures:** Preoperative CSF diversion using ventriculo-peritoneal shunt was carried out in nine patients with advanced hydrocephalus because of the insidious onset of deterioration of their conscious level and advanced papilledema. We took in consideration the cautious insertion of the ventricular catheter without rapid egress of CSF. Prior to the definitive surgery, an informed consent was signed, based upon the conclusion of the study of Kondziolka et al.\(^4\). Anesthetic considerations were taken, aiming at optimum cerebellar relaxation. Following exposure of the suboccipital surface of the cerebellum, we performed cervical laminectomy in sixteen (53.3%) of our patients according to the level of descent of the tonsils, and the caudal end of the tumor. Regarding patients without preoperative ventriculo-peritoneal (VP) shunt insertion, an intraoperative external ventricular drainage (EVD) was used to control the elevated intracranial pressure. Using a small dural incision near the region of the cerebellomedullary fissure, a gradual release of CSF after opening of the cerebellomedullary cistern is carried out, after adequate relaxation of the cerebellum, the durotomy was then completed.

**Fig. 1a-d:** Shows operative steps. a: Dissection along the right uvulotonsillar space with the right tonsil (asterisk) and contralateral PICA (white arrow) seen. b: Further dissection reveals the edge of Magendie Foramen (white arrow head) with fourth ventricular choroid plexus seen through it. c: Dissection of the fourth ventricular tumor (double arrow head). d: Inspection of the aqueduct (black arrow head) with free egress of CSF seen after gross total removal of the tumor.

We aimed at identification of the Foramen of Magendie; and according to the microsurgical anatomy of this region \(^5\)^\(^6\) we aimed at dissection cautiously at the edges of the foramen of Magendie in the tela choroidea alongside the uvula upward to inferior medullary velum (Figure 1). Actually, the uvulotonsillar and medullotonsillar spaces on both sides were opened along with the lateral dissection around the surfaces of the tonsil and the biventral lobule without dividing any neural tissue; this resulted in further release of CSF.

When the tumor has a remarkable growth, outside the fourth ventricle, the higher magnification of the operating microscope, was used. A central debulking performed using ultrasonic surgical aspirator then microdissection and separation of the lower part of the mass, from the cervicomedullary junction and thus identification of the lower medulla & the cervical cord, with early identification of the telovelotonsillar segment of the PICA on both sides that rotates around the inferior pole of the tonsils and are attached to the lateral
sides of the mass. Microdissection and separation of the lateral sides of the mass from the medial trunks of the PICA on both sides was performed. We reported a noteworthy tumor mass extending up to C5 segment (Figure 2). In this patient we extended the laminectomy to the lower end of the mass and then separating it from the cervical cord till the cervicomedullary junction as a first step followed by excision of the posterior fossa mass as mentioned above. After adequate internal decompression, a cautious separation of the capsule that lies on the floor of the fourth ventricle was applied under strict monitoring of the cardiac rate. Finally, with further lowering of the head of the operating table, in addition to maximum possible patient head flexion, it was easier to dissect the remaining capsule with the visualization of the CSF egress from the aqueduct (Figure 1-D). Irrigation with warm tepid saline to remove remaining blood clots was performed. Then the wound was closed after putting a hemostatic agent.

Post-operative follow-up: Early: a cautious extubation was applied without straining. Close observation & monitoring of the neurological status & the vital signs were carried out and then whenever possible a routine post-operative CT scan was done as a baseline. A communication with the oncologist was emphasized prior to discharge. Unfortunately, all intra-operative EVD were converted into a ventriculoperitoneal shunt in all cases because of persistently elevated intracranial pressure more than three days.

Late: regular monthly interval; either by outpatient visit or by telephone questionnaire. Karnofsky outcome scale was applied to our patients.

Fig. 2a-f: a&b: are preoperative sagittal T1 MR Images showing tumor mass (ependymoma) reaching C5. c: Postoperative sagittal CT image showing complete tumor resection. d: tumor mass after removal. (E&F) Shows preoperative and 2-year postoperative MR images for another patient showing complete resection of fourth ventricular edulloblastoma.

Fig. 3a-c: a&b: Preoperative sagittal T1 MR Images showing tumor mass (Medulloblastoma) and marked Hydrocephalus. c: 4-year postoperative MRI with contrast.
RESULTS

All of our patients suffered from pre-operative hydrocephalus and large tumor volume (grade IV) sufficient to obstruct the CSF pathways. CSF diversion was carried out in 23/30 (77%) while seven patients were managed by tumor attack without the need of CSF diversion procedures. Preoperative CSF diversion using Ventriculoperitoneal shunt was carried out in 23/30 (77%). Transient tonsillar herniation and tumor descent. Gross total upper cervical laminectomy in 16/30 (53%) due to both late VP shunt insertion (Total =23). We were in need for VP shunt with addition to two patients that underwent late VP shunt insertion (Total =23). We were in need for upper cervical laminectomy in 16/30 (53%) due to both tonsillar herniation and tumor descent. Gross total resection could be achieved in 23/30 (77%). Transient post-operative mutism occurred in 4/30 (13%), while transient oral-pharyngeal apraxia occurred only in one patient (3%). Only one patient developed shunt infection 4 months after surgery and radiation. Histopathology revealed medulloblastoma in 21/30 (70%), ependymoma in 7/30 (23%) while in 2/30 (7%) revealed astrocytomas (Table 1). Higher incidence of total resection is associated with medulloblastomas than other neoplasms (Figure 3). The outcome according to Karnofsky scale (table 2) was 100 in 20/30 (66.7%), 90 in 4/30 (13.3%), 80 in 2/30 (6.6%), 70 in 2/30 (6.6%) and 10 in 2/30 (6.6%).

Table 1: Relation Between Extent of Resection and Tumor Pathology

<table>
<thead>
<tr>
<th>Extent of Resection</th>
<th>Tumor Pathology</th>
<th>Medulloblastoma</th>
<th>Ependymoma</th>
<th>Astrocytoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>21</td>
<td>2</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Subtotal</td>
<td>0</td>
<td>5</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>

The operative findings that lead to subtotal resection revealed that in six patients, there was an amount of tumor tissue that was so adherent to the brain-stem and encasing the PICA that it was impossible to resect totally without risk, while in one patient, a small fragment was adherent to the lower medulla, near the obex. There was an arterial feeder seen to be originated from the PICA anterior segment and coursing around the lower medulla to supply the tumor; under magnification, it was cauterized near the site of attachment of the remaining tumor piece that was left. The patients in whom gross total removal was achieved, the separation of the tumor mass from the 4th ventricular floor was easy and no changes in the heart rate were reported.

Table 2: Relation between Extent of Resection and Karnofsky Scale

<table>
<thead>
<tr>
<th>Extent of Resection</th>
<th>Karnofsky Scale</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>100</td>
</tr>
<tr>
<td>Total</td>
<td>19</td>
</tr>
<tr>
<td>Subtotal</td>
<td>1</td>
</tr>
</tbody>
</table>

High scale is not necessarily associated with total resection

DISCUSSION

Owing to the association of hydrocephalus in our work, we found it necessary to highlight the strategy of management of such condition to help optimum definitive tumor removal procedure. In our work, 50% had pre-operative placement of CSF diversion procedure (VP-shunt) due to deterioration in the conscious level attributable to hydrocephalus, together with papilledema. Shimooji et al., 2009, was not supporting Pre-operative VP shunt, while Deborah et al supported such procedure. They concluded that the factors determining the need for ventriculoperitoneal shunts after posterior fossa tumor surgery included young age at diagnosis (63% of pediatric age in our work), duration of symptoms, extent of hydrocephalus, tumor location in the mid-line (all our patients had mid-line tumor), subtotal tumor resection, (in our work, we had 23% subtotal resection), presence and duration of an external ventricular drain, flow of cerebrospinal fluid (CSF) through the fourth ventricle after tumor resection, (achieved in our work in patients with total resection), presence of hemostatic cavity linings, (we used hemostatic linings in all patients), method of dural closure, (in-lay technique in all our patients), tumor type (70% of our patients revealed medulloblastoma), last, CSF infection, CSF leak, and pseudomeningocele formation. We didn't report any complication related to CSF diversion procedure except one patient, who developed 7-month post-operative and post irradiation abdominal catheter exposure and manifestation of shunt infection. Up till now, none of our patients developed extraneural spread, as reported in the literature. We didn't try ETV owing to convenience with the opinion of Duong et al.16

The prone position was applied for our patients and we had the ability to lower the head of the table through rotation around its transverse axis to visualize the most rostral structures. We separated the lower end first for early identification of the fourth ventricular floor and we also could early identify the PICA rotating around the lower pole of the tonsils. Matsushima et al., concluded that trans-cerebellomedullary fissure approach provides a clear observation of the boundary between the tumor and the floor of the fourth ventricle, thus making the surgery safe and effective. The point of
view of Matsushima et al, however, about this approach was that it requires the Concorde position to observe the interior of the fourth ventricle from the postero-inferior side.

In our study, Intravenous anesthesia was planned aiming at getting cerebellar relaxation, together with CSF release from the fissures, in spite of that, post-operative mutism occurred in 4/30 (13%), while oral-pharyngeal apraxia occurred only in one patient (3%). All of them were transient. Wells et al.\(^{11}\) determined the factors associated with development of cerebellar mutism syndrome (CMS) which was associated with pre-operative brainstem invasion, postoperative edema (92% of their series) & damage to the cerebellum and brainstem and involvement of the cerebellomedullary angle & cerebellar peduncles. In our work, patients developed post-operative mutism & oral-pharyngeal apraxia were of pediatric age and no brainstem invasion but there was involvement of the cerebellomedullary angle. We could attribute this also to cerebellar edema due to operative manipulation specially they developed their symptoms on the second post-operative day in comparison to the direct damage that results in immediate post-operative affection and might be permanent.

In our work, transient cerebellar mutism syndrome (CMS) occurred in 13% of our patients, all of them occurred on the second post-operative day; half of the them lasted 2 months while the other half lasted 2-3 weeks. All of our patients with CMS were in pediatric age, and the histopathology was medulloblastoma. Wells et al.\(^{11}\), reported that CMS is reported to affect approximately 25% of patients undergoing resection for medulloblastoma. CMS was initially believed to be either transient or persistent neurological and neurocognitive impairment. Large inferior vermian incisions were also considered the cause, nevertheless, avoidance of vermis-splitting has not prevented CMS.

Mark Souweidane,\(^{12}\) reported, regarding the post-operative edema, that the time needed for edema to spread to the critical structures may be reflected by the delayed onset of the speech impairment that is commonly observed in patients with CMS. They recommended the routine use of brain and cranial nerve electrophysiological monitoring, minimal retraction, avoiding the use of ultrasonic surgical aspiration and postoperative steroids that lessen the incidence of edema. Dailey et al.\(^{13}\) had similar opinion; moreover, the time of recovery from oral-pharyngeal apraxia is variable from 1 weak up to 2 years. In our work, the patient who developed oral pharyngeal apraxia, recovered after one week. We are also convinced by the anesthetic measures that lead to relaxation of the cerebellum, moreover, adequate flexion of the patient's head and lowering the head of the operating table may also lessen the retraction on the vermis by improving the viewing angle. We mentioned in (Table 2) that high Karnofsky scale is not necessarily associated with total resection, as minor symptoms might result from post-operative edema that might lessen the scale, even if transient and improving but less than normal.

Hermann et al.\(^{2}\) reported that mutism can be avoided by approaching posterior fossa tumors with the use of the cerebellomedullary fissure approach, without vermian splitting. Rajesh et al.\(^{14}\) reported that significant incidence of postoperative ataxia and mutism is seen with the telovelar approach in large tumors, and it could be avoidable by staged dissection of the uvulotonsillar cleft, initial arachnoid release in the uvulotonsillar cleft, facilitating easier retraction of the tonsils followed by decompression of the tumor, and further complete dissection of the uvulotonsillar cleft.

Rhoton.\(^{15}\) reported that occlusion of veins around the tonsils, on the lower vermis, and near the inferior part of the roof of the fourth ventricle, including the vein of the cerebellomedullary fissure, could result without sequela. In our work we preserved these structures by thorough microscopic dissection with relaxed cerebellum. In all of our patients, tumors were of Grade IV, the PICA was bordering the lateral capsule, and it was identified caudally, as it rotates around the lower pole of both tonsils and started our dissection from the caudal aspect first.

In our work, we could control the operating table and rotate it around both longitudinal and transverse axes to get additional working and viewing area. Vivek et al.\(^{16}\) concluded that the telovelar approach provides a greater working area and superior access to lesions at the lateral part the fourth ventricle and to lesions in close proximity to the foramen of Luschka compared with the trans-vermian approach. The horizontal angles for the obex and in the rostral and lateral portions of the fourth ventricle were significantly better for the telovelar approach with and without removal of the C1 arch than for the transvermian approach. In our work, patients who underwent upper cervical laminectomy got benefit for improving the viewing angles. Moreover, we observed that the thoracic kyphosis also an obstacle for improvement of the viewing angles. Of course it is variant from a patient to another.

Tannrover et al.\(^{17}\) through cadaveric study, reported that the main limitations of the exposure through this approach were the superior medullary velum and the junction between the pyramid and uvula, the broadest strip of vermis within the posterior incisural space that may limit the lateral and medial resections of the tonsil and the uvula at the uvulotonsillar space, respectively. The operative angle was limited by the uvulopyramidal connection in all cadaveric study cases.

Longatti et al.\(^{18}\) reported, more anatomical structures could consistently be identified by endoscopic inspection of the fourth ventricle. Neuroendoscopy offers a panoramic view of the fourth ventricle, and compared with the microsurgical view it seems to provide better visualization of the structures located in the inferior triangle. We consider this opinion for future
work to improve tumor total resection, inspect the fourth ventricular cavity and confirm patent CSF flow through the aqueduct.

**CONCLUSION**

The Cerebellomedullary fissure approach has the advantage of being through the natural cleavage planes in approaching lesions within the fourth ventricle without incising or removing parts of the cerebellum. We advise the pre-operative insertion of VP shunt for rapidly deteriorating patients attributable to hydrocephalus. Controlling of the viewing angles is possible through the proper patient positioning and operating table adjustment. Efforts should be done for early identification of the 4th ventricular floor and the PICA at the lower pole of the tumor, efforts should be done to minimize the risk of post-operative mutism and ataxia as we use the natural planes. Therefore, through this approach; large sized lesions could be excised. Gross total removal is possible unless there’s brainstem invasion. Good quality of life is also possible.

**Declaration**

The author(s) declare no conflict of interest or any financial support and confirm the approval of the submitted article by the concerned ethical committee.

**REFERENCES**

7. Greenberg MS: Outcome assessment, in Handbook of Neurosurgery: Greenberg MS (ed), Thieme Medical Publisher, New York, NY, USA; 1182. 2010