Neuroendoscopic Transventricular Approach for Cystic Craniopharyngioma

Mohammad Hassan A. Noureldine 1, Sajjad Khodmehr 2, Mohammadmahdi Sabahi 3, Puya Alikhani 4, George I. Jallo 5, Mahdi Arjipour 6, 7

1. Neurosurgery and Brain Repair, University of South Florida Morsani College of Medicine, Tampa, USA  2. Neurosurgery Research Group (NRG) Student Research Committee, Hamadan University of Medical Sciences, Hamadan, IRN  3. Neurological Surgery, Neurosurgery Research Group (NRG) Student Research Committee, Hamadan University of Medical Sciences, Hamadan, IRN  4. Neurosurgery and Brain Repair, University of South Florida, Tampa, USA  5. Neurosurgery, Johns Hopkins All Children’s Hospital, Baltimore, USA  6. Neurosurgery, Brain and Spinal Cord Injury Research Center, Neuroscience Institute, Tehran University of Medical Sciences, Tehran, IRN  7. Neurosurgery, School of Medicine, Hamadan University of Medical Sciences, Hamadan, IRN

Corresponding author: Mahdi Arjipour, mahdianripour@yahoo.com

Abstract

The literature is rich with many studies reporting different treatment modalities and approaches for cystic craniopharyngioma (CC), including microsurgery, neuroendoscopic transventricular approach, endoscopic transnasal surgery, stereotactic drainage, and Ommaya reservoir insertion. The goals of this manuscript are to report the successful treatment of an atypical case of CC using the neuroendoscopic transventricular approach (NTVA) as well as discuss the different surgical modalities for these tumors following a comprehensive review of the literature. Our patient is a nine-year-old female with a large CC who was managed using the NTVA. No complications or recurrence occurred over two years of follow-up. Results of our literature review showed lower recurrence and complication rates of the NTVA compared to other surgical modalities. The NTVA is potentially efficient, reliable, and safe for managing CC and cystic-dominant craniopharyngiomas, with low recurrence and complication rates compared to microsurgery and Ommaya reservoir insertion. Future randomized clinical studies comparing the various treatment modalities of CC are needed to solidify these conclusions.

Categories: Neurosurgery, Oncology
Keywords: ommaya reservoir, microsurgery, neuroendoscopy, cystic, craniopharyngioma

Introduction

Craniopharyngiomas arise from squamous cells of Rathke’s pouch. These benign tumors account for 10% of pediatric and 2-4% of intracranial brain tumors, are located in the intra- and suprasellar zones, and are classified as cystic, solid, or mixed [1]. Tumoral attachment to critical neurovascular tissue, such as the hypothalamus or pituitary, is a treatment challenge. Although endocrine dysfunction could be seen in total resections, the risk is increased in partial resections as well [2]. Total or partial surgical resection remains the treatment of choice, and radiation therapy may play a role in partial resections or recurrences. Neuroendoscopic transventricular fenestration and placement of an intracystic catheter with an Ommaya reservoir is a valid, minimally invasive technique in the management of large cystic craniopharyngiomas (CC) [3-4]. The objectives of this paper are to report the management of an atypical CC using the neuroendoscopic transventricular approach (NTVA), as well as discuss the different surgical modalities for these tumors following a review of the literature.

Case Presentation

The patient is a nine-year-old female who presented with headache, agitation, urinary frequency, and progressive visual deficits over the past two years. A large sellar and suprasellar lesion was detected on a CT scan, and a ventriculoperitoneal (VP) shunt was placed to relieve the progressive symptoms of hydrocephalus (Figure 1). MRI solidified the initial suspicion of CC, where the lesion was hyperintense on T1- and T2-weighted images, without any significant, enhancing solid component on postcontrast T1-weighted images, consistent with the high protein and cholesterol contents of CC (Figure 2).
FIGURE 1: Head CT scan images reveal a massive cystic lesion with extension from the anterior to posterior fossa and upward to the lateral ventricles.

Cystic contents are iso-to-hypodense and peripheral punctate calcifications (arrows) are evident (A-C). Diffuse periventricular hypodensity due to interstitial edema (D, arrow) initially persisted despite shunt placement (E & F, arrows).
A few days prior to surgery and despite no signs of VP shunt failure, the patient’s exam was notable for acute confusion, severe ataxia, and bilateral blindness. The NTVA was performed using a rigid neuroendoscope (Aesculap AG, Tuttlingen, Germany). The cyst was accessed through a right frontal burr hole, and the contents were drained. The neuroendoscope was advanced to the right lateral ventricle, where cyst wall calcifications were visible through the thinned and elevated neural elements. The choroid plexus was then identified and tracked back to the foramen of Monro. The cyst wall was sampled and the cyst was punctured, draining a copious gray-to-green liquid content with debris. Simultaneous irrigation with a Ringer’s solution and suctioning cleared the contents of the cyst; adjacent neurovascular structures were clearly visible at the end of the procedure.

The postoperative course was uneventful, and all symptoms improved in the immediate postoperative period, except for visual loss. Histopathology confirmed the diagnosis of CC. The patient did not develop any symptoms of chemical meningitis on close follow-up. Her pituitary function remained normal postoperatively as well as on long-term follow-up. We opted not to pursue any additional surgical or adjuvant therapy due to the lack of any obvious solid tumor component on postoperative MRI, very thin

FIGURE 2: MRI of the brain

Brain MRI showing a diffuse hyperintense signal on axial fluid-attenuated inversion recovery (FLAIR) (A) and axial T2-weighted images (B), which is consistent with high levels of protein and cholesterol. Sagittal (C) and coronal (D) sections of postcontrast T1-weighted images reveal no significant enhancement of the solid component. All these findings support the diagnosis of cystic craniopharyngioma. The lesion extended from anterior to posterior fossa and laterally to the middle fossa, compressing and elevating all basal elements (frontal lobes, temporal lobes, third ventricle, brain stem). The chronically compressed pituitary infundibulum has been elongated and pushed superiorly as shown (arrow in C).
residual cyst wall, and extensive basal extension with involvement of the critical structures (Figure 3). Follow-up MRIs showed no evidence of tumor regrowth or re-accumulation of cystic contents (Figure 4).

FIGURE 3: Immediate postoperative MRI
Sagittal (A) and coronal (B) postcontrast T1-weighted images showing complete evacuation of cystic contents, decompression of neurovascular elements, and no obvious solid tumor component.

FIGURE 4: Follow-up MRI at two years
Axial (A), sagittal (B), and coronal (C) postcontrast T1-weighted images revealing no re-accumulation of cystic contents. The elongated infundibulum of the pituitary gland is noted (arrow in B).

Discussion
In this paper, we report a child with a large CC, which was treated via NTVA only with no complications or recurrence after two years of follow-up. As there is a lack of consensus regarding the optimal surgical approach to manage CC, we conducted a literature review about the different treatment modalities and summarized our results in tables.

We found 17 reports that utilized the NTVA (with and without Ommaya reservoir insertion) for the treatment of CC in 67 patients, with a follow-up period ranging from six to 73 months; the recurrence rate ranged between nil to 54% (Table 1) [2-5,5-19].

| Author(s) (year) | N | Age (yrs.) | Characteristics | Location | Signs & Symptoms | Surgical drainage | Radiotherapy | Follow-up (mo.) | Recurrence | Comments |
|------------------|---|------------|-----------------|----------|-----------------|------------------|-------------|----------------|------------|---------|
| Hellwig et al. (1995) [5] | 5 | NR | Cystic; mixed | NR | NR | Endoscopy (CSF) | No | NR | No cyst recurrence | 40% reported for the solid portion |

2021 Noureldine et al. Cureus 13(9): e18123. DOI 10.7759/cureus.18123
| Authors et al. (Year) | N or mo. | Type | Site | Symptoms | Approach | N or mo. or yrs. | Clinical Outcome | Remarks |
|-----------------------|----------|------|------|----------|-----------|----------------|-----------------|---------|
| Nakamizo et al. (2001) [6] | 1 3 | Cystic | Third ventricle | Gait disturbance; somnolence | Endoscopy (CSF) | No | 24 | No | Neurolysis of cystic tumor; no clinical meningitis or neurologic deficit |
| Joki et al. (2002) [7] | 1 10 | Cystic | Sella, pons, & metastatic olbenges & | Headache; visual acuity impairment | Endoscopy (CSF) | Yes | 6 | No | Complete cystic decompression; visual acuity and headache improvement |
| Delitala et al. (2004) [8] | 7 9–72 | Cystic; recurrence (43%) | Suprasellar; parasellar; extraventricular (3 Pt); intra + extraventricular (5 Pt); purely intraventricular (2 Pts) | | NR | 14% (pre) | 38 | 28% | No chemical meningitis |
| Nakahara et al. (2004) [3] | 3 46–76 | Cystic | Suprasellar | Headache; dementia; incontinence; hemianopsia | Endoscopy (CSF) | Yes | 7 | 33% | Decompression of the optic chiasm |
| Tirakotai et al. (2004) [9] | 10 NR | Mixed | NR | NR | Endoscopy (CSF) | No | NR | None | No cyst recurrence; 20% re-operated for solid portion |
| Kamikawa & Inui (2005) [10] | 1 4 | Cystic | Suprasellar | Visual disturbance; headache | Endoscopy (CSF) | Yes | NR | NR | No chemical ventriculitis or meningitis |
| Berila et al. (2006) [11] | 1 63 | Cystic; recurrence | NR | Focal neurologcal deficits | Endoscopy (CSF) | No | 6 | No | Exent decrease of cyst size |
| Crippa et al. (2007) [12] | 1 NA | Cystic | NA | NA | Endoscopy (CSF) | No | 12 | No | None |
| Fujimoto et al. (2008) [13] | 1 74 | Mixed | Suprasellar region, extending coronally & obstructing the foramina of Monro bilaterally | Headache; dizziness & gait disturbance | Endoscopy (CSF) | No | 48 | No | Symptoms improved, except for hemianopsia; reduction of cyst size |
| Cappabianca et al. (2008) [14] | 1 3 | Cystic | Intraventricular | Headache; vomiting; bilateral papilledema | Endoscopy (CSF) | No | 36 | No | None |
| Park et al. (2011) [15] | 13 26 | Cystic | NR | NR | Endoscopy | Yes | 32 | 54% | Ommaya reservoir kept in place postoperatively; visual fields stable or improved in 12 Pts (92.3%); preservation of endocrine function |
| Mohanta et al. (2013) [16] | 3 44.6 (18–63) | Solid (2 Pt); cystic (1 Pt) | Intraventricular | Headache; visual disturbance; drowsiness; memory impairment; confusion | Endoscopy (CSF) | Yes | (5 Pt) | 11.6 | No | Weakness & memory impairment improved |
| Takano et al. (2015) [17] | 9 56.7 (38–88) | Cystic | Sellar & suprasellar (3 Pt); Suprasellar (7 Pt) | Raised intracranial pressure; headache; memory disturbance; visual disturbance; hypopituitarism | Endoscopy (CSF) | Yes | 73 | 11% | Tumor size reduction; symptoms improved |
| Shukla (2015) [18] | 3 5–12 | Cystic | Small calcified suprasellar tumor; large cyst extending into the third ventricle | Raised intracranial pressure | Endoscopy (CSF) | Yes | 6–11 | No | Collapse of the cyst; subsidence of hydrocephalus |
| More et al. (2017) [19] | 2 7 | Cystic | Suprasellar | Headache; nausea; vomiting; somnolence; behavioral changes; decreased visual fields | NR | No | 24 | No | None |
| Lauritzen et al. (2018) [20] | 8 43 (32–52) | Cystic, mixed | NR | Raised intracranial pressure; hypothalamic or pituitary dysfunction; visual disturbances | Endoscopy (CSF) | 12.5% (pre) | 56 | 12.5% | None |

**TABLE 1: Summary of the literature reporting the neuroendoscopic transventricular approach in the management of cystic craniopharyngioma**

CSF, cerebrospinal fluid; mo., month(s); N, number of patients; NA, not available; NR, not reported; Pt(s): patient(s); yrs., years
The surgical modality of choice may differ depending on patient- and/or tumor-related factors. In pediatric and adult patients with good functional status, long life expectancy, and small tumor size, microsurgical gross-total resection (GTR) is the recommended strategy [3]. GTR may be considered an acceptable option for tumors not invading the hypotalamus due to the low risk of recurrence and to avoid subsequent radiotherapy. Tumors invading the pituitary stalk, however, are potentially better managed with sub-total resection (STR) due to the high risk of postoperative endocrinopathy [18]. Tumor recurrence, reaching up to 62% (Table 2), is the main concern following microsurgery, endoscopic endonasal, and/or Ommaya reservoir insertion [15,17,20-60]. Alternatively, see Table 2 for the recurrence rate of CC managed with NTVA (without Ommaya reservoir insertion) ranged between 0% and 33% across various studies.

| Author(s) (year) | Surgical Approach | Approach Details | N | Age (yrs.) | Characteristics | Postoperative Visual Function | Follow-up (mo.) | Recurrence |
|------------------|-------------------|------------------|---|------------|----------------|-------------------------------|-----------------|------------|
| Yasargil (1996)[20] | Microsurgery | NA | 162 | NA | NA | NA | NA | NA |
| Fahlbusch et al. (1999)[21] | Microsurgery | Among 148 primary Pts: GTR 73, STR 33, PR 21, Biopsy 4, No resection 17 Among 34 Pts with recurrence: GTR 12, STR 7, PR 15 | 118 – 30; Secondary 20 | Primary (Adults) 148; Primary surgery: 36.8 (1 – 79) | Among 148 primary Pts: Purely cystic 5, Predominantly cystic 73, Predominantly solid 42, Purely solid 16, Multicystic 12; Among 34 Pts with recurrence: Purely cystic 1, Predominantly cystic 15, Predominantly solid 12, Purely solid 2, Multicystic 4 | Primary Surgery: Normalized 38%, Improved 35%, Unchanged 14%, Worsened 12%, Surgery for recurrence: Normalized 12%, Improved 48%, Unchanged 32%, Worsened 8% | 10 |
| Van Effenterre et al. (2002)[24] | Microsurgery | GTR 72; STR 35; PR 15 | 122 | 32.7 (1.5 – 78) | N/A | Of 76 Pts followed: Normalized 34%, Improved 48%, Unchanged 3%, Worsened 14% | Mean 84 | 24 |
| Im et al. (2003)[22] | Microsurgery | GTR | 6 | 10.6 | Cystic | All improved | Mean 23 | 16 |
| Karavitaki et al. (2005)[23] | Microsurgery | GTR 16 (Group A), GTR + Radiotherapy 3 (Group B), PR 51 (Group C), PR + Radiotherapy 33 (Group D), Cyst evacuation 6 (Group E), Cyst evacuation + Radiotherapy 3 (Group F) | 121 | Age <16: 42, Age ≥16: 79, Total: 2.5 – 83 | Purely or predominantly cystic 42/91, Mixed 33/91, Purely or predominantly solid 16/91 | Worsening of visual fields at 10 yrs.: Group A: 9%, Group B: 0%, Group C: 45%, Group D: 24% | Mean 103 | GTR 0%, PR 62 |
| Filis et al. (2009)[31] | Microscopic-endoscopic | GTR | 1 | 7 | Cystic | NR | 24 | No |
| Schubert et al. (2009)[38] | Microsurgery | GTR 6, SR 11 | 17 | ≤17 | Cystic | NR | 66 | 58 |
| Ichikawa et al. (2016)[34] | Microscopic-endoscopic | GTR | 4 | 6.4 | Cystic | Improved 2, Unchanged 2 | Mean 142 | 25 |
| Feng et al. (2018)[30] | Microsurgery | GTR 124, STR 37, PR 13 | 183 | 36.2 (3 – 77) | NR | Improved 54, Worsened 22 | Mean 27.3 | 12 |
| Shibata et al. (2018)[33] | Microsurgery + Endoscopic endonasal | GTR | 1 | 1 | Cystic | Improved (light perception) | Mean 18 | No |
| Abe et al. (1997)[25] | Endoscopic endonasal | GTR 15 STR 19 | 35 | 27 (8 – 72) | Cystic | Improved 18 Unchanged NR Worsened NR | Mean 24.1 | 8.6 |
| Buhl et al. (2001)[26] | Endoscopic endonasal | NR | 1 | 4 | Cystic | Improved | Mean 12 | No |
| Fujimoto et al. (2002)[32] | Endoscopic endonasal + Radiation therapy | GTR | 1 | 8 | Cystic | Improved | Mean 30 | No |
| Author(s) | Year | Endoscopic endonasal |Procedure | Patients | Follow-up | Results | Mean | % |
|----------|------|----------------------|----------|----------|-----------|---------|------|---|
| Locatelli et al. (2004) | 36 | GTR, Endoscopic endonasal | 1, Draining cyst contents to sphenoid sinus, 3, Multiphase approach | 5 | 2 – 16 | Cystic | NR | 48 | 20% |
| Chakrabarti et al. (2005) | 28 | GTR, Endoscopic endonasal | 61, PR 7 | 68 | 2.5 – 73 | Cystic & solid | Improved 54, Unchanged 12, Worsened 2 | Mean ≥ 66 | 10% |
| Rudnick & Dinardo (2006) | 37 | GTR, Endoscopic endonasal | - | 1 | 31 | Cystic | NR | 28 | No |
| Gardner et al. (2008) | 33 | GTR, Endoscopic endonasal | 16 | 55 (36 – 80) | NR | Improved 13, Unchanged 1, Worsened 0 | Mean 34 | NR |
| Stamm et al. (2008) | 40 | GTR, Endoscopic endonasal | 7 | 23.4 | NR | Improved 2, Unchanged 1, Worsened 0 | Mean 36.2 | No |
| Fatemi et al. (2009) | 29 | GTR, Endoscopic endonasal | 18 | 40 (18 – 62) | NR | Improved 11, Unchanged 1, Worsened 0, No impairment preop/postop 6 | Mean 20 | 0% (among 16 Pts) |
| Campbell et al. (2010) | 27 | GTR, Endoscopic endonasal | 14 | 45 (18 – 85) | NR | Improved 12, Unchanged 1, Worsened 1 | NR | NR |
| Jane et al. (2010) | 35 | GTR, Endoscopic endonasal | 12 | 50.77 (29 – 76) | Mixed 8, Cystic 3, Solid 1 | Improved 7, Unchanged 5, Worsened 0 | Mean 13.3 | NR |
| Coppers & Coudwell (2010) | 44 | GTR, Endoscopic endonasal | 1 | 26 | NR | Improved | 18 | No |
| Garcia-Navarro et al. (2011) | 48 | GTR, Endoscopic endonasal | 2 | NR | NR | NR | NR | NR |
| Lang et al. (2012) | 49 | GTR, Endoscopic endonasal | 24 | 43.6 (5 – 82) | Cystic 10, Solid & cystic 14 | Improved 10, Unchanged 3, Worsened 1 | Mean 32.9 | 26% |
| Koutroubis et al. (2013) | 48 | GTR, Endoscopic endonasal | 64 (Primary 47 – Recurrent 17) | 40 (4 – 82) | NR | Improved 38, Unchanged 5, Worsened 1 | Mean 38 | 34% |
| Cavallo et al. (2014) | 43 | GTR, Endoscopic endonasal | 103 | 42.5 (3 – 83) | NR | Improved 63, Unchanged 14, Worsened 2 | Mean 48 | 22% |
| Ou et al. (2015) | 47 | GTR, Endoscopic endonasal | 3 | 36.3 | Solid | Improved 3, Unchanged 0, Worsened 0 | Mean 35.6 | 33% |
| Prabhu et al. (2015) | 50 | GTR, Endoscopic endonasal | 1 | 79 | Cystic | Improved | NR | NR |
| Abou-Al-Shaar et al. (2016) | 41 | GTR, Endoscopic endonasal | 1 | 22 | Cystic & solid | NR | 12 | No |
| Bai et al. (2016) | 42 | GTR, Endoscopic endonasal | 25 (Primary 15 – Recurrent 10) | 5 – 68 | NR | Improved 15, Unchanged 10, Worsened 0 | Mean 54.7 | NR |
| Fomichev et al. (2016) | 45 | GTR, Endoscopic endonasal | 136 | 49.3 (13-73) | NR | Improved + Unchanged 121, Worsened 15 | Mean 42 | 20% |
| Mangiussi-Gomes et al. (2018) | 54 | GTR, Endoscopic endonasal | 1 | 72 | Cystic & solid | NR | NR | NR |
| Locatelli et al. (2021) | Cureus 13(9): e18123. DOI 10.7759/cureus.18123 | GTR, Endoscopic endonasal | | | | | | |
### TABLE 2: Summary of the literature reporting microsurgical, endoscopic endonasal, and Ommaya reservoir procedures for patients with cystic craniopharyngioma

| Authors                          | Procedure                      | GTR | STR | PR | Cystic/Non-Cystic | Improvement         | Mean | No. |
|----------------------------------|--------------------------------|-----|-----|----|-------------------|---------------------|------|-----|
| Vitz et al. (2001)               | Ommaya reservoir               |     |     |    |                   | Improved            | NR   |     |
| Nicolato et al. (2004)           | Ommaya reservoir + bleomycin   | GTR | STR | PR |                   |                     | NR   |     |
| Park et al. (2011)               | Ommaya reservoir endoscopy     | STR |     |    |                   | Improved            | NR   |     |
| Moussa et al. (2013)             | Ommaya reservoir + Stereotaxy  |     |     |    |                   |                     | NR   |     |
| Rahmathulla & Barnett (2013)     | Ommaya reservoir + Stereotaxy  | STR |     |    |                   | Improved + Unchanged | NR   |     |
| Srikandarajah et al. (2014)      | Ommaya reservoir               |     |     |    |                   |                     | NR   |     |
| Takano et al. (2015)             | Ommaya reservoir endoscopy     |     |     |    |                   | Improved            | NR   |     |
| Vakharia et al. (2017)           | Ommaya reservoir               |     |     |    |                   |                     | NR   |     |
| Zhu et al. (2017)                | Ommaya reservoir + Microsurgery| GTR | STR |    |                   | Improved + Unchanged | NR   |     |
| Laurol et al. (2018)             | Ommaya reservoir endoscopy     |     |     |    |                   | Improved            | NR   |     |
| Frio et al. (2019)               | Ommaya reservoir endoscopy     |     |     |    |                   | Improved + Unchanged | NR   |     |

GTR, gross-total resection; mo., month(s); N, number of patients; NA, not available; NR, not reported; NTR, near-total resection; PR, partial resection; Pt(s), patient(s); STR, sub-total resection; yrs., years.

The NTVA is recognized as a safe, efficacious, and minimally invasive procedure for intra- and paraventricular craniopharyngiomas, especially for cystic, large, and extensive lesions [2-3]. Our literature review results, as well as our experience exemplified by the case report described above, further support this notion (Table 1, Table 3). Intraoperative fenestration of the cyst wall usually drains the dense liquid content, typically described as 'engine oil' in color and texture [2]; spillage of the cyst contents into spaces containing cerebrospinal fluid (CSF) may lead to chemical meningitis and possibly secondary hydrocephalus [32]. Some authors advocate inserting a catheter with an Ommaya reservoir to allow for intermittent drainage, combined with radiation therapy [51]; long-term catheter placement, however, is associated with risks of infection, catheter displacement, content re-accumulation due to cyst septations, and pain with Ommaya reservoir tapping. Remarkably, none of the NTVA studies summarized in Table 1 reported the occurrence of chemical meningitis or delayed hydrocephalus secondary to the communication of CSF spaces with the cyst.
The low complication and recurrence rates (Table 1 and Table 3) following NTVA, as well as the endoscopic endonasal approach, compared to microsurgery and Ommaya reservoir insertion, support the validity of these minimally invasive techniques as credible and potentially better alternatives in the management of CC or cystic-dominant craniopharyngiomas. This conclusion is limited by the small number of reported NTVA cases and the heterogeneity of the published studies.

Conclusions

The NTVA is efficient, reliable, and safe for managing CC and cystic-dominant craniopharyngiomas, with potentially lower recurrence and complication rates compared to microsurgery and Ommaya reservoir insertion. Future randomized clinical studies comparing the various treatment modalities of CC are needed to solidify these conclusions.

Additional Information

Disclosures

**Human subjects:** Consent was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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