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Original Research Article

A Hospital-Based Study to Assess the Endoscopic Transcortical Transventricular Management of Cystic Craniopharyngioma

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Abstract

Aim: The aim of the present study was to assess the endoscopic transcortical transventricular management of cystic craniopharyngioma.

Methods: The present study was conducted at IGIMS, Patna, India and data of patients who underwent transcortical transventricular endoscopic approach and reservoir placement for cystic craniopharyngioma were reviewed retrospectively for the period of 2 years. 50 patients were included in the study.

Results: The mean age was 22.6 years (range 2–49 years) with a slight female preponderance. Headache (74%) was the most common presentation followed by vomiting (56%) and vision disturbances (46%). An equal number of patients presented with motor weakness (3.4%) and memory disturbances (12%). 3 patients presented with altered behavior and 2 patients with polyuria. Most of the patients had a single cyst. All patients had hydrocephalus. The presurgery hormone evaluation showed that 10% of patients had hypocortisolemia and 6% of patients had marginally increased prolactin levels. The follow-up duration ranged from 3 to 72 months with a median of 18 months. 19 (38%) patients had improvement in vision. 10 (20%) patients required thyroid hormone replacement and 9 patients (18%) steroid replacement. 21 (42%) patients received radiotherapy. 14 (28%) cases had a cyst recurrence and required repeat intervention.

Conclusion: Transventricular endoscopy is a versatile procedure that plays an important part in the management of craniopharyngioma, especially for those lesions with a predominant cystic component growing within the third ventricle. It has been used to fenestrate cystic tumors into the ventricles or subarachnoid space (ventriculocystostomy or cystocisternostomy), to establish CSF flow in case of hydrocephalus, for delivery of intracavitary chemotherapy, and to achieve variable extent of resection as a first step of combined procedure.

Keywords: endoscopy, Neuroendoscopy, hydrocephalus, craniopharyngioma, reservoir

Introduction

Craniopharyngiomas are benign sellar and suprasellar tumors, deemed to originate from the cellular residue of Rathke's pouch. [1,2] Half of all cases occur in adulthood with a peak incidence at the age of 40–44 years and another peak at the sixth decade. They have a variable worldwide incidence ranging from 2% to 5% of all primary intracranial tumors. [2,3] The malignant course is noted in long-term management due to compression of the pituitary gland and stalk, the tendency to repeated cystic recurrence with the potential invasion of the hypothalamus and third ventricle, which further complicates the clinical status due to hydrocephalus. Hydrocephalus is present in 20–30% of adult craniopharyngioma at the time of presentation. [4-6]

Cystic craniopharyngiomas present a unique challenge because of the possibility of recurrence of cyst irrespective of the modality of initial treatment. [7] Almost 90% of craniopharyngiomas have a cystic component. [8] The presence of a cyst in craniopharyngioma can account for major total bulk of the tumor, and a predominantly cystic tumor is seen in 60% of craniopharyngiomas. [9] The traditional approach to cystic craniopharyngioma is a variable extent of microsurgical resection.

Ommaya reservoir is often placed in the residual cyst for repeated aspiration or intracavitary therapy. [10] However, the malposition of reservoir is not uncommon. [11,12]

Microscopic surgical resection and endoscopic cyst fenestration along with intracystic catheter placement of the Ommaya reservoir are the commonly performed surgical techniques. [13,14] Transcortical transventricular endoscopic surgery for cystic craniopharyngioma has gained acceptance as a safe minimally invasive alternative to resection. [15,16] The traditional approach to cystic craniopharyngioma is a variable extent of microsurgical resection. Ommaya reservoir is often placed in the residual cyst for repeated aspiration or intracavitary therapy. [10] However, malposition of the reservoir is not uncommon. [11,12] In such circumstances a neuroendoscopic approach provides a less invasive and more accurate technique to deal with cystic craniopharyngiomas. [11,17]

The aim of the present study was to assess the endoscopic transcortical transventricular management of cystic craniopharyngioma.

Materials and Methods

The present study was conducted at IGIMS, Patna, India and data of patients who underwent transcortical transventricular endoscopic approach and reservoir placement for cystic craniopharyngioma were reviewed retrospectively for the period of 2 years. 50 patients were included in the study. The patients were identified and none of the patients' personal information is included in the article so that none of these patients can be identified or contacted, considering this, retrospective studies are waived off from review by our institute's ethics committee. The clinical features, imaging findings, surgical steps, complications, and outcomes were recorded. All patients underwent a neurological and neuro-ophthalmological examination before surgery and at follow- up. Basal serum levels of anterior pituitary hormones were assessed for all patients before surgery and at follow-up. All patients underwent preoperative magnetic resonance imaging of the brain, and computerized tomography scan of the head immediately after surgery and at follow-up.

Technique

The technique of transcortical transventricular endoscopic approach and Ommaya reservoir placement for cystic craniopharyngioma was similar to any intraventricular cystic tumors.11

The patient was positioned supine with the head in neutral position. A semilunar scalp flap based on coronal suture was raised. A precoronal burr hole, 3 cm from the midline, was made. The ventricles were entered with trocar and endoscope sheath. Gaab 0° telescope (Karl Storz, Tut- tlingen, Germany), was introduced in the ventricle and the cyst was identified at the foramen of Monro. The 0° scope was removed and the operating scope was introduced.

The cyst wall was coagulated, and a piece of wall was taken for biopsy. On puncturing the cyst olive green (machine oil) colored fluid, typical of craniopharyngioma was aspirated. Continuous irrigation with suction was done to remove all the fluid, till the contents of the cyst became clear. The inner wall of the cyst was inspected and a biopsy was taken from the wall of the cyst and solid portion. No attempt was made to resect the solid portion of the tumor. A further fenestra- tion at the deeper portion of the cyst for communication to the basal cistern was attempted only if the cyst wall was transparent and the basilar artery could be easily visualized. Neuronavigation was used for multiloculated cysts to ensure communication between cysts. At the end of irrigation, a ventricular catheter was passed through the operating channel of scope under direct vision till it touched the deepest wall of the cyst. There was good three-dimensional visualization and orientation. The operating scope along with the sheath was slowly withdrawn under vision leaving behind the ventricular catheter. The catheter was connected to the reservoir in the subgaleal space. The reservoir was punctured to aspirate fluid and confirm patency. The histopathology of the cyst wall confirmed the diagnosis of craniopharyngioma in all cases. All patients received perioperative antibiotics, steroids, and antiepileptic drugs. Adults and children over 7 years of age were referred for radiotherapy.

Results

Table 1: Demographic details

Parameter (*N=50*)	
Age	Mean 22.6 y (range 1.8–49)
Gender	Male 23 (46%)
	Female 27 (54%)
Clinical features	
Headache	37 (74%)
Vision disturbances	23 (46%)
Polyuria	2 (4%)
Vomiting	28 (56%)
Motor weakness	6 (12%)

Memory disturbances	6 (12%)
Altered behavior	3 (6%)
Radiology	
Single cyst	38 (76%)
Multiloculated	12 (24%)
Endocrine disturbances	
Hypocortisolism	5 (10%)
Marginally increased prolactin (stalk effect)	3 (6%)

The mean age was 22.6 years (range 2–49 years) with a slight female preponderance. Headache (74%) was the most common presentation followed by vomiting (56%) and vision disturbances (46%). An equal number of patients presented with motor weakness (3.4%) and memory disturbances (12%). 3 patients presented with altered behavior and 2 patients with polyuria. Most of the patients had a single cyst. All patients had hydrocephalus. The presurgery hormone evaluation showed that 10% of patients had hypocortisolemia and 6% of patients had marginally increased prolactin levels.

Table 2: Surgical details and follow-up data

Parameter (*n* =50)	
Surgery duration	Median 90 min
Hospital stay	Mean 3.4 d
Follow-up (*n* ¼ 29)	
Duration	Median 18 mo (3–72 mo)
Cyst recurrence	14 (28%)
Reaspiration	9 (18%)
Craniotomy	4 (8%)
Shunt	6 (3%)
Median recurrence-free survival	43 mo
Vision improved	19 (38%)
Radiotherapy	21 (42%)
Hormone replacement	
Thyroid	10 (20%)
Steroid	9 (18%)
Desmopressin	2 (4%)

The follow-up duration ranged from 3 to 72 months with a median of 18 months. 19 (38%) patients had improvement in vision. 10 (20%) patients required thyroid hormone replacement and 9 patients (18%) steroid replacement. 21 (42%) patients received radiotherapy. 14 (28%) cases had a cyst recurrence and required repeat intervention.

Discussion

Surgery for craniopharyngioma has a major negative impact on long-term quality of life (QoL). In a recent literature review on quality-adjusted life years (QALYs) of four surgical approaches to craniopharyngiomas: aggressive tumor removal (attempt at gross total resection [GTR]), planned subtotal removal plus radiotherapy, biopsy plus radiotherapy, and transnasal endoscopic resections were reviewed. [18] Cystic craniopharyngiomas present a unique challenge because of the possibility of recurrence of cyst irrespective of the modality of initial treatment. Almost 90% of craniopharyngiomas have a cystic component. [19]

Endoscopy has some advantages over other drainage techniques. Through a minimally invasive approach it is possible to perforate the capsule under direct visual control as compared with repeated attempts of perforation and the risk of deviation from the tract due to the consistency of the cystic wall in stereotactic procedures. Endoscopy offers more radical cyst drainage, partial resection of the capsule and sampling of tissue fragments under direct vision for diagnostic purposes avoiding the risks of closed biopsy, and wide marsupialization into cerebrospinal fluid (CSF) fluid. The most common procedure performed through the transventricular endoscopic approach for craniopharyngiomas is fenestration and aspiration of the cyst with catheter placement inside the cyst connected to a subgaleal reservoir. Instead of placement of reservoir Delitala et al [9] positioned a transcystic multiholed catheter with ends reaching into the cistern (caudally) and lateral ventricle (rostrally). In such stenting, ventricle acts as reservoir in which the cyst fluid collects and gets absorbed along with CSF. This stenting was devised in an attempt to promote continuous CSF circulation and prevent the cyst's reclosure. The mean age was 22.6 years (range 2–49 years) with a slight female preponderance. Headache (74%) was the most

common presentation followed by vomiting (56%) and vision disturbances (46%). An equal number of patients presented with motor weakness (3.4%) and memory disturbances (12%). 3 patients presented with altered behavior and 2 patients with polyuria. Most of the patients had a single cyst. All patients had hydrocephalus.

Nakamizo et al[16] had an early single case report of trans ventricular endoscopic craniopharyngioma drainage and Ommaya catheter and reservoir placement. Nakahara et al [13] reported three adult cases of trans-ventricular endoscopic management for craniopharyngioma with mixed solid/cystic components where they could evacuate the cyst contents and obtain a biopsy. The presurgery hormone evaluation showed that 10% of patients had hypocortisolemia and 6% of patients had marginally increased prolactin levels. The follow-up duration ranged from 3 to 72 months with a median of 18 months. 19 (38%) patients had improvement in vision. 10 (20%) patients required thyroid hormone replacement and 9 patients (18%) steroid replacement. 21 (42%) patients received radiotherapy. 14 (28%) cases had a cyst recurrence and required repeat intervention.

Previously, complex cases of cystic craniopharyngiomas that invade the third ventricle and induce obstructive hydrocephalus were treated at two steps. Initially, a transcranial trans-ventricular endoscopic fenestration of the cyst wall with placing a catheter into the cyst to release the increased intracranial pressure. This is followed by another elective intervention within the next days by an endonasal trans-sphenoidal procedure for excision of the tumour. [20] Furthermore, Frio et al[21] have recommended the sole drainage of the cyst to relieve symptoms caused by mass effect as a valid alternative option to limit surgery related morbidity, especially in recurrent cases. [21] They reported 72.7% long-term local tumour control with the improvement of visual function and intracranial hypertension. Craniopharyngiomas can produce a wide range of manifestations such as headache, nausea and vomiting, dizziness, as well as visual and endocrine deficits. [22] Preoperative evaluation of the studied patients revealed headache (100%), dizziness (72.7%), visual field changes (68.2%), nausea and vomiting (40.9%), hormonal changes (37.8%), and memory disturbances (31.8%). Correspondingly, Almelesy [23] has recently reported visual disturbance (83.3%), endocrinopathy (33.3%), and headache (75%) in adult craniopharyngioma patients recruited from Al-Azhar University Hospitals, Egypt. Park et al [24] described vision loss as the most common presenting symptom (83%), followed by headache (48%) and endocrinopathy (33%) in a multicentre study. Further, Cavallo et al [25] found visual disturbances (76.7%) and endocrinopathy (56.6%) in affected adult patients.

Conclusion

Transventricular endoscopy is a versatile procedure that plays an important part in the management of craniopharyngioma, especially for those lesions with a predominant cystic component growing within the third ventricle. It has been used to fenestrate cystic tumors into the ventricles or subarachnoid space (ventriculocystostomy or cystocisternostomy), to establish CSF flow in case of hydrocephalus, for delivery of intracavitary chemotherapy, and to achieve variable extent of resection as a first step of combined procedure. Among all the endoscopic treatment options the preferred treatment is fenestration, aspiration of cyst, and placement of catheter reservoir followed by radiation. The transventricular endoscopic approach is not shown to be particularly effective for improvement of vision in patients with complete vision loss before surgery. Residual tumor with persistent symptoms would require resection for relief of symptoms of neural compression.

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