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Original Article

Endoscopic Transcortical Transventricular Management of Cystic Craniopharyngioma: Outcome Analysis of 32 Cases at a Tertiary Care Center

Somil Jaiswal, Manish Jaiswal, Pooja Jaiswal[1], Ankur Bajaj, Chhitij Srivastava, Anil Chandra, Bal Krishna Ojha, Janu Vikas, Awadhesh Yadav

Department of Neurosurgery, King George's Medical University, [1]Department of Pathology, Integral Institute of Medical Sciences and Research, Lucknow, Uttar Pradesh, India

Abstract

Background: Microsurgical resection has been considered the gold standard treatment of craniopharyngioma, but lately, it has found less favor due to its morbidity and is being replaced by minimally invasive cyst drainage procedures. We present our experience of transventricular endoscopy and cyst drainage along with its technique and have analyzed its results. **Materials and Methods:** Clinical and radiological data of all cystic craniopharyngioma patients treated by transventricular endoscopic cyst drainage and Ommaya placement were retrieved and analyzed **Results:** Thirty-two patients underwent endoscopic cyst drainage during the study period. All patients had immediate clinical and radiological improvement. No significant complications were seen. All patients underwent adjuvant radiotherapy and six patients (18.7%) showed recurrence. Three patients died in the follow-up period. **Conclusions:** Endoscopic transcortical transventricular cyst drainage with Ommaya reservoir along with adjuvant radiotherapy is a simple, safe, and effective treatment modality.

Keywords: *Cystic craniopharyngioma, cystocisternostomy, endoscopic transcortical transventricular, Ommaya, radiotherapy*

Introduction

Craniopharyngiomas are benign neoplasm with malignant behavior and account for nearly 3% of all intracranial tumors.[1-5] About 60%–90% of all craniopharyngiomas have a cystic component and propensity to extend into the anterior third ventricle, interpeduncular cistern, and retro and parasellar region.[1,3,6] Gross total resection of this tumor is the gold standard surgical treatment, but the preservation of neurological and hypothalamopituitary functions is also an important facet in the management of this complex tumor.[5]

Microscopic surgical resection and endoscopic cyst fenestration along with intracystic catheter placement of the Ommaya reservoir are the commonly performed surgical techniques.[1,7,8,9,10] Transcortical transventricular endoscopic surgery for cystic craniopharyngioma has gained acceptance as a safe minimally invasive alternative to resection.[11,12]

In this retrospective study, an outcome analysis of transcortical transventricular endoscopically treated cystic craniopharyngioma over a period of 9 years was performed.

Materials and Methods

Records of 95 craniopharyngioma patients were retrieved from the departmental database, who were operated between January 2009 and October 2017 in the Department of Neurosurgery at King George's Medical University, Lucknow, India.

Thirty-two out of 95 patients had undergone endoscopic transcortical transventricular decompression and drainage along with cerebrospinal fluid (CSF) diversionary procedure, were included in this retrospective study. Ethical approval for conducting this study was taken from the institutional ethics committee.

Demographic details, clinical presentations, radiological features, intraoperative findings, details of CSF diversionary procedure, surgical complications, and follow-up details were noted.

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Address for correspondence:
Dr. Somil Jaiswal, Associate Professor, Department of Neurosurgery, King George's Medical University, Lucknow - 226 003, Uttar Pradesh, India.
E-mail: dr.somil26@gmail.com

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Surgical technique

All cases in this study had undergone neuroendoscopy by Karl Storz Lotta system 6° ventriculoscope of diameter 6.1 mm with a working channel diameter of 2.9 mm (KARL STORZ SE and Co. Tuttlingen/Germany). Patients were positioned supine and a linear incision was given at coronal suture. A small precoronal burr hole 3 cm from the midline and 1 cm anterior to coronal suture was made. Dura was opened either in a linear fashion or a C-shaped flap. The lateral ventricle was entered with obturator and working sheath. A freehand technique was used in all cases. The endoscope was introduced inside ventricle and cystic craniopharyngioma was visualized at Foramen Monro. The cyst wall was coagulated, fenestrated, and biopsy was taken from the wall. Dark green or machine oil colored fluid was aspirated from the cyst. Irrigation and suction of the cyst was done and inside of the cyst was visualized by the endoscope. Multiple calcified spots along with friable, suckable contents were usually seen and taken out. In five cases, floor of the third ventricle and interpeduncular cistern were well visualized though the cyst for performing cystocisternostomy, this procedure was termed as double fenestration (ventriculocystostomy and cystocisternostomy) [Figure 1]. Patients with significant hydrocephalus would also undergo endoscopic septostomy, followed by a ventriculoperitoneal shunt performed through Keen's point in the same sitting. A catheter was positioned in the cyst cavity under direct vision through the scope and was connected to the Ommaya reservoir placed at burr hole [Figure 2]. The skin was closed in two layers.

Postoperative management

The patients were administered antibiotics according to our institute protocol. Postoperative computed tomography (CT) scan was done to rule out any bleed and confirm the placement of Ommaya catheter. Anti-epileptics (phenytoin 5 mg/kg in divided doses) was given for 6 months if the patient remained seizure-free. In four patients, we witnessed seizures in the postoperative period for which the dosage of anti-epileptic drug was increased and no further episodes of seizures were reported.

Results

The age of the patients ranged from 3 years to 68 years (mean 16.28 ± 15.70). Seventy-five percent of patients were below 16 years of age. There were 23 male and 9 female patients. Headache was the most common presenting complaint, followed by the visual disturbance. Presenting symptoms are enumerated in Table 1.

Investigations

CT and magnetic resonance imaging scans were performed in all patients preoperatively. Fifty percent of the patients had hydrocephalus. All the patients had predominantly cystic craniopharyngioma with cyst diameter ranged from 3 cm to 8 cm (mean 4.5 cm). Calcification was seen in the wall of cyst in 70% of cases [Table 2].

Table 1: Presenting symptoms of the patients (*n*=32).

Symptoms	Number of patients (%)
Headache	27 (84)
Nausea/vomiting	24 (75)
Confusion	6 (19)
Visual disturbance	25 (78)
Hormonal deficiency	7 (22)

Ophthalmological investigations showed papilledema in 53% (17/32) of patients which improved significantly after surgical intervention, whereas 40% (13/32) of patients had primary optic atrophy whose visual acuity status remained nearly same as preoperative or had only slight improvement.

Outcome

All 32 patients were treated by endoscopic fenestration of craniopharyngioma cyst along with a biopsy of cyst wall and complete drainage of cyst fluid. Ommaya reservoir was placed in all cases [Figure 4]. Twelve patients required a ventriculoperitoneal shunt and six patients among them also underwent septostomy who had significant hydrocephalus. In five patients, we were able to perform cystocisternostomy (double fenestration) [Table 2 and Figure 3].

Headache improved in 92% (25/27) patients in whom headache was presenting symptom. Nausea, vomiting, and confusion improved in all patients.

External beam radiotherapy was given to all the patients. No patient developed any visual deterioration following radiotherapy. However, six patients showed clinical and radiological recurrence in the follow-up with requirement of multiple Ommaya aspirations, they were advised microsurgical intervention, but out of them, only two underwent transcranial surgical excision. The recurrence rate in the study was 18.75% (6/32). Remaining 26 patients (81%) had good tumor control on radiology, required no Ommaya aspirations and were doing well at follow-ups.

Complications

Burr-hole site skin infection was seen in one case which was treated successfully with antibiotics. Bacterial meningitis was seen in two cases (Case 1-Patient No. 9-both VP shunt and Ommaya reservoir with catheter were removed and intravenous antibiotics were given according to culture reports for 14 days, reinsertion of the shunt was done from a fresh site after CSF culture reports turned sterile). (Case 2-Patient No. 11-Ommaya reservoir with catheter was removed on the basis of positive culture reports and was treated with intravenous antibiotics). One case each of intraventricular hemorrhage and subdural

Table 2: Clinicoradiological profile and outcome data of the study patients

Patient number	Age/sex	Symptom	Cyst maximum diameter on MRI (cm)	Endoscopic cyst fenestration with Ommaya placement	Cystocisternostomy	VP shunt	Complications	Radiotherapy	Recurrence	Follow-up (months)	Expiry
1	8 years/male	HA NV VD	6	Yes		Yes		Yes	Yes	36	Lost in follow-up
2	10 years/male	HA NV VD	4	Yes			Burr hole site skin infection	Yes		18	
3	65 years/male	HA VD	5	Yes		Yes		Yes		15	
4	9 years/female	HA NV VD	4	Yes				Yes		20	
5	9 years/female	C	6	Yes		Yes		Yes	Yes	17	Lost in follow-up
6	16 years/male	HA NV VD HD	5	Yes		Yes	Seizure	Yes	Yes	22	Resurgery
7	10 years/male	HA NV HD	5	Yes				Yes		12	
8	5 years/female	HA NV VD C	4	Yes	Yes			Yes		3	Expired
9	6 years/female	HA NV VD	4	Yes		Yes	Shunt infection, meningitis, seizure	Yes		6	
10	68 years/male	C	6	Yes		Yes		Yes		12	
11	25 years/male	HA NV VD	4	Yes			Meningitis	Yes		18	
12	18 years/female	HA NV VD HD	5	Yes		Yes		Yes	Yes	25	Lost in follow-up

Contd...

Table 2: Contd...

Patient number	Age/sex	Symptom	Cyst maximum diameter on MRI (cm)	Endoscopic cyst fenestration with Ommaya placement	Cystocisternostomy	VP shunt	Complications	Radiotherapy	Recurrence	Follow-up (months)	Expiry
13	10 years/male	HA NV VD	6	Yes	Yes			Yes		12	
14	35 years/male	HA VD	5	Yes				Yes		15	
15	12 years/male	HA NV VD	4	Yes			Seizure	Yes		48	
16	15 years/male	HA NV VD HD	6	Yes		Yes		yes	Yes	20	Resurgery
17	10 years/male	HA NV VD	4	Yes				Yes		6	Expired
18	7 years/male	HA NV VD HD	4	Yes	Yes			Yes		9	
19	8 years/female	C	5	Yes				Yes		32	
20	10 years/ female	HA NV VD	4	Yes			Burr hole site EDH	Yes		76	
21	45 years/maleh	HA VD	5	Yes		Yes	Shunt malfunction, CSF leak	Yes		50	
22	11 years/ female	HA NV VD	6	Yes	Yes			Yes		24	
23	7 years/male	C	8	Yes		Yes		Yes	Yes	46	Lost in follow-up
24	16 years/male	HA NV VD HD	5	Yes			Seizure	Yes		32	

Contd...

Table 2: Contd...

Patient number	Age/sex	Symptom	Cyst maximum diameter on MRI (cm)	Endoscopic cyst fenestration with Ommaya placement	Cystocisternostomy	VP shunt	Complications	Radiotherapy	Recurrence	Follow-up (months)	Expiry
25	3 years/male	HA NV	3	Yes				Yes		11	
26	8 years/male	HA NV VD	5	Yes	Yes			Yes		15	
27	10 years/male	HA NV VD HD	5	Yes		Yes	Shunt malfunction, CSF leak	Yes		33	
28	16 years/male	C	5	Yes			IVH	Yes		10	
29	7 years/female	HA NV VD HD	5	Yes				Yes		6	Expired
30	8 years/male	HA NV VD	5	Yes				Yes		17	
31	15 years/male	HA NV VD	4	Yes		Yes	Subdural collection	Yes		25	
32	19 years/male	HA NV VD	4	Yes				Yes		36	

HA – Headache; NV – Nausea/vomiting; VD – Visual deficit; C – Confusion; HD – Hormonal deficiency; IVH – Intraventricular hemorrhage; EDH – Extradural hemorrhage; CSF – Cerebrospinal fluid

collection was managed on conservative lines and fared well. Burr hole site extradural hemorrhage was seen in 1 case and was evacuated. CSF leak seen in two cases was due to shunt malfunction, which subsided on shunt revision. Shunt complications were treated by reinserting fresh shunts from the changed side [Table 3].

Mortality

Three patients expired in the follow-up. The exact cause of death could not be ascertained but telephonic conversation linked them to seizures due to poor compliance with anti-epileptics.

Follow-up

The patients were regularly followed up clinically and radiographically. CT scan was initially done at 3 months thereafter at 6 months and then at yearly interval. The mean follow-up was 22.7 ± 15.8 months (3–76 months) [Table 2].

Discussion

Craniopharyngiomas are commonly seen in two age groups (5–15 years) and (60–70 years).[1,5,13,14] Similar observation was seen in our series. Majority of the patients were of pediatric age group.[1,5,13,14] The mean age of the patients in this study was 16.28 ± 15.70 which is close to the reported mean age in other series.[1,5,13,14] The clinical profile of patients in this series was similar to previously reported other series. Features of raised intracranial pressure and visual disturbance were commonly seen.[5,10,13,14]

Table 3: Complications encountered in the study patients

Complications	Number of patients
Burr hole site skin infection	1
Meningitis	2
Intraventricular hemorrhage	1
Subdural hygroma or collection	1
Shunt malfunction	2
Shunt infection	1
CSF leak	2
Burr hole site EDH	1
Seizures	4

EDH – Extradural hemorrhage; CSF – Cerebrospinal fluid

Gross total removal of cystic craniopharyngioma is considered as the gold standard treatment, but this procedure is plagued with significant morbidity and mortality following damage to the hypothalamus, optic apparatus, and stalk.[1,5] To avoid the torrid postoperative course, surgical resection of cystic craniopharyngioma has largely been replaced by cyst fenestration and drainage, and it has become one of the commonly performed procedures for predominantly cystic craniopharyngioma.[11,12,15] Over the years, cyst fenestration along with Ommaya placement has evolved as a less invasive and more accurate surgical procedure, while it was initially performed percutaneously, or ultrasound-guided, but lately with endoscopy, it has led to better visualization of tumor cavity, accurate placement of catheter, as well as minimal damage to the hypothalamus or optic apparatus.[3,4,9,6,16,14,17] Rachinger *et al*. in their series concluded that cystic fenestration is as effective as microsurgery with less hypothalamic damage.[18] The present study further reiterates the above fact.

Lauretti *et al*. analyzed and stated the superiority of neuroendoscopy over stereotactic cyst aspiration as well as cyst CSF communication as a protective factor for recurrence.[19] In our series, patients had a collapse of cyst after cyst fenestration and widening of the cyst opening along with aspiration. Ommaya catheter holes had communication with both craniopharyngioma cyst and ventricle CSF. Patency of cyst and ventricle communication due to widening of fenestration and catheter holes both in cyst and ventricle is the probable reason which is believed to be the cause of no recollection in these patients. The same explanation was mooted by Al-Abyad and El-Sheikh and Moussa *et al*. in their series on cystic craniopharyngioma.[13,19,20,21] In some series, chemical

Table 4: Summary of series describing endoscopic cyst drainage with Ommaya shunt placement

Study	Type of study	Number of patients	Radiotherapy	Follow-up (months)	Outcome	Recurrence
Hellwig *et al*., 1995	Retrospective	5	No	NA	Improved	No
Nokamizo *et al*., 2001	Retrospective	1	No	24	Improved	No
Joki *et al*., 2002	Retrospective	1	Yes	6	Improved	No
Delitala *et al*., 2004	Retrospective	7	Yes	38	Improved	28%
Nakahara *et al*., 2004	Retrospective	3	Yes	7	Improved	33%
Tirakotai *et al*., 2004	Retrospective	10	No	NA	Improved	No
Berlis *et al*., 2006	Retrospective	1	No	6	Improved	No
Cinalli *et al*., 2006	Retrospective	1	No	12	Improved	No
Fujimoto *et al*., 2007	Retrospective	1	No	48	Improved	No
Park *et al*., 2011	Retrospective	13	Yes	32	Improved	54%
Takano *et al*., 2015	Retrospective	9	Yes	73	Improved	11%
Shukla 2015	Retrospective	3	Yes	6-11	Improved	No
Lauretti *et al*., 2018	Retrospective	8	Yes	56	Improved	12.5%
Present series	Retrospective	32	Yes	3-76	Improved	18.7%

N/A – Not available

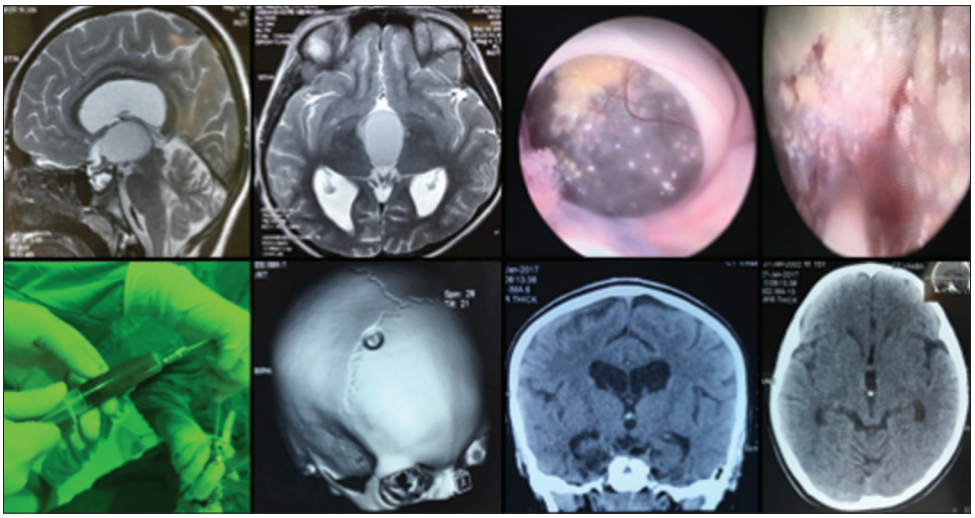

Figure 1 : Endoscopic cystic decompression and cystocisternostomy. (Patient no 13 [Table 2]) Top row images 1 and 2 – Magnetic resonance imaging T2 images show cystic craniopharyngioma with extension to the 3rd ventricle. Top row images 3 and 4 – Endoscopic images of cystic craniopharyngioma occluding the foramen of Monro and endoscopic intracavitary image showing flecks of calcification. Bottom row image 1 – Fluid aspirated from cystic craniopharyngioma -"Machine oil." Bottom row images 2-4 – Computed tomography images shows site of burr hole, shows decompression of cystic cavity and regression of ventriculomegaly (follow-up computed tomography at 6 months)

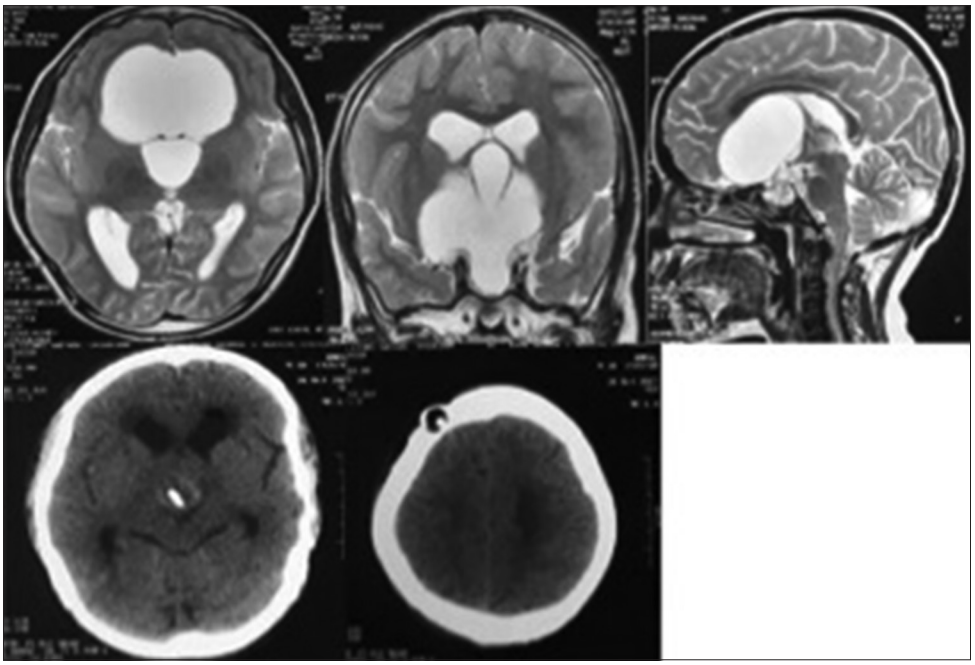

Figure 2: Top row images1-3 – Magnetic resonance imaging T2 images axial, coronal, and sagittal section shows hyperintense cystic craniopharyngioma extending into the third ventricle with ventriculomegaly. Bottom row image 1 and 2 – Computed tomography axial sections show catheter tip into the cystic cavity and Ommaya reservoir at the burr hole site

meningitis subsequent to spillage of cyst contents "machine oil" was reported.[22,23] No complication pertaining to the cyst-ventricle communication or spillage and absorption was seen in this study and none were reported by the other series.[7,13,20,24] The careful fenestration and aspiration of the cyst helps in avoiding the spillage of cyst contents into the ventricle thus minimizing chances of chemical meningitis. It was not seen in this study and has been rarely reported.[13,20] No intracavitary treatment with bleomycin or interferon alpha was used in the present study. There are no conclusive results described in the literature supporting the use of intracavitary chemotherapy.[2,3,23,25,26,27]

Complications noted in the series were similar to any series with procedures related to endoscopic transcranial procedures and all were managed effectively.[9,28,29,30,31] They were far less as compared to those encountered in the resection procedures and surely had lower morbidity.[1,29,32,33,34]

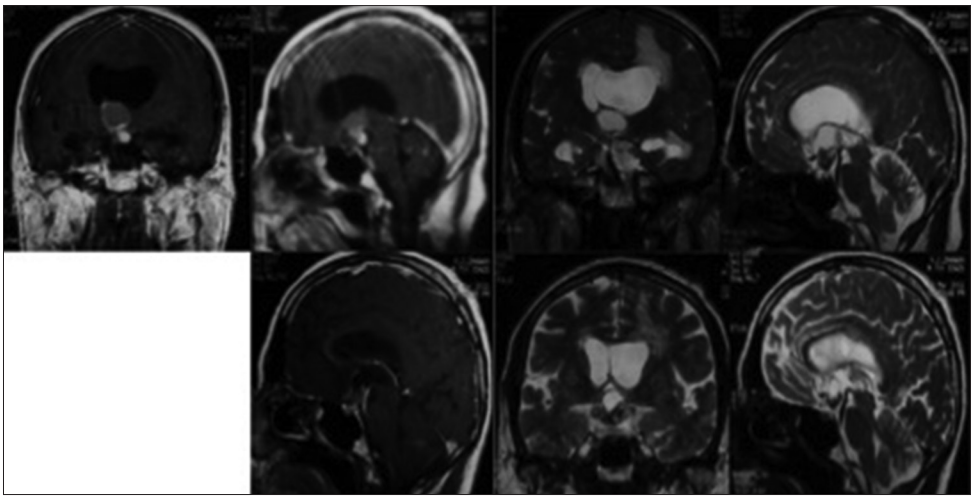

Figure 3: The patient underwent cystocisternostomy. Top row images 1 and 2 – Magnetic resonance imaging T1 contrast coronal and sagittal section shows contrast-enhancing cystic craniopharyngioma reaching up to foramen of Monro with ventriculomegaly. Top row images 3 and 4 – Magnetic resonance imaging T2 coronal and sagittal section shows hyperintense cystic craniopharyngioma Bottom row images 1-3 – Magnetic resonance imaging T1 contrast sagittal, T2 coronal and sagittal sections show reduction and regression of cystic cavity and improvement of ventriculomegaly

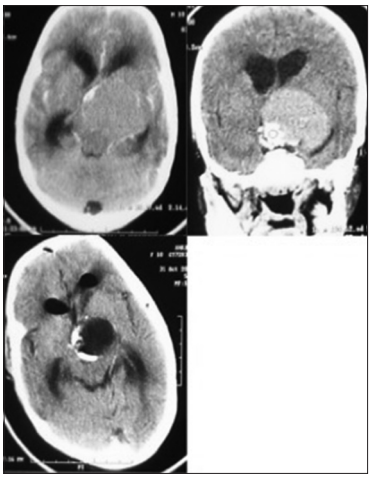

Figure 4: Top row images 1 and 2 – Computed tomography axial and coronal sections show large cystic suprasellar tumor with calcification. Bottom row image - catheter in situ in cystic cavity with reduction in cyst volume

Transventricular endoscopic management begets superior results as compared to microsurgical techniques in terms of less postsurgical morbidity and complications along with drastically improved patient's symptomatology. It is also better than other techniques of cyst drainage as it gives real-time direct visualization of the procedure and the catheter placement in the cavity.

Radiotherapy's role in craniopharyngioma is controversial, and currently, adjuvant radiotherapy is given in cases of residual tumors after resection to prevent relapse.[35,36,37] Though concerns regarding damage to optic apparatus and endocrinopathies as well as inducing malignant transformation have been raised.[38,39,40] Rachinger *et al*. recommended to withhold or postpone radiotherapy in majority of cystic craniopharyngioma.[18] Lauretti *et al*. suggested that radiotherapy can be avoided after cyst drainage and should be given for recurrent or progressive cases[41] but several other series have conclusively prescribed radiotherapy for progression-free survival in cystic craniopharyngioma [Table 4].[20,36,37,41,42] In the present study, external beam radiotherapy was given in all 32 patients with good tumor control in follow-up with only 18.7% (6/32) patients showing recurrence. Post radiotherapy, no visual deterioration was noted in the follow-up. Therefore, we suggest radiotherapy to be given as adjuvant therapy following neuroendoscopic fenestration and drainage.

Limitation of the study

This study has few limitations. First, this is a retrospective single-institution series which stands low on the pyramid of the level of evidence. Second, the sample size is not large enough even though it is the largest series on the cystic craniopharyngioma treated by neuroendoscopy.

Conclusions

Neuroendoscopic transcortical transventricular drainage of cystic craniopharyngioma is a simple, safe, and effective treatment and lacks many risks associated with microsurgery. Cyst ventricle communication in this procedure effectively prevents reaccumulation and Ommaya reservoir catheter further reinforces this communication. Radiotherapy is to be given as adjuvant modality following neuroendoscopic decompression. Consensus on ideal management of craniopharyngioma is still far from over therefore multi-institutional collaborative study involving all treatment modalities should be formulated for obtaining more relevant statistical information to come to a definite conclusion.

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Conflicts of interest

There are no conflicts of interest.

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