

Role of Ommaya Reservoir System in Management of Grossly Cystic Craniopharyngioma

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ABSTRACT

Introduction: This is a retrospective study of 16 patients with grossly cystic craniopharyngiomas (primary cystic or recurrent cystic craniopharyngiomas) whom were treated surgically by implantation of Ommaya reservoir system (ORS) in department of neurosurgery, Cairo university hospitals and Nasser institute hospital. **Objective:** The objective of this study was to evaluate the role of percutaneous drainage using the Ommaya reservoir system as a therapeutic option in the management of grossly cystic craniopharyngiomas. **Patients and methods:** Between January 2004 and December 2007, 16 patients were referred for insertion of Ommaya reservoir system for drainage of their cystic craniopharyngiomas. All procedures were performed at Neurosurgery Department, Cairo University hospitals and Nasser Institute Patients were selected on the basis of having their CT scan and MRI disclosure of: 1- Unilocular cystic craniopharyngioma, either primary or recurrent. 2- The craniocaudal dimension of the cyst was at least 3cm. 3- The cyst wall was not too thick or calcified to be easily punctured. There were 9 males and 7 females vary in age from 7 to 52 years. **Results:** The results with regard to cyst response after Ommaya reservoir system placement with radiosurgery or radiotherapy. Three patient (18.75%) the cyst disappear completely, Nine patients (56.25%) the cyst decrease in size and four patient (25%) need infrequent aspiration. **Conclusion:** Treatment of grossly cystic craniopharyngiomas (primary or recurrent) by percutaneous aspiration via Ommaya reservoir system can be considered as a suitable treatment option. The drainage procedure is expected to improve the results of subsequent treatment either using radiotherapy or radiosurgery by reducing the volume of the target and overcoming problems related to the fluid portion of the tumor on which the ionizing radiation has no effect.

Keywords: Craniopharyngioma, Ommaya reservoir, Therapeutic option.

INTRODUCTION

Craniopharyngiomas are benign slowly growing tumours that are located within the sellar and parasellar region of the central nervous system. They constitute from 2.5 to 4% of all intracranial tumors⁽⁵⁾. The onset of symptoms is normally insidious with most patients at diagnosis having neurological (headaches, visual disturbances) and endocrine (growth retardation, delayed puberty) dysfunctions. Craniopharyngiomas are thought to arise from epithelial remnants of the craniopharyngeal duct or Rathke's pouch (adamantinomatous

type) or from metaplasia of squamous epithelial cell rests that are remnants of the part of the stomadeum that contributed to the buccal mucosa (squamous papillary type)⁽¹³⁾. The neuroradiological diagnosis is mainly based on the three components of the tumour (cystic, solid and calcified) in the characteristic sellar/para sellar location. Definitive diagnosis is made following histological examination of a surgical specimen. The differential diagnosis includes other tumours in this region (pituitary adenoma), infectious or inflammatory processes (eosinophilic granuloma), vascular malformations (aneurysm) and

congenital anomalies (Rathke's cleft cyst)⁽¹⁷⁾. The current treatment is gross total excision of the tumour, but in the presence of hypothalamic invasion, a subtotal resection with post-operative radiotherapy will be beneficial^(19,20).

The ultimate goal of treating patients with craniopharyngioma is to maximize local control while minimizing acute and long-term related complications. Despite recent advances in aggressive and conservative approaches to these tumors and their benign histological nature, they are associated with a significant morbidity^(3,8). There are two major opposing views as to the surgical approach to these lesions. The ideal approach is a total excision, owing to the fact that a partial or subtotal excision will bring sooner or later to a recurrence. However, because a total excision is not always possible in every case and often results in major deficits such as hypothalamic dysfunction and adjacent nerve injury, a palliative operation as insertion of ommaya reservoir in combination with radiosurgery or radiotherapy may be preferred^(2,4).

Special therapeutic options are open to mainly cystic craniopharyngiomas, which represent as much as 60% of cases. When the tumor is predominantly composed of fluid, its reduction by percutaneous aspiration may enable palliative relieve of most of the problems connected with the neoplastic mass and allow using alternative therapies for the tumour. The Ommaya reservoir system is used as a permanent percutaneous draining system consisting of an inert catheter, provided with few holes at its distal end, indwelling within the cyst and connected with a subcutaneous reservoir. Since the therapeutic role of percutaneous drainage of

craniopharyngioma has seldom been stressed, precise knowledge about its usefulness and its main technical features is not easily obtained from the literature^(1,16).

The aim of this study was to evaluate the role of percutaneous drainage using the ORS implanted by open surgery as a therapeutic option in the management of primary and recurrent grossly cystic craniopharyngiomas.

PATIENTS & METHODS

Between January 2004 and December 2007, 16 patients were referred for insertion of ORS for drainage of their cystic craniopharyngiomas. All procedures were performed at Neurosurgery Department, Cairo University hospitals and Nasser Institute Patients were selected on the basis of having their CT scan and MRI disclosure of:

1. Unilocular cystic craniopharyngioma, either primary or recurrent.
2. The craniocaudal dimension of the cyst was at least 3cm to ensure that the whole distal perforated portion of the catheter which is 1.6 cm. in length was totally introduced into the cyst cavity at the time of placement and to ensure that cyst aspiration would be of clinical benefit.
3. The cyst wall was not too thick or calcified to be easily punctured by the ventricular catheter with its inner stylet.

Surgical procedure:

Two different models of ORS were used. In 10 patients (62.25%), the standard ORS with flat-bottom reservoir and side-inlet catheter was used, while in 6 patients (37.75%), the

burr-hole reservoir with bottom-inlet catheter was used. Each model has its advantages and disadvantages. For the flat- bottom model, a twist drill hole is sufficient to pass the catheter into the brain. Its disadvantage is that the reservoir is dome-shaped, 7.2 mm in height, and this might exert some difficulty in subsequent puncturing and maintaining the needle within the reservoir during aspiration. The burr-hole model is conical in shape (1.5 cm in height) making subsequent aspiration much easier. However its use is more time consuming as it requires performing a burr hole. Moreover, attachment of the catheter to the bottom inlet reservoir may inadvertently push the catheter deeper into the brain.

In all cases. A transfrontal approach with an entry point 2 to 3 cm off midline at the coronal suture was chosen to access all lesions. A U-shaped flap centered over the entry point was drawn with its base. The stylet accompanying the standard ventricular catheter was inserted directly to the predetermined target distance. Then the catheter was cut at the correct length, connected to the reservoir which was secured to the periosteum by a silk suture. In case of the side-inlet reservoir, a subcutaneous pocket was made away from the incision and the reservoir was inserted

inside and connected to the catheter using a right-angle connector. Before skin closure gentle cyst aspiration was performed using an insulin needle (a 21 gauge needle) aiming at reducing the cyst pressure and not emptying the cyst completely.

CT scan was obtained within 24 hours after placement to verify position of the catheter, ensure satisfactory cyst drainage and exclude cerebral hemorrhage.

Follow up:

Patients were requested to be seen in the out-patient clinic every month for the first 3 months and then every 3 months for the first year. Assessment involved full neurological and ophthalmologic examination as well as hormonal investigations whenever indicated. Follow-up contrasted CT was performed at one, and 6 months' visits, and whenever there was clinical suspicion of cyst recurrence. MRI was performed routinely at the end of the first year, and then annually thereafter.

RESULTS

In this study there were 9 males and 7 females vary in age from 7 to 52 years characteristics of patients are shown in table 1.

Table (1): Characteristics of 16 cystic craniopharyngioma cases

Characteristic	Classification					
		No.	%	No.	%	
Age	≤ 16 years	11	68.75	> 16 years	5	31.25
Sex	Males	9	56.25	Females	7	43.75
Tumor Type	Purely cystic	5	31.25	Mixed	11	68.75

The most prominent clinical manifestations were related to increased intracranial pressure (ICP)

and to visual and endocrinological impairment. The preoperative clinical findings are illustrated in table 2.

Table (2): Clinical findings in 16 patients with cystic craniopharyngiomas prior to the drainage procedure

Symptoms and /or signs	No. of cases	Percent
⇒ Headache	9	56.25
⇒ Depressed mental status	3	18.75
⇒ Related to visual dysfunction:		
- decreased visual acuity	13	81.25
- papilloedema	6	37.5
- optic atrophy	4	25
- 6 th nerve palsy	2	12.5
⇒ Related to endocrine dysfunction:		
- increased prolactin level only	1	6.25
- panhypopituitarism (growth retardation, diabetes insipidus, asthenia).	11	68.75

Early clinical outcome 14 patients (87.5%) showed significant clinical improvement in the early postoperative period. The remaining 2 patients (12.5%) did not improved In table (3). This consisted of either improvement in symptoms and signs of raised ICP or improvement of with preoperative impairment of visual acuity and/or field. Moreover, there was complete improvement in the recent 6th nerve palsy encountered in 2 patients preoperatively. Of the 2 patients presented with depressed mental status,

1 patient improved significantly, while the other had not improved. We facing technical problems during insertion of Ommaya reservoir system catheter in two patients due to difficulty of penetrating the cyst in two cases. Catheter obstruction with debris was encountered in 2 patients and necessitated revision.

Procedure related morbidity such as haematoma, infection or new neurological deficit was not present. Table (4).

Table (3): Clinical improvement after insertion of Ommaya reservoir system

Clinical improvement	No. of cases	Percent
Improved	14	87.5
Not Improved	2	12.5

Table (4): Difficulty and complications with insertion of Ommaya reservoir system

	No. of cases	Percent
Difficulty of penetrating the cyst wall	2	12.5%
Catheter obstruction	2	12.5%
Haematoma	0	0%
Infection	0	0%

Subsequent aspiration:

Three patients (18.75%); 2 with purely cystic and 1 with mixed tumors, showed spontaneous and complete regression of their cysts and this was confirmed by repeated post-operative neuro-imaging. Thirteen patients (81.25%) required multiple aspirations for symptomatic recurrence of their cysts performed at intervals of 4 to 8 weeks. Nine of these patients (56.25%) finally stabilized over a period of 6 months and did not require further aspiration afterwards. The remaining 4 patients (25%) continued to require infrequent aspiration at intervals of 6

months to two year. Two of them underwent open craniotomy.

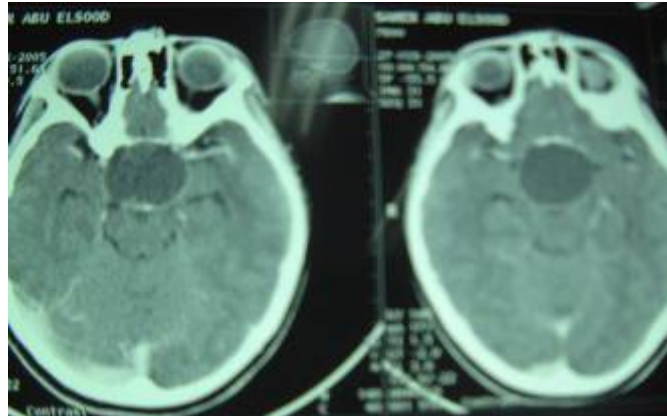
Final outcome:

The duration of follow-up was determined as the time elapsed between ORS placement and the time the patient was last seen or underwent subsequent craniotomy. For all patients, the duration of follow up ranged from 1 to 43 months. The results with regard to cyst response after Ommaya reservoir system placement with radiosurgery or radiotherapy are shown in table(5). Three patient the cyst disappear completely, Nine patients the cyst decrease in size and four patient need infrequent aspiration.

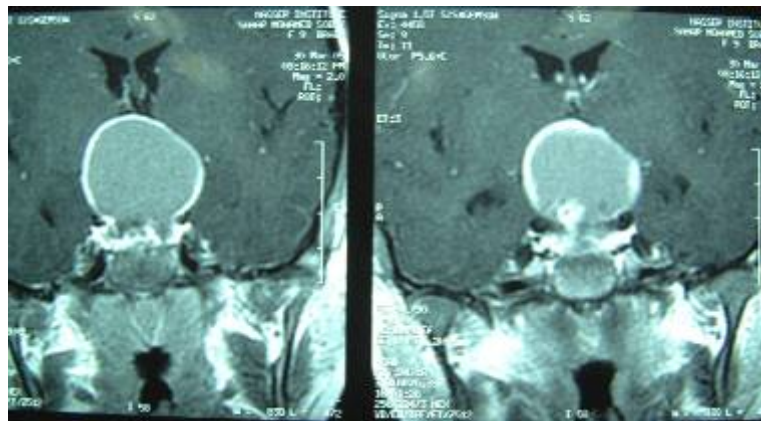
Table (5): Tumor cysts' response after treatment in the 16 survived patients

Response	Nature of the tumor					
	Purely cystic		Mixed			
	No.	%	No.	%	Total	%
Cyst disappeared	2	12.5%	1	6.25%	3	18.75%
Cyst decreased	3	12.5%	6	37.5%	9	56.25%
Required infrequent aspiration	0	0	4	0	4	25%
Total	5	25%	11	68.75%	16	100%

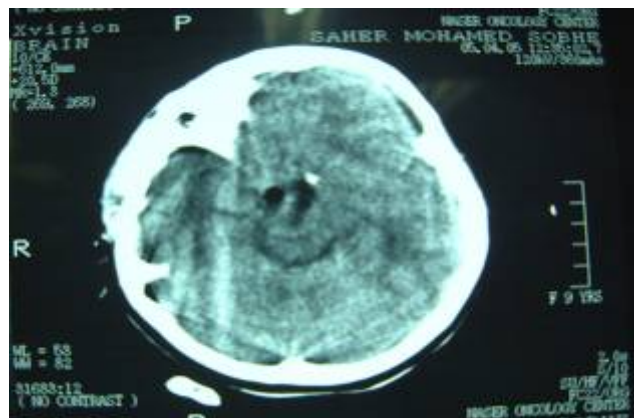
Case (1):



CT scan axial cut showing unilocular cystic craniopharyngioma

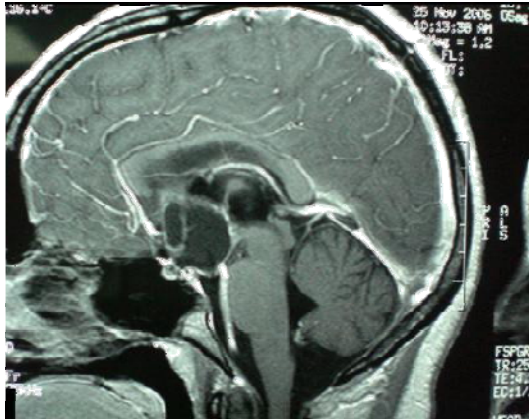


MRI brain coronal cut showing unilocular cystic craniopharyngioma

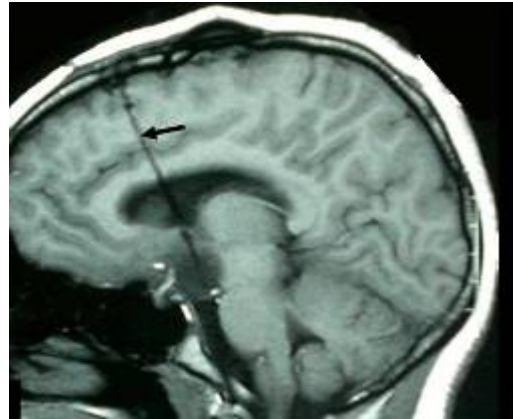


CT scan axial cut showing Ommaya reservoir system tube in cystic craniopharyngioma

Case (2):

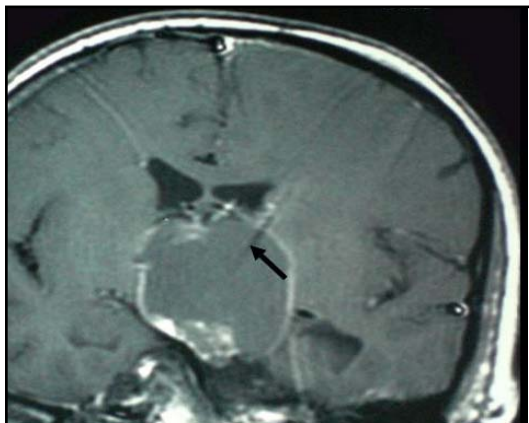


Sagittal gadolinium enhanced MRI. A cystic craniopharyngioma in a 16 years-old female.

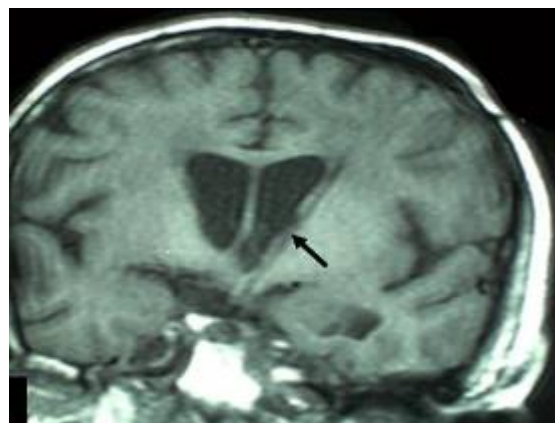


Sagittal gadolinium enhanced MRI. Obtained 12 months following ORS placement and radiotherapy showing marked cyst regression

Case (3):



Coronal gadolinium enhanced MRI obtained in sellar and supra-sellar cystic craniopharyngioma with minimal solid component. Following Ommaya reservoir system placement, the cyst required a single episode of aspiration followed by spontaneous regression and the patient received radiotherapy.



Coronal gadolinium enhanced MRI obtained in sellar and supra-sellar cystic craniopharyngioma with minimal solid component. Solid tumor and complete cyst regression 28 months following treatment.

DISCUSSION

Craniopharyngioma very frequently presents a cystic component (about 80% of cases and almost 100% of recurrences). For the past decade, the management of craniopharyngioma has emphasized both long term quality of life and tumor removal. Unfortunately, high rates of post-operative morbidity were noted. Radical resection remains important for patients with amenable solid or mixed tumours^(1,3,10,16). In the literatures a series of 168 patients treated with surgical resection, the authors reported recurrence-free survival rates of 87% at 5 years after complete resection and 49% after subtotal removal. Partial surgical resection followed by fractionated radiotherapy is also an important strategy that can provide tumor control in many cases. The recurrence rate following limited surgery and RT was found to range from 0 to 44% (mean 18%) which is favorable compared with the rate associated with surgery alone⁽¹⁴⁾.

Surgical treatment of recurrent craniopharyngioma is considered more challenging than primary surgery because scars produced by previous surgery may enhance the technical difficulty of second surgery and therefore increase the risks of morbidity and mortality. As a result, many authors who prefer to perform radical resection in the first surgery recommend radiotherapy or multimodal approach as a salvage treatment of recurrent cases^(6,7).

Special therapeutic options are open to grossly cystic craniopharyngioma. Given that this

tumor is histologically benign, in theory it should be enough:

1. To reduce production of fluid in the cyst and/or
2. To prevent accumulation and encourage its discharge either to the outside or into the CSF pathways.

The first goal is difficult to achieve because of the neoplastic nature of the cyst wall so, it is necessary to act by means which reduce its proliferation. This can be obtained by radiotherapy via the interstitial route, using colloidal isotopes, or via the external route. The second goal, that is drainage of the cyst contents, is a palliative treatment which can be easily and rapidly achieved^(9,11).

Because in many cases frequent drainage procedures are required, the indwelling Ommaya reservoir system was used for draining a recurrent, cystic craniopharyngioma. Other series have since documented the indications for and the safety of implantation of this type of access into cystic craniopharyngioma. However, clear evaluation of clinical results is somewhat difficult because only few reports regard such treatment as therapeutic option^(12,15).

In this retrospective study, the effectiveness of this method combined with external beam radiotherapy or radiosurgery as a definitive treatment in certain cases of primary or recurrent cystic craniopharyngiomas were looked at by assessment of the early post-operative results as well as the long-term outcome with regard to the clinical and local tumor responses.

Technical problems with placement of ORS in cystic craniopharyngiomas have rarely been discussed. One of the main technical problems is the

difficulty of penetrating the cyst capsule due to its consistency, as well as the presence of multilocular partitions which prevent the complete drainage of the cyst contents⁽²⁰⁾. In the present series, patients were selected on the basis of having unilocular cyst with thin non-calcified wall in the pre-operative imaging to avoid these problems. However, a catheter resistance to penetrate the cyst wall was encountered in 2 patients. In these 2 occasions, the standard brain canula was used to approach the cyst. Once the cyst wall was felt yielding underneath the needle, the inner trucker was removed and the stylet accompanying the standard ventricular catheter was introduced to puncture the cyst. The needle was then removed and the Ommaya catheter with its inner stylet was introduced into the cyst through the same trajectory.

Another major technical difficulty has been in achieving adequate catheter placement because of the proximity of these cysts to normal CSF spaces or partial collapse of the cyst cavity following aspiration and then the presence within the ventricle of part of the distal perforated portion of the catheter.

This is why the initial cyst puncture should aim to only reduce the intracranial hyperpressure and not to empty the cyst. Accurate catheter placement can also be lost because of the manipulation required in connecting the catheter to the reservoir. Review of the results in the literatures series shows that unsuccessful catheter placement occurred in 7% of their patients^(15,20). In the present study, this difficulty was tried to be overcome by pre-operative selection of patients having cysts with craniocaudal length of 3cm. or more, and the target was chosen in the bottom of the cyst. This

is to ensure that the whole distal perforated portion of the catheter which is 1.6 cm. in length was totally introduced into the cyst cavity at the time of placement.

Although complications following these procedures are very rare, reports of frontal lobe hemorrhage and subarachnoid hemorrhage resulting in death have been documented⁽¹⁵⁾.

In the present study, no hemorrhage occurred as cleared by early post-operative CT scan.

In the present study, early post-operative clinical improvement was noticed in 14 out of 16 patients. The remaining 2 patients remained stable. This was coping with many literatures that reported excellent early clinical results in their series of 9 patients with primary (3 patients) or recurrent cystic craniopharyngiomas (6 patients) treated by drainage procedures⁽²¹⁾.

Possible late complications related to the use of Ommaya reservoir system include infection and obstruction of the catheter with debris or viscous fluid⁽¹⁵⁾. literature reported a series of 20 patients who had Ommaya systems placed to access cystic craniopharyngioma with no case of infection or obstruction of the catheter⁽²⁰⁾.

In the current series, there was no case of infection, while catheter obstruction with debris was encountered in 2 patients and necessitated revision.

In the literature, operative mortality ranges from 0-14% and averages 3.7% in a multiseries review⁽¹⁸⁾. In the present study, no operative mortality.

CONCLUSION

Treatment of grossly cystic craniopharyngiomas (primary or

recurrent) by percutaneous aspiration via Ommaya reservoir system can be considered as a suitable treatment option. The drainage procedure is expected to improve the results of subsequent treatment either using radiotherapy or radiosurgery by reducing the volume of the target and overcoming problems related to the fluid portion of the tumor on which the ionizing radiation has no effect.

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