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NEUROENDOSCOPY IN CYSTIC CRANIOPHARYNGIOMA

Neurosurgery

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ABSTRACT

Objective - Gross total resection of cranipharyngioma is technically challenging because it is surrounded by vital structures. Ideal management of such patient should be safe excision of the tumor without adding neurological deficits. Multidisciplinary care like endoscopic decompression of cystic craniopharyngioma helps in reducing the operative morbidity. The aim of this study was to evaluate the role of neuroendoscopy in the placement of ommaya reservoir in patients of cystic craniopharyngioma.

Method - Thirteen patients with symptomatic cystic craniopharyngioma associated with hydrocephalus were treated by an endoscopic transventricular approach for the insertion of an intracystic catheter

Result - All patients except one show significant improvement in immediate post operative period. There was no evidence of chemical meningitis in post operative period. Three patients require ventriculo- peritoneal shunt at a later date for hydrocephalus. In two cases cysto-ventriculo-cisternostomy was performed for managing hydrocephalus.

Conclusion - This type of neuroendoscopic management has proven minimally invasive, safe, and effective and could be considered as an alternative management technique for some cystic craniopharyngiomas

KEYWORDS

Craniopharyngioma, Cystic, Endoscopy, Multidisciplinary, Ommaya, Transventricular

Introduction

Craniopharyngioma are solid or mixed solid- cystic benign tumors that arise from remnants of rathke's pouch. They are usually located in suprasellar and parasellar location and account for 1.2–4% of all primary intracranial neoplasms and 5–10% of intracranial tumors in children.[1]

Ideal management of such patient should be safe excision of the tumor without adding neurological deficits to the patient. Gross total resection is technically challenging because it is surrounded by vital structures like optic nerves and chiasm, hypothalamus, third cranial nerve and vessels of circle of willis. These tumors are mostly adhered to these structures thus limiting the ability to remove the entire tumor. Almost all craniopharyngiomas have an attachment to pituitary stalk, and radical surgery in such patients may lead to life-long pituitary hormone replacement therapy. [2,3]

Multidisciplinary care like limited surgery followed by radiation therapy helps in reducing the operative morbidity. Out of many minimally invasive procedures used, one approach described in literature is the placement of ommaya reservoir in cystic cavity of craniopharyngioma followed by cyst decompression and radiation therapy. Ommaya reservoir helps in repeated aspirations of cyst if needed during the course of therapy. Ommaya can be placed blindly or by stereotactic methods. Endoscopic guided placement of ommaya reservoir helps in taking biopsy, and for optimal placement of tip of ommaya catheter.

Material and methods:

Patient Population

Thirteen patients with symptomatic cystic craniopharyngioma associated with hydrocephalus were treated by an endoscopic trans ventricular approach for the insertion of an intracystic catheter. Clinical, radiological and endocrine evaluations were done in all the patients. Ten patients belong to pediatric age group (<16 years). Male: Female ratio was 8:5. Their signs and symptoms included headache (10 cases), vomiting (7 cases), visual disturbances (10 cases), altered behavior (3 cases) and endocrine disturbances (4 cases). In all cases, MR imaging showed a large cyst with a solid component, with contrast enhancement of the solid component and the cyst wall (Fig. 3& 4). A summary of the clinical data is presented in Table 1.

Surgical Technique:

Neuroendoscopy was performed with Karl Storz rigid endoscope. A single right pre-coronal bur hole is made 1 cm anterior to the coronal suture and 3.5 cm from the midline. This position of burr hole is little lateral to usual burr hole made for endoscopic third ventriculostomy, so as to have ease in performing septostomy. After entering the frontal horn of the lateral ventricle, the dome of craniopharyngioma cyst is usually visualized immediately under the foramen of Monro. (Figure 1) Small portion of the translucent cystic surface is coagulated and a catheter is inserted into the cyst to aspirate the cystic content. Through the same opening thus made, endoscope is entered into the cystic cavity. Interior of the cystic cavity is inspected and septations if present are broken down to convert multi-cystic compartment into a single cavity. Tissue is taken from the solid portion of the tumor for histopathological examination. In two of our patients floor of the third ventricle was visualized through the transparent cystic wall. A stoma for third ventriculostomy was made in pre-mammillary area through the cystic cavity (cysto-ventriculostomy) in these two cases. Ventricular catheter is inserted under endoscopic guidance (Figure 2).

Results:

Mean follow up was two year. All patients except one show significant improvement in immediate post operative period. One of our initial patients requires craniotomy with excision of tumor due to non-improvement in visual symptoms. On retrospective analysis we found that in that case the solid portion of tumor was compressing the optic chiasm, hence visual symptoms did not improve even after decompression of cyst. We recommend that in patients of cystic craniopharyngioma with primary optic atrophy, if there is significant compression on optic pathways due to the solid portion of tumor than this procedure is not useful.

There was no evidence of chemical meningitis in postoperative period. No cognitive disturbance was observed in the patients. There was neither intra, nor peri-operative mortality. Three patients require ventriculo- peritoneal shunt (univentricular) at a later date for hydrocephalus. Patients were subjected to radiotherapy after wound healing.

Discussion:

Craniopharyngiomas being a benign tumor should have relatively good long-term outcome. Its optimal management is controversial. Ideal management of such benign entity should be gross total removal without adding co morbidities to the patient. As the tumor is surrounded and adhered to vital structures hence its complete resection may lead to serious co morbidities.

Multiple surgical series in which total excision of craniopharyngiomas was attempted in children using microsurgical techniques have noted considerable morbidity, an operative mortality rate as high as 20 percent, a recurrence rate of 23 to 50 percent, and frequent long-term neurologic, endocrine and ophthalmic complications (1,2,5).

After radical surgery endocrine deficiency requiring lifelong hormone replacement is reported in 70 to 100% of patients. (4,5,6) Visual complications, including decreased visual acuity and constriction of the visual field is seen in up to 67% of patients (7). Vascular complications of surgery for suprasellar tumors include carotid artery

laceration, vasospasm, and delayed aneurysmal dilatation of the carotid artery. In one series of pediatric patients, fusiform dilatation of the carotid artery was reported in 15.7% of patients. (8)

Morbidity due to hypothalamic damage can result in obesity, defective short-term memory, limited concentration span, defective thirst sensation, and sleep disturbances. (9) Morbid obesity after radical surgery has been reported in 30 to 50% of children secondarily to hypothalamic injury (10,11,12).

Neurocognitive dysfunction, including difficulties with concentration, learning, and memory are well-known complications associated with radical surgery.(10,13) 50% of patients experienced significant problems in school.(14) Difficulties in interpersonal relationships have been reported in 9 to 50% of patients who had radical surgery (15,16,17). One series has demonstrated anxiety in 42% (18).

Thus, many surgeons have adopted a more conservative approach, which combines less aggressive surgery with radiotherapy.

Proponents of subtotal resection in conjunction with radiotherapy argue that this less aggressive approach can yield equivalent control rates with lower morbidity.

Studies have found that craniopharyngioma patients treated with partial resection and radiation have 10-year survival of 83.3% and 20-year survival rates of 67.8% . This is comparable to the rates mentioned for complete surgery alone, but with much lower rates of added complications.

Conservative surgery in cystic craniophayngioma

Cystic craniophayngioma due to large cyst and associated hydrocephalus usually present with sign and symptoms of raised intracranial pressure. For such large cystic craniopharyngiomas implantation of an intracystic catheter with a subcutaneous reservoir (ommaya) may be a valuable alternative treatment option.

Placement of ommaya catheter has been performed under direct vision, (19, 20) stereotactic guidance,(21) ultrasound and ventriculoscopic guidance,(19) and fluoroscopic guidance,(22) as well as with stereotactic endoscopic technique.(23)

Moussa et al,(24) has inserted ommaya in 52 patients of cystic craniopharyngioma and followed them for 7 years. 73% did not develop recollection of cyst after single aspiration, while 19% required re-aspiration after every 6 month. He concluded that majority of patient did not need any further treatment except for observation after ommaya reservoir placement in cystic craniopharyngioma.

Another advantage of ommaya reservoir is that in case of any deterioration during radiotherapy, tapping of ommaya reservoir can be done so as to complete the radiation therapy with a good outcome.

Many researchers have used stereotactic approaches for cystic craniophryngioma.(25) It helps in the taking biopsy from the solid part of the tumor or from the wall of a cyst. Further, it can be used to evacuate the cystic parts of the lesion by aspiration or by the implanting the catheter of ommaya reservoir, which can be used for re-aspiration at later date if needed. Cyst evacuation results in lowering of pressure over surrounding structures like optic pathways and hypothalamus. It also prepares the patients for subsequent, fractionated, external radiotherapy. Presence of ommaya reservoir also facilitates various intracystic therapies. Leakage of dye has to be checked before installation. Various Substances used for intracystic installation in cystic craniopharyngiomas are Radioisotopes, Bleomycin, and Interferon.

Endoscopy in cystic brain lesions

The endoscopic transventricular approach has been reported as a safe and minimally invasive technique for the management of many intraventricular cystic lesions including suprasellar cysts, (26,27) Rathke cleft cysts, colloid cysts,(28) and paraventricular cystic lesions(29)

Another advantage of using the endoscopic approach is the possibility to perform a septostomy at the same time so as to establish a connection between the two lateral ventricles, which in case of subsequent requirement facilitates successful univentricular shunting (in comparison to bi ventricular shunting usually required in cystic craniopharyngioma due to blockage of bilateral foramen of monro). We have done septostomy in all our cases. Three patients who later developed hydrocephalus were managed by a univentricular shunt.

Hellwig(30) have used endoscopic techniques in 70 patients of cystic brain lesions. The main diagnoses were colloid cysts, cystic craniopharyngeoma, arachnoidal and pineal cysts. Operative morbidity below 3% was reported in his series. In a paper by Tirakotai W et al (31) 46 patients with newly diagnosed solid or solid-cystic peri- and intraventricular tumours underwent neuroendoscopic procedures. He has used endoscope for taking tumour biopsy and for management of hydrocephalus.

Abdullah J et al (32) has used rigid endoscope for aspiration of cyst and removal of capsule by holding it with forceps in three patients. Total removal was achieved in one while partial removal of capsule was possible in two of his patients.

In a study by Alberto Delitala et al (33) steerable, flexible neuroendoscope was used for drainage and wide marsupialization of cyst into CSF spaces (cysto-ventriculo-cisternostomy). This procedure helps in control of mass effect, continuous dilution, and reabsorption of cystic fluid. In our series floor of the third ventricle was visualized through the transparent cystic wall in two patients. A stoma for third ventrculostomy was made in premammilary area through the cystic cavity (cysto-ventriculo-cisternostomy) in these two cases.

We found that under endoscopic guidance intra-cystic catheter can be placed successfully through single burr hole. We have not used navigation as the tip of catheter can be visualized by taking the endoscope inside the cyst ensuring its correct placement.

Conclusions:

This type of neuroendoscopic management has proven minimally invasive, safe, and effective and could be considered as an alternative management technique for some cystic craniopharyngiomas. Our preliminary experience should stimulate the recruitment of larger groups of patients in order to define the role of neuroendoscopy in the multimodal treatment of craniopharyngiomas.

Table – clinical presentation and results

Presentation	No. of patients (n=13)	Improvement in symptoms
Headache	10/13	9/10 (90%)
Vomiting	7/13	7/7 (100%)
Visual disturbances	10/13	8/10 (80%)
Altered behavior	3/13	3/3 (100%)
Endocrine disturbances	4/13	2/4 (50%)
Hydrocephalus	13/13	10/13 (77%)
Seizure	7/13	2/7 (28%)

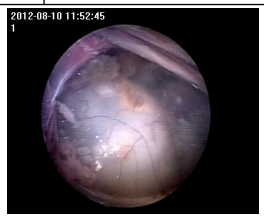

Fig.1 craniopharyngioma cyst protruding at foramen of monro

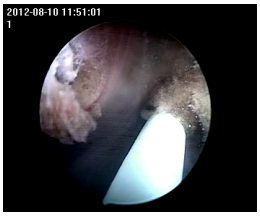

Fig 2. ventricular catheter of ommaya in cystic cavity

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