Long Term Outcome of Drainage of Midline Deeply Seated Recurrent Brain Cysts by Ommaya Reservoir

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ABSTRACT

Background: Midline deeply seated brain cysts carry great risk during its surgery especially when they are recurrent and extend vertically via the third ventricle to the floor of lateral ventricle. Objectives: To evaluate the effect of Ommaya Reservoir in controlling recurrent cystic midline pathologies and improve their functional outcome. Patients and Methods: This prospective study was conducted on 19 recurrent cases (13 cystic recurrent craniopharyngiomas 68% and 6 cystic recurrent gliomas 32%) had been treated between June 2005 and June 2011 at neurosurgery department of Zagazig University Hospitals – Egypt, by using Ommaya Reservoir and evacuation of the cyst content to decompress the brain. All cysts had cytological examination of their contents to reconfirm the previous pathological diagnosis. Decompression of the brain cysts was carried out by periodic aspiration of the reservoir according to neurological condition of the patient and radiological assessment of the cyst. All cases received optimum dose of radiotherapy according to the pathology and the surrounding structures except 6 kids younger than 3 years who periodically decompressed by Ommaya reservoir till that age when referred to radiotherapy. All cases were followed up to 36 months. Results: The age ranged from 18M to 43 years in craniopharyngioma group with mean age 16 years and from 19 months to 18 years in glioma group with mean age 6 years. All craniopharingioma cysts were considered responding except 2 cases (10.5%) had reasonable residual. 5 cases (26%) showed reduction in initial volume more than 75% and 8 cases (42%) more than 50%. Amount of aspirated fluid ranged from 5 to 15 mls in craniopharyngioma group and from 10 to 20 mls in glioma group. In 7 of 9 cases (78%) with ventricular dilation improved by cyst decompression and the ventricles became normal within 3 months without the need for a shunt. Visual function improved in 6 of 12 patients and remained stable in the rest of the patient at the end of the follow up period. 2 kids with unstable gait improved. 4 lethargic patients improved and became alert. Neither major clinical deterioration nor mortality reported with this conservative technique during follow up period. Conclusion: Because limited surgery for midline deeply seated recurrent brain cyst does not prevent recurrences and radical surgery carries unacceptable morbidity and mortality, postoperative external-beam radiotherapy has been added to limited surgery in an effort to improve local control. Children younger than 3 years may not be candidates for such radiotherapy because they can develop unusually severe long-term adverse effects. In those patients, stereotactic implantation of an intracystic catheter with Ommaya reservoir may be a valuable alternative treatment option. The benefits of this procedure include temporary relief of fluid pressure by serial drainage, may prolong the interval to or obviate the need for radiation. Key words: Ommaya reservoir (OR) – stereotactic-craniopharyngioma (CP) – cystic glioma (CG)

INTRODUCTION

Midline deeply seated brain cysts carry great risk during its surgery especially when they are recurrent and extend vertically via the third ventricle to the floor of lateral ventricle. One of these pathologies is craniopharyngioma that represents 2.5-4% of all intracranial tumors. Although half of these occur in adults, they account for a greater percentage of childhood tumors (5-13%) and are responsible for 54% of seller region pathology in children. Peaks occurring at ages 5-10 and 55-65 years. Craniopharyngioma was first described by Zenker in 1857. In 1899, Mott and Barrett stated that craniopharyngioma may arise from the hypophyseal-pharyngeal duct or Rathke’s pouch, but may arise by metaplasia of normally developed anterior pituitary cells. New craniopharingiomas represent 0.5–1.0 per million per year. Craniopharyngiomas are difficult to resect and there is usually significant long-term endocrine disturbance due to pituitary involvement; should the hypothalamus be involved, behavioral changes, obesity, learning and memory difficulties may occur. The effects of the tumor and its surgical treatment can result in a child with defective vision, in addition to an altered personality with behavioral problems; in some cases, life expectancy may be shortened because of vulnerability to a hypothalamic mediated metabolic crisis. Surgical mortality following attempts at radical resection of craniopharyngioma still high, varying in most series from 0 to 15% mainly due to endocrine, ophthalmologic and neuropsychological disturbances. Recurrence of craniopharyngioma occurs in approximately 35% of patients regardless of primary therapy. Repeat attempts at gross total resection are...
difficult and long-term disease control is less often achieved (38).

Due to the morbidity associated with radical surgical removal, more conservative methods of treatment are currently being attempted, including cyst aspiration and radiotherapy. Intracavitary brachytherapy and use of radioisotopes into craniopharyngioma cyst have been documented in the last two decades which introduced by Pollack (28). Radiotherapy has been developed by Kramer (18) and its role has a major controversy in treatment of craniopharyngiomas, despite evidence that radiotherapy alone provide long-term disease control for most patients, there has been much concern about its potential impact on growth, development and on performance (2). Radiation therapy is often not used or postponed in children younger than 3 years old to avoid damage that might affect brain development (6).

Tumors of the central nervous system often have associated cystic components. Gross total resection of the tumor and cyst is often enough to prevent recurrence of the cyst (29). Lesions in the brain stem, however, are often not surgically resectable. Stereotactic aspiration of brain stem cysts may provide temporary improvement in the clinical status of the patient. However, cysts regularly recur and require multiple aspirations. This necessitates frequent operative procedures and compounds the risk of passing the biopsy probe deep into the neuraxis (11).

Ommaya Reservoir systems have been placed to access fluid cavities within the brain since their original description in 1963 (26). Ratcheson and Ommaya in 1968 reported a series of 60 patients with implantation of Ommaya Reservoirs (29).

Stereotactic placement of an Ommaya catheter avoids multiple passes through the neuraxis and allows for aspiration of the cyst in an outpatient setting (41). This procedure may also be helpful in allowing the surgeon to perform a two-staged approach, whereby first the cyst is drained by the implanted catheter to relieve pressure and complicating symptoms, followed by tumor resection (34).

Stereotactic neurosurgery offers useful minimal-invasive treatment options in the interdisciplinary treatment regime of craniopharyngioma. These options must especially be considered if the solid part of the tumor is small, if there is a hypothalamic involvement, or if the probability for a complete resection is not favorable (39).

Stereotactic surgery involves accurate localization of intracranial target using a special frame fixed to the patient’s head, a preoperative imaging study and dedicated computer software (44). Experience has shown that outpatient stereotactic surgery is safe and effective option for selected patients with brain tumors, and that they are more resource friendly than standard approaches. Implementation of outpatient neurosurgery is not only an organizational but a social, political and cultural challenge. The time has come to embrace this new idea (29).

**Aim of the work:**

To evaluate the functional outcome of midline deeply seated recurrent predominantly cystic lesions continuously decompressed by Ommaya Reservoir.

### PATIENTS & METHODS

This analytical prospective study was conducted on 19 recurrent cystic brain lesions at neurosurgery department of Zagazig University Hospitals from June 2005 through June 2011 by using Ommaya reservoir and periodical aspiration. The catheter inserted by free hand in 2 cases (10.5%), CT-guided in 2 cases (10.5%) targeting the cyst center, endoscopically-guided in 2 cases (10.5%), manual Stereotactic technique in 5 cases (26%) and computed stereotactic technique in 8 cases (42%) targeting the most dependent part of the cyst.

We have two groups: **Group A:** 13 (68%) cystic craniopharyngiomas: All the patients were recurrent and received a course of radiotherapy except 4 cases (21%) who referred for Ommaya reservoir implantation till reaching the acceptable age of radiotherapy **Group B:** 6 (32%) recurrent cystic glioma after surgery, 4 (21%) of them had post-operative radiotherapy and 2 (10.5%) had not because of the age (under 3 years). All cases had full CT and MRI evaluation to define the exact topography of lesion.

Moreover, craniopharingiomas were classified by size according to the classification of Yasargil et al. (38) into (small <2 cm), moderate [2 to 4 cm], large [4 to 6 cm] and giant (>6 cm). The horizontal extension was classified according to Hoffman into sellar, anterior extension (prechaismatic), posterior extension (retrochaismatic) and lateral extension and also grows into various directions (giant) (34). The vertical extension classified according to Samii into Grade I: Intrasellar tumor, Grade II: Intracisternal tumor with or without intrasellar component, Grade III: Intracisternal tumor extending to the lower half of 3rd ventricle, Grade IV: Intracisternal tumor extending to the upper half of 3rd ventricle and Grade V: Intracisternal tumor extending to septum pellucidum or lateral ventricle (32). The inclusion criteria were recurrent monocular craniopharyngioma and glioma (Cystic component >60% of the tumor volume) and the exclusion criteria were multicystic, solid craniopharyngioma or glioma.

**Cyst Volume Calculation:** It was possible to analyze and compare the pre and the post-treatment tumor volume using the modified ellipsoid volume equation: A x B x C x 0.52; where A, B and C are the major diameters measured in the 3 special planes and...
chosen by convention (A transfrontal approach with a small skin flap and twist-drill hole is performed at entry that passes through the shortest appropriate trajectory to volume (software calculation). Moreover, visual path Y, Z and distance between entry and target points, cyst both ring and arc angles, target point by calculating X, Y, Z of the target point (the most dependent part of the cyst) on CT console and our team work fashioned endoscopic application.

Target Point: The length of Ommaya catheter was measured to be directed into the cyst center approximately by using the reformatted CT for the first 4 cases (two giant cysts compromising of right frontal lobe by free hand and two by CT guidance). With advent of stereotactic application, we preferred to target the most dependent portion (13 cases). The rest (2 cases with ventriculomegaly) had a catheter during endoscopic application.

5 cases (26%) were done manually by calculating X, Y, Z of the target point (the most dependent part of the cyst) on CT console and our team work fashioned fixed templates (to avoid any personal calculation errors) based on the Cartesian coordinates of the geometric frame center is 100,100,100 and the coordinates of the right, upper, posterior point of the frame is 0, 0, 0 (Figure III). The entry point was chosen by convention (A transfrontal approach with an entry point 2 to 3 cm off midline at the coronal suture).

8 cases (42%) were managed by computerized stereotactic technique. Geometric space registration of both entry and target points by SurgiPlan software that can easily determine entry point by software calculating both ring and arc angles, target point by calculating X, Y, Z and distance between entry and target points, cyst volume (software calculation). Moreover, visual path that passes through the shortest appropriate trajectory to the cyst floor, avoid superficial cortical vessel, deep cerebral veins, avoid ventriciles (if possible) and maintain the trajectory within neuraxis on targeting brain stem cyst on reformatted coronal CT or MRI. Virtual path can be tried on the station prior to surgery (SurgiPlan Software – Elekta).

After appropriate planning, the patient was taken to the operating room for the procedure. Under general anesthesia for kids and local anesthesia for adults, a small skin flap and twist-drill hole is performed at entry point which is predetermined by Leksell SurgiPlan workstation or by convention for the manual use (A precoronal burr hole was used for transfrontal approach into the cyst). A barium-impregnated ventricular catheter with an outer diameter of 2.1 mm and the appropriate length predetermined on the software was used to cannulate all cysts. The stylet accompanying the ventricular catheter was used in place of a standard biopsy probe. No modifications were made to the stereotactic system. A standard guide block and tube were used to direct the catheter into its target. The catheter was then inserted directly through the guide tube to the predetermined target distance. Ommaya Reservoir was inserted and connected to the catheter without right-angle connector. The reservoir was then anchored to the edge of the drill hole with a silk tie in the subgaleal space. Perioperative stress prophylaxis with hydrocortisone and optional single-shot antibiotic are applied. Following the procedure (for CP) the liquid balance is documented in order to detect a temporary central diabetes insipidus, which would require Desmopressin substitution.

Catheter Insertion: The standard ventricular catheters typically have holes starting 3 mm behind the tip, and continuing as far back as 18 mm from the tip, so that the cyst needs to be at least 2 cm in diameter to allow all the holes to be within the cyst. Therefore, we shifted from targeting the cyst center to the cyst floor to keep the holes contained inside the cyst to keep optimum function of the reservoir especially after cyst collapse.

Cyst Aspiration: Careful intraoperative aspiration of the cyst content as well as postoperative CT scans obtained to ensure accurate position of catheter and adequate cyst decompression. The amount of fluid removed based on the symptoms of the patient and the size of the cyst. We stopped the cyst drainage when more than 15 mL was removed or when the patient had headache, thereby avoiding the complete emptying of the cyst and subsequent traction injury to the surrounding eloquent structures. Cytological examination of the cystic fluid to reconfirm the initial diagnosis by presence of cholesterol crystals in the cyst fluid of craniopharingioma group. Periodical aspiration of the cyst content is performed meticulously through the Ommaya reservoir based on clinical and radiological justification (symptomatic chiasmatic compression, intracranial hypertension or third ventricle amputation).

Postoperative Radiotherapy: All patients received fractioned radiotherapy, delivered in 25-30 fractions and at a median time period of 40 days after surgery. The median total dose of radiotherapy was 5000 cGy (range 4000-5400 in all patients). It was postponed for 4 craniopharingioma and 2 cystic glioma kids under 3 years old.

Follow up: Along 36 months, every patient was followed up at a regular 3-month interval. Assessment included clinical, neurological, fundus examination, endocrinal and functional performance in addition to lab investigations for pituitary functions. CT scan and/or MRI to detect any cyst refill.
**Figure I**: Stereotactic application of Leksell G for lady with recurrent craniopharyngioma after surgery and radiotherapy.

**Figure II**: Calculation of X, Y, Z coordinates (the most dependent part of recurrent craniopharyngioma cyst) by CT and Leksell G fiducials (Manual stereotactic application).
RESULTS

19 cases of deeply seated midline recurrent cysts usually unreachable by the conventional surgery. Managed by Ommaya Reservoir and radiotherapy, 13 (68%) of them were craniopharyngiomas and 6 (32%) were cystic gliomas. Clinical and radiological improvement was achieved in 68% of the cases.

Morphological Results (Both Groups): All cysts showed adequate degree of evacuation of their contents with subsequent cyst reduction except 2 (10.5%) cases that had reasonable residual one in each group. The patients were classified according to volume reduction into Group A: Five cases (26%) showed substantial reduction in which size reduction was > 75%. 4 of them within the craniopharyngioma group and one case was large thalamic cyst. Group B: Eight cases (42%) showed considerable reduction in which size reduction was 50%-75%. Four cases within the craniopharyngioma and 4 cases within the glioma. Group C: Four cases (21%) showed fair response. All were within the craniopharingioma group in which size reduction was 25%-50%. Group D: Two cases (10.5%) showed poor response (1 in each group) with size reduction <25%. Two cases (10.5%) showed cyst re-expansion after malpositioning of the catheter and subsequent reservoir dysfunction.

7 out of 9 cases (78%) that had ventricular dilation got smaller in the first follow up CT (3-6 months after the procedure) without the need for shunt placement. 6 cases (32%) inserted with Ommaya catheter non-
stereotactically, 4 of them required stereotactic re-insertion.

The Cranipharyngioma Group included 8 (42%) children (up to 16 years), 4 of them (21%) were younger than 3 years, and 5 (26%) adults (more than 16 years). 8 cases (42%) were male and 5 cases (26%) were female. On preoperative MRI study, the majority of the cases (10 cases) were of moderate to large size and 3 cases were giant according to Yazargil et al, classification. According to sami classification: Five cases (26%) at grade V, 5 cases (26%) at grade IV and 3 cases (16%) at grade III. All cases proved as craniopharyngioma after first surgery by two neuropathologist and reconfirmed on recurrence. All cases had postoperative radiotherapy except those who were younger than 3 years (4 cases). Amount of aspirated fluid ranged from 5 to 15 mls per session for 2-4 times per year.

The Non-craniopharyngioma Group: Six cases (32%) include 4 cases were males and 2 cases were females. Two cases were under 3 years. Regarding the location, 3 cysts were brain stem (pontine, pontomedullary and pontomesencephalic), 2 cysts were purely thalamic and one large cyst was mesodiencephalic. Regarding the histopathology, 3 cysts were astrocytoma grade II, 2 cysts were Juvenile Pilocytic Astrocytoma (JPA) and one post-irradiation pontine cyst. The range of maximum diameter was 20-45 mm (Table III). Amount of aspirated fluid ranged from 10 to 20 mls per session for 3-5 per year.

The Overall Functional Results (Neurological, visual and endocrine function): At admission, 8 of 19 patients experienced deficiencies of visual function. In 11 of 19 patients, impairment of endocrine function was diagnosed. Four of the latter patients had additional visual deficits. Other tumor-related symptoms were cognitive disturbances (3 patients) and hemiparesis (3 patients). Visual function improved in 6 of 12 patients and remained stable in the rest of the patient at the end of the follow up period. The preoperative status did not change in 6 patients with endocrine dysfunction. In 2 of 3 cases had cognitive disturbances, showed improvement after 2 years. Hemiparesis improved significantly in 2 of 3 patients with physiotherapy. 2 kids (10.5%) with unstable gait improved within 9 months. 4 lethargic patients (21%) improved and became alert.

Catheter Related Complications: In one patient who had catheter infection, the catheter was removed and re-inserted after 4 weeks. Another patient had asymptomatic intracystic silent hemorrhage that washed through Ommaya Reservoir by regular saline. One case operated by manual stereotactic technique with small cortical hemorrhage presented with seizure because of the injury of one of the cortical vessel and the patient controlled conservatively and discharged in health. Rapid collapse of 2 large cysts (one in each group) required catheter repositioning after the cyst refill.

Figure IV: A 35-year-old female with recurrent cystic cranipharyngioma managed by Ommaya reservoir insertion and radiotherapy (follow up period 35 months).
Table I: Patient characteristics among craniopharyngioma group (13 cases):

<table>
<thead>
<tr>
<th>Total No. of Patients</th>
<th>13</th>
<th>100%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>8</td>
<td>61.5%</td>
</tr>
<tr>
<td>Female</td>
<td>5</td>
<td>38.5%</td>
</tr>
<tr>
<td>Mean age (years)</td>
<td>16</td>
<td></td>
</tr>
<tr>
<td>Range (Age)</td>
<td>18m-43y</td>
<td></td>
</tr>
<tr>
<td>Adults (&gt;16 years)</td>
<td>5</td>
<td>38.5%</td>
</tr>
<tr>
<td>Children (&lt;16 years)</td>
<td>8</td>
<td>61.5%</td>
</tr>
<tr>
<td>Initial Symptoms</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Headache</td>
<td>12</td>
<td>92%</td>
</tr>
<tr>
<td>Nausea-Vomiting</td>
<td>8</td>
<td>61.5%</td>
</tr>
<tr>
<td>Seizures</td>
<td>6</td>
<td>46%</td>
</tr>
<tr>
<td>Unsteadiness</td>
<td>2</td>
<td>15%</td>
</tr>
<tr>
<td>Hemiparesis</td>
<td>3</td>
<td>23%</td>
</tr>
<tr>
<td>Visual Symptoms</td>
<td>12</td>
<td>92%</td>
</tr>
<tr>
<td>Impaired Consciousness</td>
<td>4</td>
<td>30%</td>
</tr>
<tr>
<td>Cognitive Disturbance</td>
<td>3</td>
<td>23%</td>
</tr>
<tr>
<td>Amenorrhea</td>
<td>2</td>
<td>15%</td>
</tr>
<tr>
<td>Obesity</td>
<td>4</td>
<td>30%</td>
</tr>
<tr>
<td>Growth Retardation</td>
<td>2</td>
<td>15%</td>
</tr>
</tbody>
</table>

Table II: Response of recurrent craniopharyngioma cyst to Ommaya reservoir and subsequent radiotherapy (13 cases):

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Previous Surgery (S)</th>
<th>RTX</th>
<th>(Maximum Size)</th>
<th>Initial Size</th>
<th>Minimum Size</th>
<th>% Volume Reduction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>35 y F</td>
<td>Combined</td>
<td>RTX</td>
<td>44.22 cm³</td>
<td>3.68 cm³</td>
<td>91.7%</td>
<td></td>
</tr>
<tr>
<td>Case 2</td>
<td>1.5 y M</td>
<td>Bi-frontal</td>
<td>OR</td>
<td>36.14</td>
<td>9.81</td>
<td>73%</td>
<td></td>
</tr>
<tr>
<td>Case 3</td>
<td>6 y F</td>
<td>RT. FT</td>
<td>RTX</td>
<td>94.56</td>
<td>8.22</td>
<td>91.3%</td>
<td></td>
</tr>
<tr>
<td>Case 4</td>
<td>3.5 y F</td>
<td>Bi-frontal</td>
<td>RTX</td>
<td>12.88</td>
<td>2.88</td>
<td>77%</td>
<td></td>
</tr>
<tr>
<td>Case 5</td>
<td>18 y M</td>
<td>TS</td>
<td>RTX</td>
<td>25.82</td>
<td>4.8</td>
<td>81.4%</td>
<td></td>
</tr>
<tr>
<td>Case 6</td>
<td>2.5 y M</td>
<td>Endoscopy</td>
<td>OR</td>
<td>22.62</td>
<td>8.6</td>
<td>62%</td>
<td></td>
</tr>
<tr>
<td>Case 7</td>
<td>43 y M</td>
<td>TS</td>
<td>RTX</td>
<td>12.82</td>
<td>4.72</td>
<td>63%</td>
<td></td>
</tr>
<tr>
<td>Case 8</td>
<td>3y M</td>
<td>Lt. FT</td>
<td>OR</td>
<td>34.44</td>
<td>27.55</td>
<td>20%</td>
<td></td>
</tr>
<tr>
<td>Case 9</td>
<td>40y F</td>
<td>Bi-frontal</td>
<td>OR</td>
<td>26.42</td>
<td>11.89</td>
<td>55%</td>
<td></td>
</tr>
<tr>
<td>Case 10</td>
<td>14y M</td>
<td>Rt. FT</td>
<td>RTX</td>
<td>18.46</td>
<td>11</td>
<td>40%</td>
<td></td>
</tr>
<tr>
<td>Case 11</td>
<td>33y M</td>
<td>TS</td>
<td>RTX</td>
<td>38</td>
<td>26.6</td>
<td>30%</td>
<td></td>
</tr>
<tr>
<td>Case 12</td>
<td>2y F</td>
<td>Bi-frontal</td>
<td>OR</td>
<td>18.22</td>
<td>9.5</td>
<td>48%</td>
<td></td>
</tr>
<tr>
<td>Case 13</td>
<td>8y M</td>
<td>Rt. FT</td>
<td>RTX</td>
<td>16.8</td>
<td>9.24</td>
<td>45%</td>
<td></td>
</tr>
</tbody>
</table>

Rt. (Right), Lt. (Left), FT.(Frontotemporal), TS.(Trans-spheniodal),OR (Ommaya Reservoir), RTX (Radiotherapy)
Table III: Glioma Group (6 Cases):

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Location</th>
<th>Surgery (S)</th>
<th>Histopathology</th>
<th>Recurrence Duration</th>
<th>OR±RTX</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>19M – F</td>
<td>Ponto-medullary junction (20 mm)</td>
<td>Biopsy</td>
<td>Astrocytoma II</td>
<td>4M</td>
<td>OR inserted and periodic decompression until age 3y when radiotherapy launched</td>
</tr>
<tr>
<td>2</td>
<td>6Y- Male</td>
<td>Right thalamic cystic mass filling lateral ventricle (45 mm)</td>
<td>Biopsy of the enhanced fleshy portion</td>
<td>Astrocytoma II</td>
<td>13M</td>
<td>OR with immediate RTX</td>
</tr>
<tr>
<td>3</td>
<td>20M- Male</td>
<td>Large right mesodiencephalic cyst (25 mm)</td>
<td>Biopsy + Cyst aspiration</td>
<td>JPA</td>
<td>6M</td>
<td>OR inserted and periodic decompression until age 3y when radiotherapy launched</td>
</tr>
<tr>
<td>4</td>
<td>5Y- Male</td>
<td>Large cyst with small nodular mass spanning dorsal part of the midbrain and pons (30 mm)</td>
<td>Biopsy of the enhancing nodule with cyst aspiration</td>
<td>Astrocytoma II</td>
<td>3M</td>
<td>OR with immediate RTX</td>
</tr>
<tr>
<td>5</td>
<td>4Y- Male</td>
<td>Large left thalamic cystic mass (40 mm)</td>
<td>Stereotactic biopsy and cyst aspiration</td>
<td>JPA</td>
<td>4M</td>
<td>OR with immediate RTX</td>
</tr>
<tr>
<td>6</td>
<td>18Y-F</td>
<td>Posterior fossa ependymoma</td>
<td>Complete excision + RTX</td>
<td>Ependymoma</td>
<td>2Y – Post RTX- Pontine cyst (20 mm)</td>
<td>OR</td>
</tr>
</tbody>
</table>

OR (Ommaya Reservoir), RTX (Radiotherapy)

DISCUSSION

Microsurgery remains the technique of choice for the treatment of solid tumors, particularly when they are large. Long-term results after multi-modality treatment are not inferior to long-term results obtained by microsurgery. However, when hospital costs are considered, the difference between stereotactic and microsurgical approaches is considerable. Lastly, if we consider morbidity with respect to the treatment of giant cysts, the multi-modal approach should be preferred rather than the microsurgical approach alone.

Many procedures have been used in the treatment of intracranial cystic lesions, mainly percutaneous cyst aspiration, marsupialization, percutaneous ventricle cystostomy with or without plasty, and cyst drainage. In some patients, the procedures were unsuccessful and the cyst increased in size, causing neurological disturbances. Therefore, repeated evacuations that include the use of different methods are necessary.

Craniosphenomeningioma is the most frequent midline intracranial cystic neoplasm of non-glial origin, representing 9% of pediatric brain tumors. In larger series, 54.0 to 94.4% of the evaluated patients had tumors with significant cystic parts. It is important to keep in mind that 90% of pediatric craniopharyngiomas have these characteristics whereas only 10% of craniopharyngiomas are purely solid.

Craniosphenomeningiomas are benign tumors for which the best therapeutic option should be complete resection. However, because of the tumor location, the management of this tumor must be considered differently. In fact, the morbidity associated with radical operation remains high, notably because of adherences to critical surrounding structures such as vascular structures, optic pathways, the pituitary stalk, and hypothalamus. In addition despite radical surgery, recurrences are not rare.

Zuccaro demonstrated a 77% rate of total resection with 87% of 153 patients having a hormone deficit.

According to the literature, the condition in the vast majority of patients will progress to visual and endocrine dysfunction, hypothalamic dysfunction, disorders of hunger, psychiatric disorders, and a poor quality of life.

In the series of Yasargil et al., the overall mortality rate was 17% and the recurrence rate was 7% even after aggressive radical excision of the tumors. In addition, marked differences in the rates of good outcomes (range, 52%-87%), according to the
experience of the surgeons, were reported for the radical surgery-treated groups\(^{(19)}\).

Over the last 40 years there exists an open-ended controversy concerning the best treatment for craniopharyngioma. One group favors open surgery which is in many cases associated with increased morbidity. The other group proposes minimal-invasive procedures combined with subsequent radiotherapy to minimize risk and morbidity\(^{(22)}\).

Due to the high variability in the appearance of these tumors the treatment strategy must be individually tailored to the patient\(^{(22)}\).

In the past decade, the goal of treatment of craniopharyngioma has been not only to obtain long-term tumor control but also to preserve the patient’s quality of life\(^{(8)}\).

Preserving the patient’s quality of life and being the great majority of craniopharyngiomas having significant cystic component have pushed many neurosurgeons to look for more long term conservative technique based on Craniopharingioma is a benign chronic disease.

An insertion of a catheter into a cystic craniopharyngioma may prevail over the transient success of a cyst fenestration by allowing repetitive drainage of the tumor cyst and the opportunity of instillation of intracystic substances. Different neurological techniques are employed for the placement of catheters\(^{(3)}\).

We included in this series the difficult recurrent complicated monocystic craniopharyngioma and glioma that their cystic component more than 60% and this agrees with the definition of cystic craniopharyngioma in the literature that considers a cystic craniopharyngioma to be one in which greater than 60% of the tumor volume is cystic\(^{(33)}\).

In our series, we have had overall successful rate of 68% (>50% reduction of the initial cyst volume) over 36 months of follow up. Thirteen cases (9 cases of craniopharyngioma and 4 cases of glioma) 68% passed 50% cyst volume reduction including 2 cases of radiotherapy received craniopharingioma completely disappeared at the end of the follow-up period. Cavalheiro et al, considered cystic craniopharyngioma to be controlled when a tumor decreased more than 50%\(^{(8)}\). However, the comparison between studies in terms of cystic craniopharyngioma is very difficult because, whereas some authors considered cyst volume stability a good morphological result. Other authors considered that cyst reduction should reach 50% of the initial volume of 9 cases (78%) that had ventriculomegaly improved by cyst decompression without the need of a shunt. However, one patient developed secondary hydrocephalus and was finally shunted. Lena et al, reported 7 children were shunted before surgery and only 3 had permanent shunt\(^{(16)}\).

That means 4 out of her 7 cases were not shunt dependent and may improve by simple cyst decompression. In Cavalheiro series, 6 out of 19 cases with hydrocephalus did not need a shunt\(^{(8)}\).

We found our results in this selected group of recurrent midline cysts were comparable to other modalities. Over 36 months of follow up, vision improved in 6 of 12 cases (50%) and remained stable in the rest. the visual field deteriorated in 3 cases (25%) and remained stationary in the rest of the cases. No improvement in endocrine function in 6 of 11 cases (54.5%) who kept on hormonal support. However, In the group of children, following stereotactic treatment by Michael and Guido, the patients’ vision improved in 61%, remained unchanged in 39%, and the visual field ameliorated in 75% or remained stationary (25%). While 6% experienced temporary minor visual degradation, which recovered within days, there was no permanent visual deficit due to stereotactic surgery. Following cyst drainage the endocrinological state remained unchanged in 93%\(^{(26)}\). Cognitive function and hemiparesis showed improvement in 2 of 3 patients supported with psychotherapy and physiotherapy. Gait improved in two kids.

Neither major clinical deterioration nor mortality reported with this conservative technique during follow up period (36 months). This agrees with large recent series of Michael and Guido who stated that no procedure related lethality or permanent morbidity was observed with 208 patients with craniopharyngioma were performed in the Department of Stereotactic Neurosurgery in Freiburg , Germany From 1990 to 2010\(^{(23)}\).

Rapid decompression of two large non-stereotactically inserted cysts (one in each group) might push the catheter into the subarachnoid space with subsequent malfunction and required repositioning after the cyst refill. We shift from targeting cyst center to the cyst floor that ensured catheter accommodation by cyst and minimized the malpositioning. In our study, 4 out of 6 patients who had surgical implantation had undergone new implantation stereotactically.

The standard ventricular catheters typically have holes starting 3 mm behind the tip and continuing as far back as 18 mm from the tip, so that the cyst needs to be at least 2 cm in diameter to allow all the holes to be within the cyst. Steinbok and Hukin have used a modified catheter containing holes that go back only 8 mm from the tip to make the margin of greater safety\(^{(35)}\). Furthermore to minimize leakage of cyst content into subarachnoid space, they tried to insert the catheter using a push technique with a stylet in place or a minimal incision, if one is using an open approach to visualize the cyst directly\(^{(16)}\).
Stereotactic methodology especially when supported with the software, obviously reduced the catheter malposition and reservoir dysfunction for many reasons: 1) Reduced the incidence of catheter malpositioning; 2) The stereotactic created cyst wall hole is too small to leak; 3) Ideal placement of the tip of the catheter into the most dependent point inside the cyst with respect to its elliptical contour is ensured; 4) Surgical insertion of catheter is not a good idea as surgical disturbance of the peri-cystic adhesions and cisterns. Moreover, normal brain tissue retraction through the surgical corridor predispose to generous cyst wall gapping; 5) it is preferable to use the visual path in penetrating the cyst wall through the most accessible thick part of the cyst wall.

In report of the Canadian experience, Hukin et al, stated that the complications of the non-stereotactic catheter insertion occurred in 7 of 19 patients (37%) and included the acute epidural and intraventricular hemorrhage in a patient with a previously undiagnosed coagulation defect, intracystic blood (2 patients), fluid collection outside the cyst (1 patient), and contrast leakage on postoperative CT scan after instillation of contrast into the cyst (3 patients)\(^{(16)}\). However, Pettorini et al, advocated that neuro-endoscopic positioning of the catheter is a safer option rather than open or stereotactic approaches\(^{(5)}\). Bartels et al, did not find a difference in complications between the different surgical methods (free hand, operative, endoscopic and stereotactic)\(^{(6)}\).

There are anatomic constraints that may limit the stereotactic accessibility of a craniopharyngioma cyst which is related to the infra- or suprasellar location and the vulnerability of the adjacent critical structures such as optic apparatus, pituitary gland, and hypothalamus\(^{(16)}\). However by using the SurgiPlan station and its visual path, we secured these critical structures. Stereotactic neurosurgery provides save, minimal invasive and cost-efficient options in the treatment of childhood craniopharyngioma\(^{(23)}\).

Chronic progressive shrinkage of the cyst might have caused the movement of one or more holes of the ventricular catheter into the subarachnoid space. Therefore, we were intended to push the catheter deep inside the cyst. Steinbok and Hukin used a modified ventricular catheter with holes over 5 mm instead of the regular one over 15 mm to get optimum function of the reservoir.

Perhaps the craniopharingiomas are not tumors themselves but embryonic immune defects. Indeed, few genetic changes are found in craniopharingiomas. The balance between the aggressiveness of the tumor and the patient’s immune response will determine the winner of this battle\(^{(37)}\). This idea pushed some surgeon to try Interferon α as new modality for intracystic injection.

The mechanism of action of Interferon α in tumor control is not definitively known but preliminary studies have suggested that it may be tumoricidal by activating apoptosis with the simultaneous modulation in patient immune response\(^{(17)}\).

In a retrospective study of Schubert and his colleagues compared three groups of 32 children (<18 years of age) with craniopharyngioma. The first group included patients treated with microsurgical resection. The second group underwent stereotactic cyst drainage, implantation of a Rickham catheter, and fractionated three-dimensional conformal multi-field radiotherapy with 54 Gy volume dose. The third group received various combined approaches. In this study, the 8.5-years of freedom from tumor recurrence was 24% in the resection group as compared to 71% for children with combined stereotactic and radio-therapeutic treatment \((p = 0.05)\). The target volume can be significantly reduced by the evacuation and drainage of large cysts preceding fractionated external radiation therapy. There was no permanent postoperative morbidity related to stereotactic cyst puncture and drainage\(^{(34)}\).

Because limited surgery does not prevent recurrences and radical surgery carries unacceptable morbidity and mortality, postoperative external-beam radiotherapy has been added to limited surgery in an effort to improve local control. The literature seems to support this approach, with a reported long-term control of approximately 80-95% at 5-20 years and a low risk of long-term morbidity\(^{(36)}\). Nowadays, craniopharyngioma must be considered a complex molecular disease, and a detailed explanation of the mechanisms underlying its aggressive biological and clinical behavior, despite some benign pathological features, would be the first step toward defining the best management of craniopharyngioma. Indeed, advances in the knowledge of the molecular mechanisms at the base of craniopharyngioma oncogenesis will lead to comprehension of the critical checkpoints involved in neoplastic transformation. The final research target will be the definition of new biological agents able to reverse the neoplastic process by acting on these critical checkpoints\(^{(16)}\).

Cystic lesions of the brain stem often have substantial mass effect and may be responsible for neurological deterioration in patient. Drainage of the cystic lesion often leads to neurological improvement\(^{(1)}\). Surgical resection of cystic lesions of the brain stem carries substantial risks; often gross total resection is not possible and may lead to recurrence. Even with the gross total resection, cysts may recur\(^{(20)}\). Stereotactic aspiration of cystic lesions of the brain stem is an accurate and safe method that carries low surgical risks\(^{(41)}\).
Cyst recurrence after stereotactic aspiration is common and often requires multiple procedures to control cyst growth. Brain stem cystic regression is a gradual process and can take from 3 to 12 months\(^{(30)}\).

In report of Rogers and Barnett, who presented 21 deeply seated brain cysts including 2 cases of craniopharingioma and one case of brain stem cystic glioma, they stated that stereotactic insertion of Ommaya Reservoir system did achieve cyst control at follow up period of 4 to 114 weeks\(^{(30)}\).

In our series, we did achieve cyst control at relatively long follow up period (36 months). Moreover, in 2 cases under 3 years, we carried out periodical aspiration through Ommaya Reservoir till the appropriate age for of radiotherapy without neurological deterioration.

The hyperfractionated radiotherapy be used to reduce the possibility of radiation injury to normal surrounding brain in the treatment of pediatric brain stem glioma. However, a possibility remains that, even with this, intratumoral radiation injury can occur and cause clinical deterioration\(^{(13)}\).

We believe that Periodical decompression of brain stem and diencephalic cysts and their low grade histopathological features without malignant transformation together with appropriate radiotherapy helped make our patients stable over relatively long follow up period.

**CONCLUSION**

Because limited surgery for midline deeply seated recurrent brain cyst does not prevent recurrences and radical surgery carries unacceptable morbidity and mortality, postoperative external-beam radiotherapy has been added to limited surgery in an effort to improve local control. Children younger than 3 years may not be candidates for such radiotherapy because they can develop unusually severe long-term adverse effects. In those patients, stereotactic implantation of an intracystic catheter with Ommaya reservoir may be a valuable alternative treatment option. The benefits of this procedure include temporary relief of fluid pressure by serial drainage, may prolong the interval to or obviate the need for radiation.

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