

Case Series:

Purely Neuro-endoscopic Transventricular Approach for Cystic Craniopharyngiomas



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ABSTRACT

Background and Importance: Craniopharyngiomas are tumors made up of mixed components which can present intraventricular cystic portion. The aim of our work is to evaluate the outcome of the endoscopic marsupialization as a surgical approach.

Case Presentation: We report 11 cases presenting craniopharyngioma with intraventricular cystic portion inducing hydrocephalus managed at Neurosurgery Unit of Fann Hospital between June 2013 and June 2017. Endoscopic marsupialization was realized for all patients with a rigid neuroendoscope Karl Storz.

Conclusion: The mean age of cases was 30.18 years with a ranged 07-69. There was a male predominance with a sex ratio of 1.75. All patients were suffering from an intracranial pressure syndrome. Lowering visual acuity including two (02) blindness cases was found in patients under 15 years. Frontal lobe syndrome was found in all patients of more than 50 years. A cerebral CT scan was realized for 9 patients and an MRI for 4 patients. A cystic marsupialization with biopsy was realized by precoronal approach. Fluid looked like "waste oil" for 9 patients. Ventriculocisternostomy of the 3rd ventricle was realized in 2 cases. Progress was favorable with intracranial pressure signs disappearance in 8 cases. We noticed 3 failures with cyst persistence at control CT Scan. Three patients had a recurrence, requiring revised marsupialization complicated by death in 1 case. Endoscopic marsupialization represents a seductive technique which is an easy and reproducible treatment for intraventricular cystic craniopharyngioma.

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Highlights

- We report 11 cases with patients presenting craniopharyngioma with intraventricular cystic portion inducing hydrocephalus.
- A cysto-ventriculostomy was performed in 9 patients and additional ETV (Endoscopic Third Ventriculocisternostomy) in 2 cases.
- The global outcomes were good in the 9 patients.
- It has advantages especially in practical conditions which do not always provide adequate safety conditions for complex microsurgical approaches.

Plain Language Summary

Craniopharyngiomas are benign intracranial tumors which mainly occur in intrasellar and especially suprasellar topography in 60% of cases. Total microsurgical excision remains the reference treatment. However, the intimate relationship with delicate neurovascular structures suggests a less aggressive treatment. We report 11 cases of patients who presented a craniopharyngioma with an intraventricular cyst complicated by hydrocephalus. An endoscopic intraventricular marsupialization of the cyst was performed in all patients. We did not note any intra-operative incident. The outcome was favorable with regression of signs of intracranial hypertension in all patients, and good results in 9 patients in the medium term. The endoscopic transventricular approach for cystic craniopharyngioma is a safe and effective alternative to microsurgery in the short and medium term.

1. Background and Importance



Only 10% of craniopharyngiomas are totally solid, while more than half are purely or predominantly cystic [1, 2]. Their development in the 3rd ventricle is inevitably responsible for biventricular hydrocephalus.

From these cases, the treatment of hydrocephalus becomes the immediate objective because it directly threatens the life of the patient. In such cases, control of mass effect may represent a suitable alternative by neuroendoscopic trans-ventricular approach. It restores the CSF (Cerebrospinal Fluid) pathways and lifts compression on the hypothalamus. Moreover, from an economic point of view and in our working conditions, this method has multiple advantages. In fact, radical surgery can in many cases be complicated by hormonal deficits that require supplementation for life and the cost of drugs is most often difficult to bear for patients with modest financial incomes. Also, aggressive methods necessarily require well-resourced resuscitation that most countries in sub-Saharan Africa do not have.

The aim of this work is to show the results of trans-ventricular endoscopic treatment of cystic craniopharyngiomas as an alternative to microsurgical approach through a consecutive series of 11 patients.

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2. Case Presentation

Patients

We performed a retrospective review of all surgeries for craniopharyngiomas between October 2007 and October 2017. Eleven patients underwent purely endoscopic treatment for cystic craniopharyngiomas. The decision to use this technique was made after reviewing pre-operative imaging. Pre- and post-operative clinical, radiographic, and biological data including age, gender, presenting symptoms, tumor grade using Yasargil's classification, the existence or non-existence of prior treatment were obtained and reviewed. The outcomes were assessed by clinical examination post-operatively. The average follow-up duration was 11 months.

Endoscopic system and instrumentation

We used a rigid endoscope with 30 degrees view angle, a perforator, a suction tip, Foragy balloon, grasping forceps, and scissors.

Surgical technique

The patient was placed supine and the head was elevated 20-30 degrees. An incision was made and a burr hole was done 1 cm in front of the coronal suture on the mid-pupillary line (side depending on the cyst location).

The endoscope was introduced in the frontal horn of the lateral ventricle. The cyst was easily located at the entrance to the foramen of Monro. We punctured it with a needle and aspirated the contents avoiding rupture in the ventricle as much as possible. Once the cystic fluid well decreased, we washed the inside of the cyst with a Ringer's lactate solution until a significant dilution of the contents was observed. Then we coagulated the wall of the cyst before opening it widely with scissors or forceps. After this step, we introduced the endoscope into the cyst and tried to make a stomy of the floor of the cyst with the basal cisterns. If anatomy allowed it, we completed the procedure with an ETV (Endoscopic Third Ventriculocisternostomy) in the interpeduncular cistern. We finished by washing with the Ringer's lactate solution to avoid chemical meningitis. We checked the absence of active bleeding and removed the endoscope (Figure 1 shows intra-operative view of 1 case).

3. Results

There were 7 male patients versus 4 females, with a sex ratio of 1.75. The mean age of our patients was 30.18 years, with extremes ranging from 05 years to 69 years.

Clinically, they all had Increased Intracranial Pressure (ICP) syndrome due to hydrocephalus. All patients under the age of 18 years had a decrease in visual acuity, including 2 cases of blindness. An endocrine syndrome was present in 2 patients aged 13 years and 17 years. Neuropsychologic and psychiatric disorders were found in 3 patients, all aged over 5 years.

A cerebral Computed Tomography (CT) was performed in 9 patients and complementary cerebral Magnetic Resonance Imaging (MRI) in 4 patients. In terms of imaging, in all patients, the cystic lesion was located extra and intraventricular (third ventricle) and could be classified as "D" according to Yasargil et al. classification (Figure 2).

The precoronal route was used and found an obstruction of the foramen of Monro by the cyst in all cases. The classical brownish appearance of the cystic fluid was found in 9 cases. A cysto-ventriculostomy was performed in 9 patients and additional ETV in 2 cases. There were no intra-operative incidents. We had 3 cases of post-operative chemical meningitis treated with corticosteroid. The global outcomes were good in the 9 patients available for follow-up with a regression of the ICP in all patients, improvement of visual acuity in 2 cases, and regression of the frontal syndrome and psychiatric disorders in 3 cases. The early control CT showed a clear regression of cyst and hydrocephalus in 7 cases.

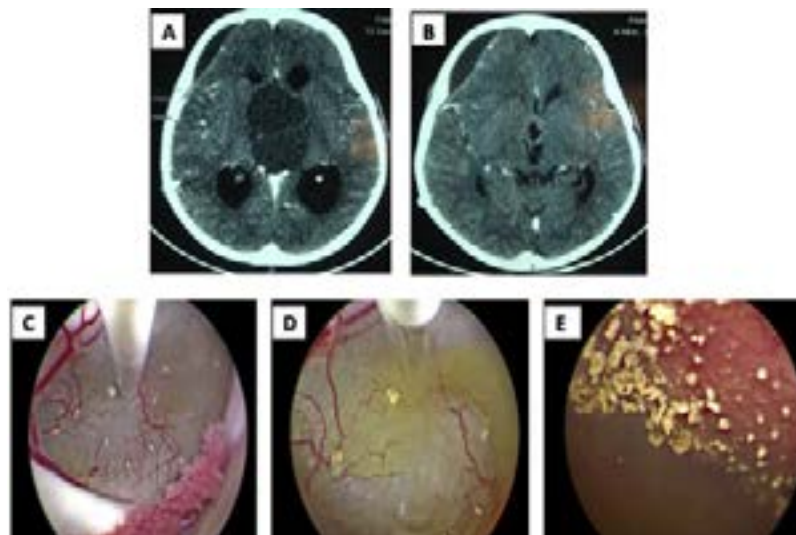


Figure 1. Pre-, post- and intra-operative images from case 1

A: CT demonstrating a huge predominantly cystic craniopharyngioma, developed into the 3rd ventricle responsible for biventricular hydrocephalus; B: Post-operative CT showing a little cyst remnant and total regression of hydrocephalus; C, D: Intraoperative view showing the cyst into the 3rd ventricle being punctured (note the yellow cyst fluid flowing into the ventricle); E: Intracystic view after huge opening of the cyst wall, note the presence of micro calcium fragments.

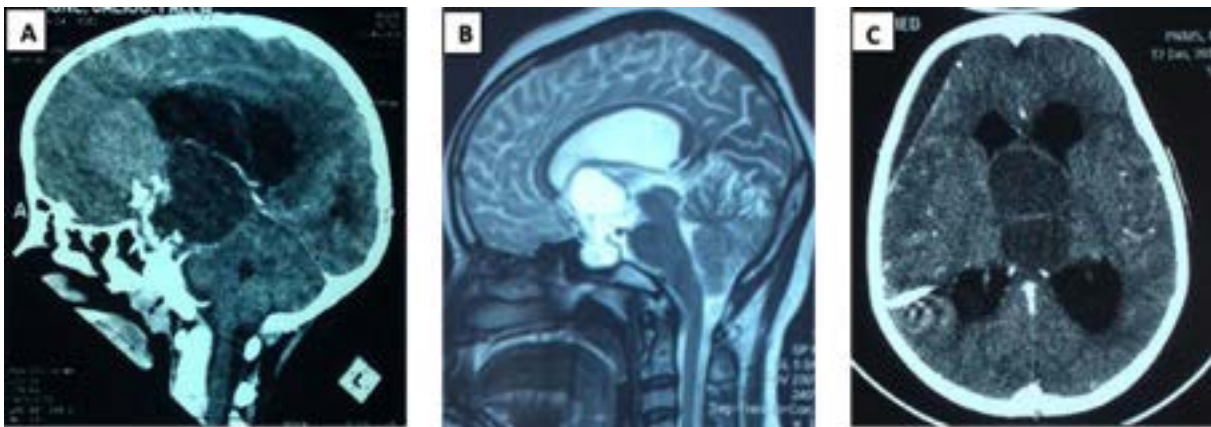


Figure 2. Pre-operative imaging (type “d” of Yasargil)



A: A 12-year-old patient with mixed craniopharyngioma with predominant and compressive cystic portion; B: A 32-year-old patient with mixed craniopharyngioma with predominant and compressive cystic portion; C: A 7-year-old patient with a large intra- and extra-ventricular cyst who initially had a microsurgical approach followed by Ventriculo Peritoneal Shunt (VPS) in the presence of hydrocephalus.

A failure of marsupialization was noted in 3 cases with persistent cyst imaging. Surveillance was retained in a patient with regression of Increased ICP signs, a Pterional surgery was secondarily performed in 1 case, and the 3rd patient was lost to follow-up.

Three patients (Table 1; cases 7, 8, 9) underwent a second procedure for cyst recurrence respectively 4, 10, and 8 months after the first operation. They all were re-operated on with good results in 2 cases (Figure 3). The 3rd patient died 4 days after operation who had waking delay with protuberant hematoma at the control CT.

4. Discussion

Craniopharyngiomas comprise 5.6% to 15% of all intracranial tumors among children, mostly occurring be-

tween 5 to 14 years of age. Direct surgical approaches on craniopharyngiomas have extremely developed within the last half-century. Surgical mortality has reduced from 41% before replacement therapy and became available to 2% or less in recent series [3]. Endocrinological sequelae are the rule and 80% of Yasargil’s patients needed substitution therapy [4].

However, these results can only be conceived in certain centers with recognized expertise and wide experience in the treatment of this pathology. In sub-Saharan Africa, it is very often difficult to meet all the conditions for an optimal microsurgical resection of these lesions. Furthermore, replacement therapy can be problematic for financially poor patients. Therefore, less invasive solutions should be favored to avoid the complications inherent in microsurgery.

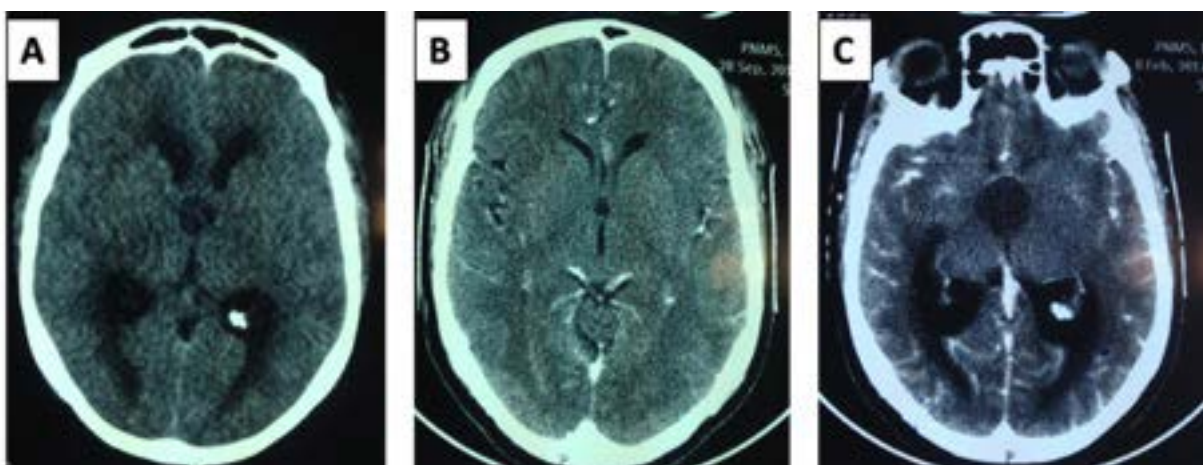


Figure 3. Recurrence 8 months after surgery in a 50-year-old patient with good initial control



A: Initial CT-scan; B: Good control post-operative to M3; C: Recurrence to M8.

Table 1. Patients' characteristics

Case	Age/Gender	Previous Treatment	Clinical Features	Surgery	Outcomes (Follow-up)
1	5 y/M	Partial removal/ pterional approach; calcified subdural hematoma complicating a VPS	Inc ICP, Visual impairment	Cyst opening in ventricles	Partial visual recovery, ICP normalized (2 years)
2	9 y/F	No	Visual impairment, Inc ICP	Cyst opening in ventricles	ICP normalized, Lost after 3 months
3	12y/M	No	Visual impairment, Inc ICP, Loss of consciousness (GCS 14)	Cyst opening in ventricles and basal cisterns, ETV	ICP normalized, total visual recovery (5 months)
4	13 y/M	VPS	Inc ICP, blindness	Cyst opening in ventricles	No visual recovery, ICP normalized (7 months)
5	17 y/F	No	Inc ICP, Blindness, Dwarfism	Cyst opening in ventricles	Failure Pterional surgery
6	21 y/M	No	Inc ICP	Cyst opening in ventricles and basal cisterns, ETV	ICP normalized (4 years)
7	34 y/F	No	Loss of consciousness, Seizure	Cyst opening in ventricles	Regression of symptoms (4 months). Recurrence
8	40 y/M	Pterional surgery	Inc ICP, Blindness, focal deficit	Cyst opening in ventricles	Regression of symptoms (10 months). Recurrence
9	50 y/M	No	Psychiatric disorders	Cyst opening in ventricles	Regression of symptoms (8 months). Recurrence
10	62 y/F	No	Frontal syndrome	Cyst opening in ventricles	Regression of frontal syndrome (9 months)
11	69/M	No	Memory disorders	Cyst opening in ventricles	Regression of symptoms (13 months)



GCS: Glasgow Coma Scale; Inc ICP: Increased Intracranial Pressure; ETV: Endoscopic Third ventriculocisternostomy; VPS: Ventriculoperitoneal Shunt.

A predominantly cystic lesion characterizes 60% of craniopharyngiomas [4]. When mass effect can cause symptoms, cyst drainage is an acceptable compromise for most of the patients.

In the last 2 decades, neuroendoscopy has been increasingly used in the treatment of hydrocephalus and also intracranial cystic lesions. Cysts growing into the ventricular system (Yasargil's C to F types) [5] are appropriate for an endoscopic approach. This procedure has been widely used by many [5, 6] and has been proven to be safe, effective, and repeatable. This technique is well standardized and does not require complex instrumentation. In our experience, we have been using the endoscope for about 10 years. As a result, our experience makes it easy to practice this procedure safely. It

has 4 essential steps: catheterization of the lateral ventricle, puncture and emptying of the cyst, coagulation and wide opening of the wall, and abundant washing with Ringer's lactate solution. If the anatomy allows it we complete with a cystostomy in the skull base cisterns. We had no intra-operative complications. This procedure, even if it is oncologically palliative, allows normalizing the ICP with a control of the mass effect in more or less long term. During follow-up, 3 patients in our series presented a recurrence that required revision surgery. Cysto-ventriculostomy, as proposed by Spaziante et al. [2] discusses about an evolution in the treatment of intraventricular cystic lesions: the authors explained the concept of marsupialization of the cyst into the ventricles, focusing on continuous dilution of

the cyst's fluid and resorption through the CSF pathways [7]. Clinical improvements in all patients were remarkable.

Other less invasive options can be proposed in the treatment of cystic craniopharyngiomas. The mostly used one is intracavitary irradiation [8, 9]. The management options include the stereotactic placement of a catheter to allow repeated aspiration and furthermore intracystic irradiation with radionuclide (32P or 90Y) [10]. Although no controlled comparative studies have been performed so far, the latter approach seems to have the highest chance of local control with minimal morbidity, being the favored treatment in many countries [11]. This technique could be indicated in our patients but the radioisotopes are not available in our country (Senegal).

5. Conclusion

The endoscopic transventricular approach for cystic craniopharyngioma is a safe and effective alternative to microsurgery in the short and medium term. It has advantages especially in practice conditions which do not always provide adequate safety conditions for complex microsurgical approaches. Senegal is a country where the majority of patients have limited financial resources and access to care. This technique has the advantage of offering patient care even if the technical platform is still limited. It is a reproducible and effective surgical technique especially when the intervention is controlled, without major increase in morbidity during the re-exploration. However, it should be remembered that this treatment is palliative on the oncological level and must always be part of a multimodal approach whenever possible.

Limitations

Our work has certain limitations. This is a retrospective study with a small sample of the series due to a selection bias linked to the absence of digitized patient records. Also, the possibility of a surgical revision in the medium term should be noted because it is a non-radical technique.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this research.

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Authors' contributions

Conception and design: Sagar Diop, Alioune Badara Thiam, Maguette Mbaye; Data collection: Sagar Diop; Data analysis and interpretation: Sagar Diop, El Hadji Cheikh Ndiaye Sy; Drafting the article: Sagar Diop, El Hadji Cheikh Ndiaye Sy; Critically revising the article: Mbaye Thioub; Reviewing submitted version of manuscript: Mouhamed Abdoulaye Cisse, Momar Codé Ba; Approving the final version of the manuscript: Seydou Boubakar Badiane.

Conflict of interest

The authors declared no conflict of interest.

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