

## Surgical Experience with Lateral Ventricular Choroid Plexus Papilloma in Children

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### Abstract

**Background:** Surgical experience with lateral ventricular choroid plexus papilloma in children introduction choroid plexus papilloma is the most common lateral ventricular tumor in the children with frequent location in the atrium surgical access to the lateral ventricle remains one of the challenging procedures in the neurosurgery because of the deep location and relation to vital structures especially eloquent area of the brain. In this study, the transcortical microscopic surgery was used to reach the tumor and identify its vascular and structural relation with minimal cortical incision. Endoscopic assisted microscopic surgery was done in the last two cases.

**Aim of Study:** To understand and learn more about one of the most challenging brain tumors (lateral ventricular choroid plexus papilloma) in children and to overcome the difficulties and complications in treating such as these kind of brain tumors to improve the quality of life in these children.

**Patients and Methods:** The seven cases of trigonal location were operated using posterior parietal lobule incision in the dominant side or small tumors in nondominant side and parietotemporal junction incision in large tumors stretching the cortex in nondominant side.

The case located in the frontal horn of lateral ventricle was operated by transcortical frontal incision Endoscopic access to the ventricle and navigation of the tumor were done through small incision which was followed by microscopic removal were done in the last two cases.

**Results:** Total removal was done in six cases and in the remaining two another surgical setting were needed to achieve total removal two cases had postoperative visual deficit and one case transient motor deficit and one case had transient sensory aphasia, endoscopic assisted approach provide short and safe way to the atrium with excellent identification of tumor extension, vascular pedicle and relation and also can help and check total removal.

**Conclusion:** Choroid plexus papilloma is uncommon tumor that need special surgical plane and knowledge. Endoscopic assisted access to the lateral ventricle enhances safe

and total microscopic removal of this vascular tumor through small cortical.

**Key Words:** *Choroid plexus papilloma – Lateral ventricular tumors – Atrium of the lateral ventricle.*

### Introduction

**CHOROID** plexus papilloma (CPT) is neuroepithelial tumors the constitute one of the commonest lateral ventricular neoplasms in children.

Generally, it accounts for about 1% of all brain tumors and 2-6% around of pediatric brain age group tumors [1-3].

This fragile and vascular neoplasm usually reaches large size before causing any manifestations; which usually result for increase intracranial tension rather than compression of adjacent neural structures [4]. CPTs occur usually in the atrium of the lateral ventricle or less commonly originating from the atrium and extend to frontal or occipital horns [2].

Surgical access to this tumor represents special importance for any pediatric neurosurgeon not only because of the deep location which is the same for all intra-ventricular [2,5,6] but also because the vascular nature of this tumor which aid more challenge of pediatric age group [7-9].

In this study the trans-cortical microscopic surgery was used to reach the tumor and identify its vascular and structural relationship. The clinical, radiological and surgical results are analyzed.

### Patients and Methods

Eight cases with choroid plexus papilloma of the lateral ventricle were operated between Jan. 2006 – Jan. 2015.

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All cases were subjected to preoperative clinical evaluation, full laboratory and Radiological investigation in the form of MRI (magnetic resonance imaging) brain with and without contrast with or without CT (computed tomography) brain with contrast. Median follow-up period was 18 months.

#### *Operative technique:*

There should be cooperation with special anesthetic team specialized in pediatric neurosurgery that should be aware about the vascularity nature of this type of tumors and hence accurate replacement with crystalloid, fresh frozen plasma and blood can be done.

#### *Proper surgical approach:*

Choroid plexus papilloma occupying the atrium of the lateral ventricle are approached using transcortical approach that performed in parietotemporal Junction or posterior parietal or posterior parietal parietotemporal variety is used in large tumors stretching the angular and supramarginal gyrus in the non-dominant side. Skin incision was done 3cm above and 3cm behind the ear pinna (the usual incision for parietal ventriculo peritoneal shunt).

Posterior parietal variety is used in the dominant side whatever the size of the tumor and in medium sized tumors in the non-dominant side to avoid affection of the optic radiation by long distance cortical dissection if the another variety (parietotemporal) is used the skin incision is performed as in parieto-temporal variety but the selection of cortical incision variety is based an anatomical data or regard to eloquent are, optic radiation and vascular pedicle, posterior parietal incision through superior parietal lobule is considered more safe as regard to eloquent speech area and optic radiation which pass in the superolateral aspect of the atrium but the vascular pedicle control by this variety is less than direct access provided by parieto-temporal junction in both varieties cortical incision is done parallel to long axis of cerebral hemisphere for 2-3 can length. The lateral ventricle is approached by using two spatulas for cortical dissection. This step is facilitated by ventricular dilatation which is evident in most of the cases and so, any preoperative CSF (cerebrospinal fluid) diversion should be avoided.

Choroid plexus papilloma occupying the frontal horn (originating from the body and extending to the incision though the middle frontal gyrus. Skin incision is done as that for frontal ventriculoperitoneal shunt but is larger.

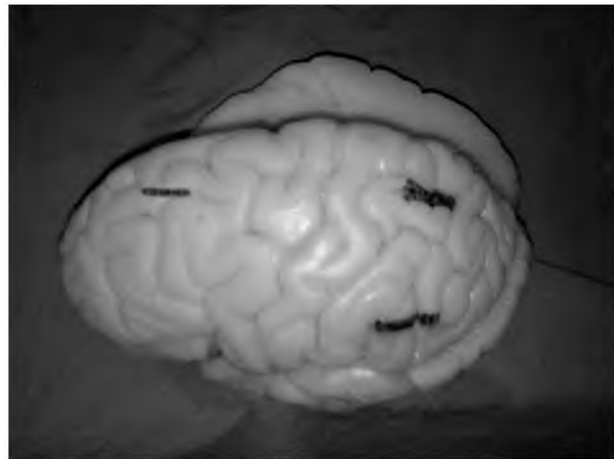


Fig. (1): Shows the varieties of the cortical incisions (middle frontal gyrus incision, parietotemporal incision and posterior parietal incision).

#### *Tumor removal:*

After ventricular opening the tumor appear as large, fragile and vascularized lesion with distinct cleavage planes this tumor is removed by one of two ways. In medium sized tumors the first goal is identification and obliteration of the vascular pedicle to decrease blood loss. The exact location of vascular pedicle is variable because it is usually displaced by the tumor but in most of cases, it can be identified in relation with the choroidal fissure. Once the vascularization is obliterated surgical removal of the tumor as "en bloc" or piecemeal can be done. In large sized tumors, the vascular pedicle can't be identified and any attempts for retraction and dissection to show it with such huge sized tumors will result in hazardous effect for the thalamus and fornix with also inevitable bleeding from just vascular tumor piecemeal removal is performed step by step with control of tumor bleeding and any feeding vessels. The operation continued until identification and obliteration of vascular pedicle (which is usually multiple vessels) that followed by removal of the remaining part of the tumor. The operation should be stopped when the blood loss endangers the child life and the rest of tumor can be removed at another session.

In the last two case of this study endoscopy was used for only accessing the ventricle and navigation of the tumor but the rest of surgery is completed as used by microscopic removal. Before closure, external ventricular drain is inserted.

#### *Postoperative findings:*

The patient is placed in intensive care unit for at least one day for multi-parameter monitoring, including vital signs, laboratory investigation and conscious level.

External ventricular drain should be followed and ventriculo-peritoneal shunt is inserted in cases

of persistent increase in the intracranial tension by 5-7 day postoperatively.

Table (1): Shows the patients included in this study.

	Sex	Age	Location	Chosen approach	Complication
1	♂	3 months	Atrium (left)	Posterior parietal	Transient paresis
2	♂	2 years	Atrium (Rt)	Parieto-temporal	
3	♀	One year	Atrium (Rt)	Parieto-temporal	
4	♀	2 years	Atrium (Rt)	Posterior parietal	
5	♂	5 months	Atrium (Rt)	Parieto-temporal	Visual field defect
6	♀	3 years	Atrium (Rt)	Parieto-temporal	
7	♂	9 months	Atrium and occipital horn (Lt)	Posterior parietal	Visual field defect
8	♀	10 years	Frontal and atrium (Rt)	Rt frontal middle frontal gyrus	

**Results**

Eight patients were included in this study. 4 male and 4 females with age ranged from 3 months to 10 years.

*Clinical and radiological analysis:*

Manifestation of increase intracranial tension as headache vomiting, head enlargement and excessive crying were reported in all cases.

Focal manifestations were limited to visual field affection in large tumors.

CT brain with contrast showed enhancing, copulated and globular lesion. This tumor was 150 in

tense on T 1 and hypertense on T2 weighted MRI with enhancement after contrast injection all cases showed dilatation of cerebral ventricles to varying degrees.

*Tumor location:*

Seven cases were located in the atrium of the lateral ventricle with extension to the occipital horn in one of them.

Frontal horn location was reported in one case. It should be noted that the case with frontal horn location was originated from the atrium and extended anteriorly.

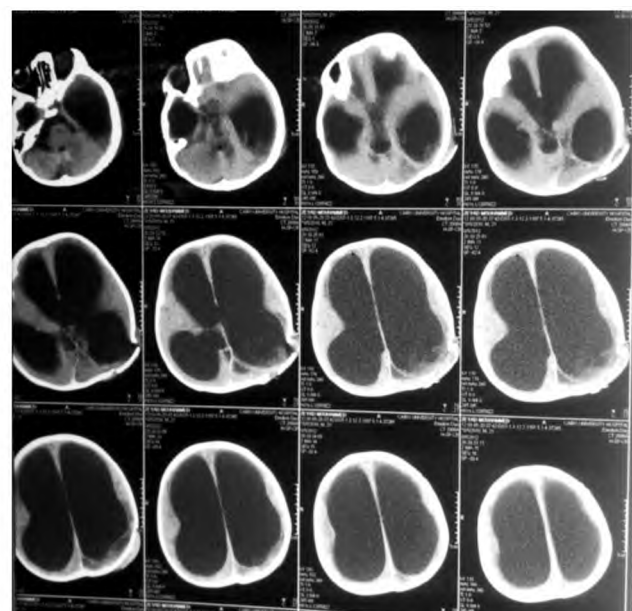
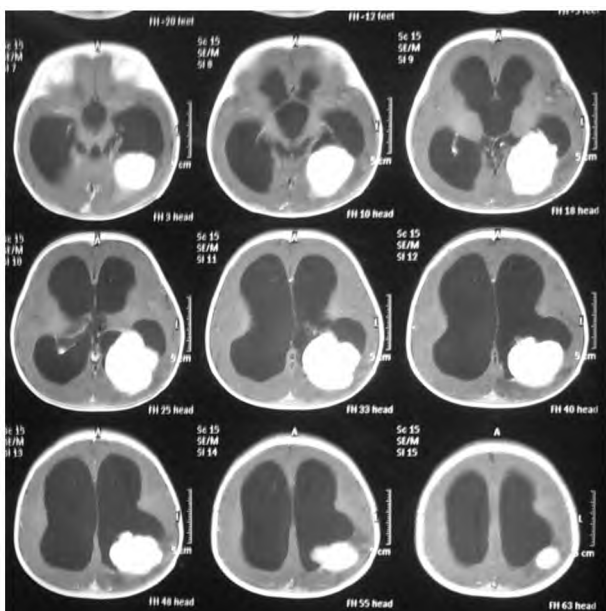


Fig. (2): Pre and postoperative films shows complete removal of the choroid plexus papilloma extending from left atrium to occipital horn through posterior parietal incision.

*Surgical results:**Cortical incision:*

Posterior parietal incision was done in three cases occupying the atrium (two of them were in dominant hemisphere and the remaining case was medium sized tumor not stretching the cortex in the non-dominant side.

Parieto-temporal incision was done in four cases with large stretching the cortex in non-dominant side.

The case with frontal location was approached through Rt middle frontal gyrus.

*The tumor and its vascular pedicle:*

All the transcortical approaches that used allow rapid access to the tumor especially that ventricular dilatation was reported in all cases. The traversing route is shorter in parieto-temporal and middle frontal gyrus incision but the degree of exposure was limited in all transcortical approaches which can be compensated to a degree with changing the angle of the microscope.

Endoscopic access to the ventricle and navigation of the tumor was used in last two cases with more detailed finding about the tumor and the

vascular pedicle especially in medium sized tumors the vascular pedicle is hidden by the tumor in all cases and identified before resection in a single case with medium sized tumor in the atrium and in the rest of the cases debulking of this fragile and vascularized tumor was needed for his identification, in this study the pedicle has no specific location to displacement by this large tumor and usually include multiple feeding vessels.

*Tumor removal:*

All cases in this study showed complete removal except in one case with marked vascular tumor that necessitated another surgical setting.

*Histopathology:*

All cases were choroid plexus papilloma without any malignancy (choroid plexus carcinoma).

*Outcome:*

There is no intra-operative mortality in this study. There were two cases with postoperative visual field deficit one case with transient hemiplegia and one case with transient sensory aphasia five case required permanent CSF diversion (four cases need ventriculoperitoneal shunt and one case need subdural-peritoneal shunt.

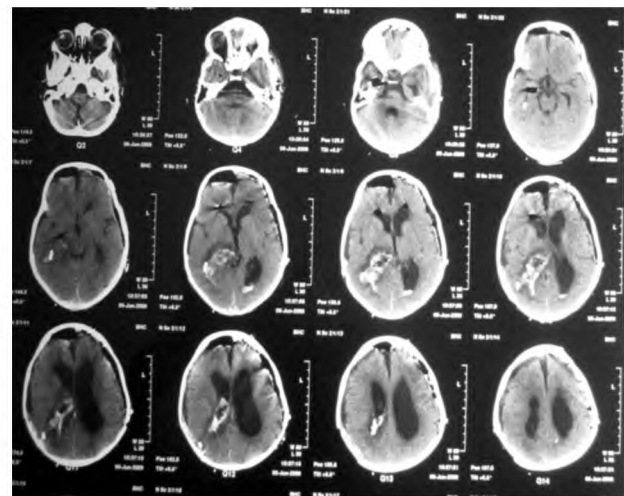
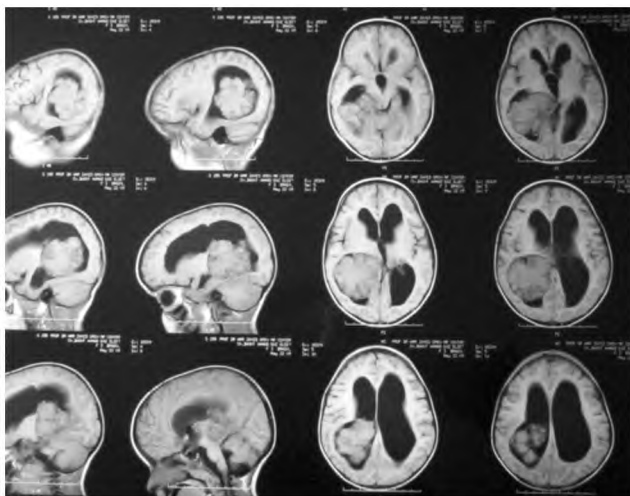


Fig. (3): Pre and postoperative films shows complete removal of the choroid plexus papilloma occupying right atrium to occipital horn through parietotemporal incision.

## Discussion

Choroid plexus papilloma is uncommon benign brain tumor that originates from choroid plexus epithelium.

It occurs in both pediatric and adult age group with incidence 2-6% and 0.5% respectively [1,2,10].

It has great tendency to present in the first and second decades of life but has been seen in other age groups also.

This tumor occurs in the lateral ventricle in pediatric age group and in the fourth ventricle in adults. However, it is reported at other uncommon

sites as third ventricle, cerebellopontine angle, suprasellar region and cerebellum. This study is concerned only about lateral ventricular location [4,9,11].

Choroid plexus papillomas located mainly in the atrium of the lateral ventricle.

This is related to anatomical origin of the tumor from the choroid plexus that extend in the atrium which is the region of confluence of body, occipital and temporal horns and the feeding vessels can be identified in this region. On the other hand, the frontal and occipital horn contain no choroid vessels, so they are not the primary location but can host the tumor originating in the atrium, which coincides with the results obtained in this study [2,4].

This tumor usually affects CSF pathway by obstruction, overproduction and deposition of proteins in subarachnoid space with subsequent impaired reabsorption. Hence, patients having choroid plexus papilloma remain asymptomatic until hydrocephalus with manifestation of increase intra cranial tension. The patients included in this study are of pediatric age group and coming mainly from rural area which aid for late diagnosis and enlargement of the tumor proper knowledge about microsurgical anatomy is very important for choosing the surgical corridor and manipulating the tumor intra operatively without injuring the neurological structures [4,11].

Each lateral ventricle includes frontal horn, body atrium, occipital horn, temporal horn. They are between the foramen of Monroe and the splenium of the corpus callosum which constitute the body of the lateral ventricle is of specific importance in surgery choroid plexus papilloma. The anterior thalamo-caudate and posterior caudate vein runs laterally in this region draining into the thalamo-striate or internal cerebral veins. Also, this region is related to choroidal fissure the separate the thalamus from the fornix and attached to both of them. In the posterior part of this fissure which extends in the atrium, the two choroidal arteries can be identified, one passing medially following the choroid plexus and the other coursing in the body of the ventricle. These feeding vessels of the tumor originate in this region [6,8,12].

It should be noted that knowledge about the anatomy of cerebral hemisphere eloquent area and tract is very important for approach selection [6,8,12]. Optic tract and radiation are of specific importance during surgery of this tumor as it is

closely related to the roof and lateral aspect of the atrium and occipital horn [13].

The trans-cortical approaches were preferred in this study as it passes a short way especially with dilated ventricle. Incision through middle frontal gyrus is used for frontal location to avoid the disconnection syndrome that reported with transcallosal corridor [14].

Choroid plexus papilloma occupying the atrium were approached by posterior parietal incision (through superior parietal lobule) or parieto-temporal junction. Posterior parietal variety is considered anatomically safer than trans-temporal or parieto-temporal approaches because it carries low risk to motor, language areas and in addition optic radiation as it passes high and medial to its pathway (as previously mentioned) [2,5,13].

However, this approach has a relatively late control of the feeding blood vessels which pass in relation to choroidal fissure.

On the other hand, the trans-temporal and parieto-temporal approaches provide lateral opening of the atrium with early exposure of choroidal fissure and vessels, so it is preferred in large tumors stretching the cortex in the non-dominant hemisphere (no risk for language area) [2,5,15].

In spite of fact that obliteration of vascular pedicle is essential before removal of choroid plexus papilloma, the vascular supply can't identify early in this study except in one case with medium sized tumor.

Tumor debulking and hemostasis was performed in all other cases until identification, and obliteration of the feeding vessels with subsequent removal of the residual tumor this match with other studies dealing with large tumors [5]. However it should be mentioned that way can result in marked hemorrhage which happen in one case of this study and the surgery was terminated and repeated again after six weeks.

Preoperative embolization reduces this type of complication but mandates radiation exposure and major risks of vascular injury and stroke [8].

The important limitation of this study is especially in the first five case is the lack of endoscopic application which is compensated in the last two cases with better exposure and identification of the tumor. In the future it is advised to perform another study with endoscopic assisted surgery as this technique when performed with more experi-

ence is expected to provide more favorable results [2].

Surgical outcome for choroid plexus papilloma in all studies including this study is satisfactory.

Chemotherapy and radiotherapy are not used for this type of tumors [7,16,17]. But can be used in patients with choroid plexus carcinoma (rarely-reported) [10] to improve their survival [18].

Proper postoperative multi-parameter monitoring, is very important with special attention to vital signs, conscious level, seizure, hydrocephalus and/or subdural hygroma [9]. The parents should be instructed for long term follow up as there is reported recurrence up to 10 years [16,17].

### Conclusion:

Choroid plexus papilloma is uncommon tumor that need special surgical plane and knowledge. Endoscopic assisted access to the lateral ventricle enhance safe and total microscopic removal of this vascular tumor through small cortical incision.

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## مع الورم الحليمى للضفيرة المشيمية فى البطن الجانبى للمخ فى الأطفال

يهدف هذا العمل البحثى والذى تم عمله ودراسته فى مستشفيات جامعة القاهرة - لدراسة كاملة وتفصيلية لورم يعد من الأورام الغير منتشرة بشكل نسبياً وهو الورم الحليمى للضفيرة المشيمية فى البطن الجانبى للمخ.

تعد الأورام الحليمية بالضفيرة المشيمية من أكثر الأورام حدوثاً فى البطن المخى الجانبى بالأطفال ولكنها لازلت تعد من أكثر الحالات صعوبة فى جراحة المخ والأعصاب لأنها موجودة فى مكان حساس جداً وصعب الوصول إليه فى المخ.

تمت هذه الدراسة على ثمانية حالات من الأطفال عن طريق استئصال الأورام باستخدام الميكروسكوب الجراحى وقد تم استخدام المنظار الجراحى المساعد فى حالتين فقط.

تم التوصل إلى استئصال كلى للورم فى ستة حالات وحيث أنه احتجنا إلى عمل تدخل جراحى ثانى لاستكمال استئصال الورم فى حالتين فقط لتحقيق الوصول إلى استئصال كلى. حيث أنه حدث مضاعفات فى حالتين حيث تأثر الأبصار وحدث ضعف بالإطراف مؤقتة فى حالة واحدة، كما أنه حدث إصابة مؤقتة فى الكلام عند حالة واحدة فقط.

لقد توصلنا فى بحثنا هذا إلى أن استخدام المنظار الجراحى المساعد المستخدم مع الميكروسكوب الجراحى فى مثل هذه الحالات يساعد بشكل كبير جداً فى الوصول إلى الاستئصال الكلى للأورام وبشكل سريع و آمن وعن طريق فتح جراحى صغير يمكن الوصول إلى البطن المخى بشكل سهل.

استخدام المنظار الجراحى فى استئصال الأورام فى بكين المخ الجانبى يمكننا من تحسين نتائج ما بعد التدخل الجراحى فى مثل هذه الأورام المحبة الصعبة وحيث أنه يعد سهل و آمن وسريع ويقلل نسبة حدوث اية مضاعفات من الجراحة ويمكننا من الاستئصال الكلى للورم

نهدف إلى التوسع فى دراسة مثل هذه الأورام المخية للوصول إلى نتائج أفضل وتحسين الحياة العامة فى ما بعد الجراحة فى الأطفال المرضى.