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Experience of choroid plexus papilloma in children at Mansoura University Hospital

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Abstract

Choroid plexus papilloma (CPP) are rare, benign tumours of neuroectodermal origin; they represent 1-3% of central nervous system (CNS) tumours in paediatric patients. Authors present their experience in the management of such lesion in Mansoura University Hospitals.

Methods. For children with Choroid plexus papilloma who were treated via microsurgical excision over 4 years from January 2012 to January 2016 in Mansoura University Hospitals, a retrospective analysis was done for age, sex, clinical manifestations, surgical treatment and follow up.

Results. Twenty-three paediatrics were treated over 4 years. Age ranged from (7 months - 8 years). There were 13 female and10 males. Tumours were located in the lateral ventricle in all cases. Features of raised intracranial pressure were the predominant presentation. Total microsurgical excision was achieved in all cases. All cases had intraoperative blood loss < 100 ml with mean Haematocrit 28%. Follow up period (7-53 months). Complete relief of all symptoms was reported in all cases with no mortality. One of our patients needs postoperative ventriculoperitoneal shunt for persistent CSF leakage and another one required subdural peritoneal shunt for persistent subdural CSF collection.

Conclusion. Total excision of Choroid plexus papilloma is usually the rule with an excellent outcome. Routine external ventricular drainage for at least 3 days is effective in lowering shunt-dependent cases as it allows the release of bloody CSF and small tumour residue. With proper microsurgical technique through superior parietal lobule to access lateral ventricle then tumour coagulation at the same time of irrigation that helps the tumour to shrink and thus, decrease the incidence of bleeding. The tumour vascular pedicle should be resected with the last part of the tumour to avoid pedicle retraction that may lead to ventricular haemorrhage.

Keywords
choroid plexus papilloma, surgical resection, complications

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Introduction

Choroid plexus papilloma (CPPs) are benign neoplasms of the choroid plexus, a structure made from tufts of villi within the ventricular system that produces cerebrospinal fluid (CSF) [13, 24]. Lateral ventricle of the brain is the commonest site of CPPs in paediatrics, but they can occur in adults too. Although the major percent of these tumours are considered benign in nature, a small percent is classified to be malignant [1, 3].

CPPs represent about 1% of intracranial neoplasms but this percentage tends to be higher in children (2-4%). The most common

location is the atrium of the lateral ventricle in paediatric population while 4th ventricle represents the commonest location in adults. Cerebellopontine angle (CPA), 3rd ventricle, and parenchyma of the cerebrum are considered rare locations of these tumours [20].

Clinically infants and children with CPP presented with; headache, vomiting, progressive head enlargement and excessive crying, as reported by the patients mothers, usually caused by hydrocephalus that results from; CSF pathway obstruction, CSF overproduction, or arachnoidal fibrosis due to recurrent occult bleeding from tumour [8].

Computed tomography (CT) and magnetic resonance imaging (MRI) show iso- or hyperdense, T1 isointense, and T2 hyperintense masses inside the ventricles that enhance with contrast, generally associated with hydrocephalus [17].

Grossly, these tumours arise as well-circumscribed cauliflower-like exophytic growths from the ventricular wall. Histologically, choroid plexus papilloma appears as papillary proliferation with a fibrovascular core lined by a single layer of monotonous cuboidal to columnar epithelium with a basement membrane [5].

Gross total excision is considered the main line of treatment of these tumours as it is associated with excellent outcomes after surgery. Moreover, adjuvant radiotherapy can be used for progressing tumours that were treated by subtotal resection. A meta-analysis of 566 choroid plexus tumours found no difference in survival when comparing patients with recurrent CPPs and those with stable disease [10].

Choroid plexus tumours can be distinguished from each other via mouth intestinal bacteria (MIB-1) labelling, status of p53, and histology into CPP, atypical papilloma, and carcinoma, and this differentiation can direct follow-up and adjuvant treatment plans later on [22].

In management of choroid plexus papilloma, there are two main problems: treatment of associated hydrocephalus, rich tumour vascular supply that is medially located in these young patients, can lead to a hazardous intraoperative blood loss. Preoperative embolization, to minimize risk of intraoperative loss, is associated with exposure to radiation and also to vessel injury and stroke [21].

We present our experience in management of such lesions in the neurosurgical department located at Mansoura University Hospitals.

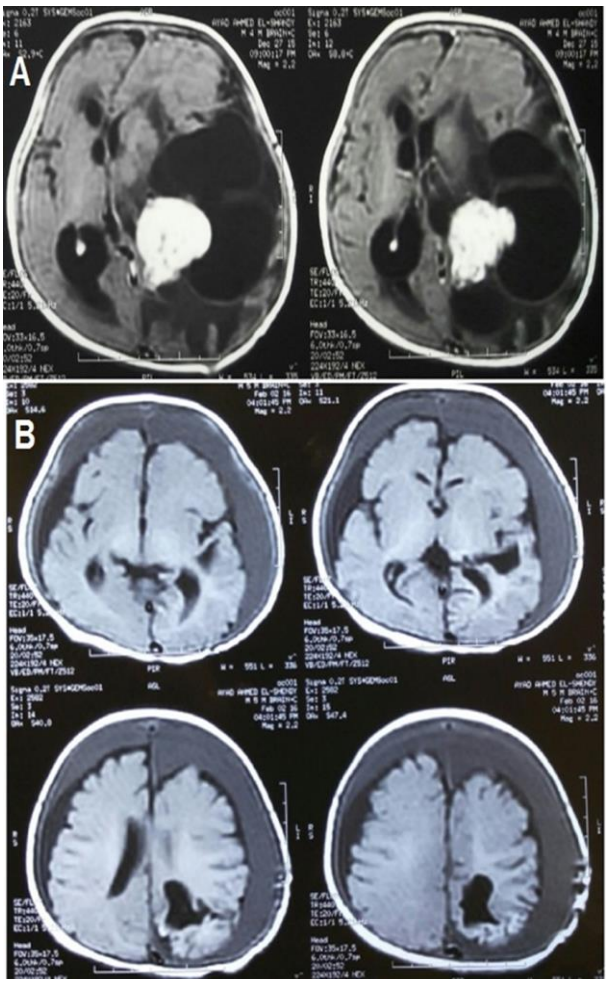

Figure 1. A 2-year-old female child with left lateral ventricle choroid plexus papilloma. A: preoperative MRI brain with contrast; B: 3-month follow up MRI brain with subdural CSF effusion and total tumour excision.

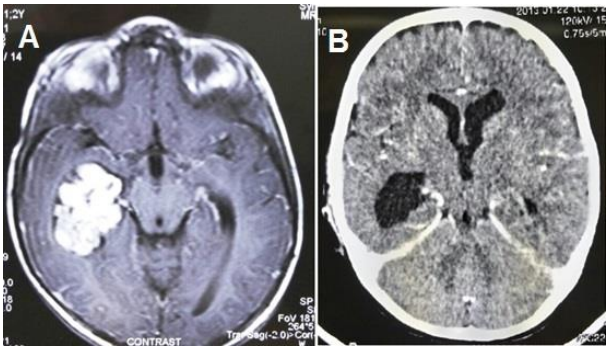

Figure 2. A 6-year-old female child with right lateral ventricle choroid papilloma. A: preoperative MRI brain with contrast, B: 1-month follow up CT brain with contrast showing total tumour excision.

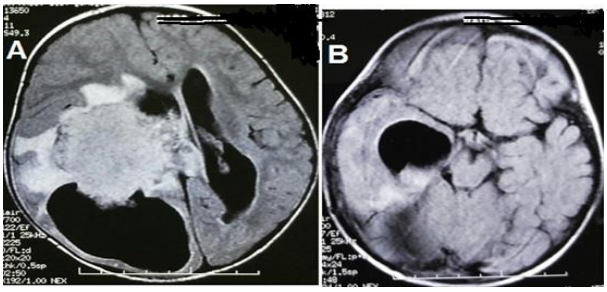

FIGURE 3. Right lateral ventricle choroid papilloma which was managed by microsurgical resection. A: preoperative MRI brain, B: 3-month follow up MRI brain showing no residual or tumour recurrence.

METHODS

Retrospective analysis of children with choroid plexus papilloma who were treated by microsurgical resection over 4 years from January 2012 to January 2016 in Mansoura University Hospitals.

Each patient was subjected to complete medical history taking, through physical examination, routine pre-operative laboratory tests, and radiological studies (CT and MRI brain). The tumour size was calculated as the maximum diameter of the lesion measured by CT and MRI.

All research activities were approved by Mansoura University local ethical committee.

Post-operative complications were classified into two categories; major or minor. Denovo permanent neurological deficits and functional status worsening were classified as major complications. Minor complications included temporary post-operative problems that faded with time like transient cranial neuropathy, temporary postoperative seizures, deep venous thrombosis (DVT), or wound healing problems such as infection, cerebrospinal fluid (CSF) leak, and pseudo meningocele.

The collected data were coded, processed and analysed using the SPSS (Statistical Package for Social Sciences) version 22 for Windows® (SPSS Inc, Chicago, IL, USA). Qualitative data was presented as number (frequency) and Percent. Quantitative data was expressed as median (range).

RESULTS

TABLE 1: Demographic, clinical, operative, and follow up data.

Variable	*Range or number*
Age (month)	39 (7 – 96)
Sex Male Female	 10 (43.47%) 13 (56.52%)
Symptoms -Headache -Vomiting -Progressive head enlargement -Excessive crying	 13 (56.52%) 8 (34.78%) 9 (39.13%) 6 (26.08%)
Tumour size (cm)	3.1 (2.4 – 4.2)
Calcification	3 (13.04%)
Major complications	0 (0%)
Mortality	0 (0%)
Minor complications -CSF leakage -Persistent subdural effusion	2 (8.69%) 1 (4.34%) 1 (4.34%)
Follow up (month)	20 (7-53)
Need for shunt	2 (8.69%)
Residual in follow up CT	0 (0%)

Twenty-three paediatrics were treated via micro-surgical resection over 7 years. Age ranged from (7 months - 8 years). There were 13 females (56.52%) and 10 males (43.47%). Features of increased intracranial pressure were present in all our cases with varying combination like headache, vomiting, excessive crying, and progressive head enlargement.

All cases were evaluated preoperatively via CT and MRI brain. Lateral ventricle of the brain was the site of the CCP in all our paediatric cases. The CT scans showed a mass that was lobulated, globular, and brightly enhancing in the lateral ventricle. Median tumour diameter measured by CT or MRI was 3.1 cm. Calcification was diagnosed radio-logically in 3 cases (13.04%), whereas all the cases had varying degrees of hydrocephalus. In all patients, MRI showed an enhancing mass in the lateral ventricle that was isointense on T1- and hyperintense on T2-weighted images.

Total microsurgical excision was performed via the transcortical approach through the superior parietal lobule in the all patients who had the tumour located the lateral ventricle trigone. The tumour was identified and then, its surface was coagulated. Moreover, it was excised after shrinking its size via combined coagulation and irrigation. Coagulation, then division of the vascular tumour pedicle was achieved on removal of the last tumour part.

Routine closed ventricular drainage was applied in all patients to allow CSF drainage for three days post-operatively. That ventricular drain was removed after exclusion of ventricular hematoma by CT scan and absence of red colour in the draining CSF fluid. In one case (4.34%), CSF leakage from the operative

incision occurred and that child needed ventriculo-peritoneal shunt for persisting hydrocephalus. Another patient (4.34%) needed subduro-peritoneal shunt because of persistent, subdural effusion.

The follow-up ranged from 7–53 months (median: 20 months). All patients experienced no symptoms at follow-up as well as the post-operative CT and MRI showed no residual lesions.

Discussion

Choroid plexus papilloma are considered as WHO grade I tumours and are among the most common tumours in the paediatric group less than 2 years. They are of neuroectodermal origin representing 1-3% of central nervous system (CNS) tumours in paediatric patients [23].

Choroid plexus papilloma has been reported to be associated with Aicardi syndrome [4, 25, 28] and Li-Fraumeni syndrome [6]. Leptomeningeal dissemination may occur [7].

Usually, CPPs occur in the lateral ventricles in paediatric population, while the 4th ventricle is the commonest location in adults; other rare sites included 3rd ventricle and the cerebellopontine angle (CPA). CPPs have also been reported in other regions like the suprasellar area, frontal lobe, and cerebellum [17]. In our study, all tumours were located in the lateral ventricle (23 cases).

In children, before closure of the sutures and fontanelles, the sequel of raised intracranial pressure such as blindness will be difficult to manifest. Seizures or mental changes may be the only presentations of these lesions while tumours involving the 4th ventricle may manifest itself with cerebellar signs. Cerebellopontine angle CPPs have been reported to cause cranial nerve deficits [8]. The predominant clinical features in our study can all be traced back to raised intracranial pressure that as consistent in all of our patients.

Clinical picture of CPP in paediatrics includes; headache, vomiting, progressive head enlargement and excessive crying. These are usually caused by hydrocephalus that results from CSF pathway obstruction, excessive CSF production, and recurrent occult bleeding from the tumour that results in arachnoidal fibrosis and adhesions [8].

In our study, headache was present in 56.52 % of cases while vomiting was experienced in 34.78 % of cases. Moreover, progressive head enlargement and excessive crying were reported by the mothers of in 39.13 and 26.08% of our cases respectively.

The mean value of maximum tumour diameter in our study was 3.1 cm, while the study conducted by safaee and his colleagues included cases with median diameter of 3 cm in the total resection group [21].

Calcification was detected in CT in 13.04% (3 cases) in our study cases. This is consistent with the findings reported in the literature stating that choroid plexus tumours calcification can be detected radiologically in around 4–20% of cases [9, 19].

Surgical gross total excision of the tumour should be attempted in all cases of choroid plexus papilloma [2]. As these tumours are highly vascular, great care should be taken during surgery to avoid excessive blood loss that may lead to mortality, as reported in some series, especially in young children. Securing the main arterial supply of the tumour, which is usually a branch of choroidal artery, is of crucial importance to avoid hazardous intraoperative bleeding followed by coagulation of the tumour and en bloc or piecemeal removal [15].

Regarding surgical excision for paediatric CPPs, there is a surgical opinion stating that there is some sort of benefit from delaying surgery till hydrocephalus develops. That is because this delay lessens the length of the corridor to the ventricles and widens the space surrounding the lesion making it more surgically accessible. The drawback to this delay is that hydrocephalus may not be the presenting symptom, leading to more tumour growing that may lead to neurological deficit that may not resolve after surgical resection [11, 16].

In addition, the drawbacks to a watchful waiting on dealing with such tumours are many. initially, repeated general anaesthesia will be needed for MRI for follow up which will be burdensome and not devoid of risks. Moreover, although CT scan may alleviate the need for MRI< but excess radiation exposure will carry risk to the child as well [11].

We reported that blood loss in this study was less than 100 ml in every patient. No major complications or mortality were reported in our study. Major complications reported in another series were rare, that may be attributed to the fact that these modern studies were conducted at a tertiary care centres [21].

Compared to published series from the modern microsurgical era, our observed morbidity was very minimal, once again confirming that these tutors can

be safely resected in both children and adults [14, 27, 29].

Whereas complete surgical excision is the ideal treatment, it could be impossible to accomplish in some cases, due to excessive blood loss or due to tumour infiltration into the surrounding vital structures. If significant residual tumour is left or recurrence is diagnosed, redo-surgery may be an option after sometime. Moreover, adjuvant therapy has been reported to be another option [18].

Pneumocephalus, pneumoventricle, subdural effusions, persistence of hydrocephalus (in those not previously shunted), and new onset hydrocephalus are the most common reported complications after surgical intervention [21, 26]. Total surgical removal of these tumours is associated with excellent survival that reaches 100% at 10-year follow up as reported in the literature [14].

In our series, only two cases required post-operative shunts; one needed ventriculoperitoneal shunt due to persistent CSF leakage, and the other needed subduroperitoneal shunt for persistent subdural CSF collection.

Although some studies have described neurocognitive sequelae related to surgery for CPP [12, 16], our study is not large enough to make any significant conclusions with respect to differential outcomes in children compared to adults.

Safaee and his associates also reported a 22 % incidence of temporary swallowing dysfunction that required percutaneous endoscopic gastrostomy tubes, tracheostomy, or both, with no difference in the incidence when comparing patients who received gross total resection with the subtotal resection group [21].

The prognosis for CPP patients treated with gross total resection (GTR) is excellent, with a report of 100% survival at 5 years after surgical resection in many series including ours, and adjuvant therapy is not indicated in these patients. Radiotherapy is not necessary after GTR, and its usefulness should be reserved for recurrent disease. Unfortunately, the prognosis for patients with choroid plexus carcinoma CPC is guarded, with an overall 5-year survival rate of 26-50%. GTR is generally considered the most important prognostic factor for CPC as was confirmed in our analysis [14].

Several limitations can be noted in this current study. Small sample size limits the statistical power of our analysis, but is an unfortunate and inevitable consequence of studying such a rare tumour. Moreover, this study is a single centre study that could affect the validity of our results as it serves a localized population.

Conclusion

Total excision of Choroid plexus papilloma is usually the rule with excellent outcome. Routine external ventricular drainage for at least 3 days is effective in lowering shunt dependent cases as it allows release of bloody CSF and small tumour residue. With proper microsurgical technique through superior parietal lobule to access lateral ventricle then tumour coagulation at the same time of irrigation that helps the tumour to shrink and thus, decrease the incidence of bleeding. The tumour vascular pedicle should be resected with last part of the tumour to avoid pedicle retraction that may lead to ventricular haemorrhage.

Authors contributions

This work was carried out in collaboration between all authors. Author Amr Farid, designed the study, Author Mohamed State wrote the protocol, Author Ahmed Zaher managed the literature research, Author Hatem Badr performed the statistical analysis and revised the final manuscript. All surgical procedures were carried out by the same surgical team including the four authors. All authors read and approved the final manuscript.

Abbreviations

CPC	Choroid plexus carcinoma.
CPP	Choroid plexus papilloma.
CPA	Cerebellopontine angle.
GTR	Gross total resection.
MIB-1	Mouse intestinal bacteria.
MRI	Magnetic resonance imaging.

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