

Case Report

Incidental choroid plexus papilloma in a child: A difficult decision

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Received: 07 October 16 Accepted: 20 March 17 Published: 26 May 17

Abstract

Background: Choroid plexus tumors (CPT) in the pediatric population are usually discovered in symptomatic patients often with symptoms of increased intracranial pressure, with hydrocephalus as the most common presentation, along with seizures, subarachnoid hemorrhage, or focal neurological deficit. Most CPTs are found to be benign choroid plexus papillomas (CPP), whereas a small number are intermediate and malignant choroid plexus carcinomas (CPC). Total surgical resection is the established definitive treatment for symptomatic CPP.

Case Description: We describe a young female who was found to have an incidental CPT during workup for recent head trauma without neurological deficits or hydrocephalus. She underwent a surgical operation to remove the tumor successful, with 1-year follow-up showing no recurrence and normal developmental milestones.

Conclusion: This rare presentation of an asymptomatic CPT brings attention to the fact that there is no clear evidence for how or when to treat such patients. Because discovery of a CPT in an asymptomatic patient is uncommon, the treatment plan appears to be developed on a case-by-case basis. We hope to generate discussion for establishing an agreed upon treatment approach for CPTs in asymptomatic patients.

Key Words: Child nervous system, choroid plexus papilloma, choroid plexus tumor, incidental, oncology

Access this article online**Website:**www.surgicalneurologyint.com**DOI:**

10.4103/sni.sni_386_16

Quick Response Code:**INTRODUCTION**

Choroid plexus tumors (CPT) in the pediatric population are usually discovered, unfortunately, when patients present symptomatically. Symptoms of increased intracranial pressure (ICP) with hydrocephalus are the most common presentation, along with seizures, subarachnoid hemorrhage (SAH), or focal neurological deficit.^[1] Most are found to be benign World Health Organization (WHO) grade I choroid plexus papillomas (CPP), whereas a small number are intermediate and malignant choroid plexus carcinomas (CPC). Total surgical resection is the definitive treatment for symptomatic CPTs.^[1] We describe a patient who was found to have an incidental

CPT following a fall. She was asymptomatic with respect to the mass and had been developing normally without radiological evidence of hydrocephalus. We discuss the decision whether to operate or not.

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How to cite this article: Laarakker AS, Nakhla J, Kobets A, Abbott R. Incidental choroid plexus papilloma in a child: A difficult decision. *Surg Neurol Int* 2017;8:86. <http://surgicalneurologyint.com/Incidental-choroid-plexus-papilloma-in-a-child-A-difficult-decision/>

CASE REPORT

An 11-month-old girl presented to the emergency room bleeding from her ear 2 days after an accidental fall rolling off the bed and striking her head. The patient was otherwise healthy with no relevant medical, birth, or social history. An initial computed tomography (CT) of the head was negative for hemorrhage [Figure 1]; however, a small hyperdensity was noted in the right temporal horn. A subsequent magnetic resonance imaging (MRI) [Figure 2] identified a well-circumscribed mass 1 cm in diameter without hemorrhage or hydrocephalus. The patient was discussed at a multidisciplinary tumor board and, after some controversy, the decision was made to proceed with surgical resection [Figure 3]. The lesion was successfully removed and the patient made a full recovery without any neurological deficit. Pathology revealed the lesion to be a CPP WHO grade I.

DISCUSSION

CPTs are rare, according to the Canadian Pediatric Brain Tumor Consortium experience, the annual age-adjusted incidence rate was 0.22 ± 0.12 (95% CI 0.16–0.28)/100,000 in children less than 3 years of age.^[3] The treatment of choice for CPTs is surgery with total resection being the goal. Complete removal of the tumor is generally curative and leads to resolution of the presenting symptoms in nearly all patients. Even in CPC, total resection (if feasible) leads to the best possible outcome. After incomplete surgery, a “wait-and-see” policy seems to be justified for all CPT. The separation of CPT based on MIB-1 labeling, p53 status, and histology into CPP, atypical papilloma, and carcinoma can direct follow-up and adjuvant treatment plans.^[2] For example, adjuvant multi-agent chemotherapy and craniospinal radiotherapy following surgery should be considered for CPC’s.^[8,9]



Figure 1: Computed tomography of the head revealing a hyperdense focus measuring up to 6.5 mm in the right temporal lobe

In the case of an incidentally found CPT, there is no evidence indicating whether prompt surgical removal is best, or that surgery should be withheld until routine surveillance shows either radiographic changes in the tumor, an increase in cerebrospinal fluid (CSF) volume (hydrocephalus), or the patient becomes symptomatic.

The benefit to waiting until hydrocephalus develops is that it lessens the length of the corridor to the ventricles and increases the space around the tumor giving greater access to its blood supply. This may allow for an easier resection of the CPT. The drawback to this is that hydrocephalus may not be the presenting symptom, and that the first neurological insult from mass effect, a SAH, or seizures may result in focal deficits that do not resolve after removal of the tumor. Furthermore, there is some evidence that cognitive deficits and regression may develop as these tumors grow.^[6] The drawbacks to a watchful waiting approach are many. First, in a child this age, general anesthesia is required to acquire serial MR scans. This is burdensome and not devoid of risks. To pursue CT imaging alleviates the need for MR scans, but then requires radiation and a contrast dye load, which again carry risks. And of course, one would not wait for the symptoms to present itself to repeat imaging and then make a surgical decision.

Matsuyama *et al.* reported a case of an incidentally found CPP in a 19-year-old patient. The CPP extended from the third ventricle into the right lateral ventricle. The tumor was operated on promptly. They reported that the slightly enlarged right lateral ventricle contributed to the decision as this made the tumor more accessible.^[5]

There may be a role for preoperative embolization to reduce the amount of intraoperative blood loss, and is therefore, associated with higher rates of total resection and lower operative morbidity and mortality.^[7] Selective embolization of CPTs has yet to be shown to be an

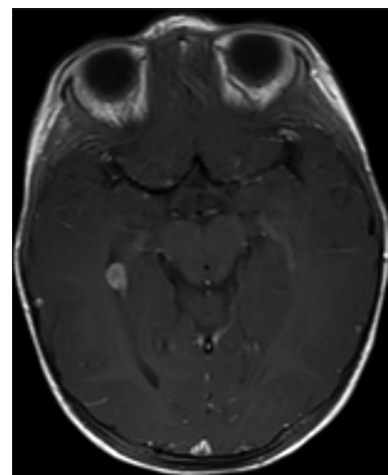


Figure 2: Magnetic resonance imaging of the head with and without contrast revealing a right lateral ventricular 8mm lesion likely associated with the choroid plexus

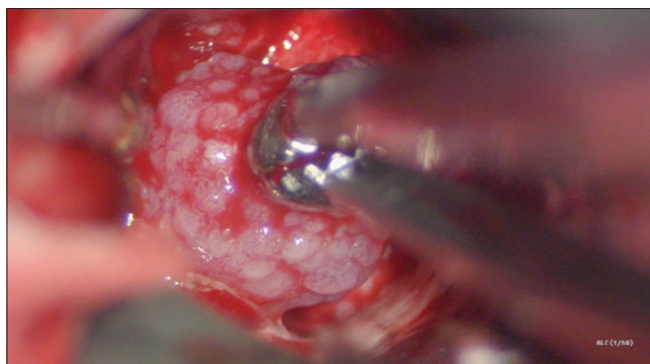


Figure 3: Tumor forceps removing the choroid plexus papilloma from the right lateral ventricle

effective stand-alone treatment strategy.^[7] As patients usually present symptomatically, embolization is unlikely to be the best treatment option and so surgical resection is generally the best choice. Furthermore, embolization is not without risk, particularly given that the anterior and posterior choroidal arteries that supply choroid plexus tumors also supply eloquent structures and may not be significantly dilated in children with these tumors.^[7]

Radiosurgery may also be a possible treatment for CPTs, however, little is known about the efficacy or use in asymptomatic patients as the risks may outweigh the benefits of use.^[2]

This rare presentation of an asymptomatic CPT brings attention to the fact that there is no clear evidence for how or when to treat such patients. We opted to operate promptly, but electively, to prevent any long-term sequelae from the CPT. A clear treatment plan is established for symptomatic patients but is not as well developed for asymptomatic patients. As discovery of a CPT in an asymptomatic patient is extremely rare, the treatment plan appears to be developed on a case-by-case basis. For the initial workup, even in the asymptomatic patient, it may be that an MRI scan is warranted when a hyperdense lesion near a ventricle is discovered, as was done for our patient. In the event that an MRI is not performed, a follow-up CT scan should be done to monitor for changes or to confirm the resolution of blood if hemorrhage was suspected.

CONCLUSION

After our literature search, we were still unclear on how to proceed given the paucity of data on the natural history of these tumors. We made our decision given that as the tumor grows and will ultimately require surgery, the psychological trauma to the child is minimized by operating in this age group as well as the need for tissue diagnosis. As a small percentage of these may be high grade, it is worth the risk to identify this patient earlier, especially because the extent of resection is significantly

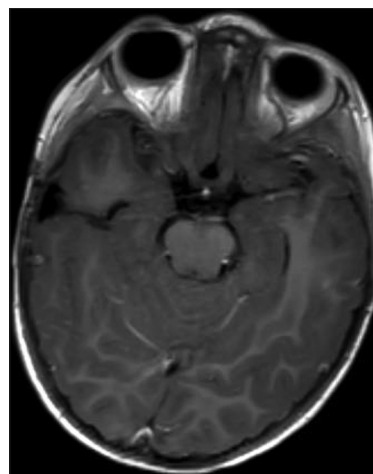


Figure 4: Status post lesion resection from the right temporal horn without evidence of abnormal enhancement or recurrent tumor

associated with increased survival.^[4] On 1-year follow-up, our patient is doing quite well as she is reaching normal growth and developmental milestones. A 1-year MRI [Figure 4] was unremarkable for any residual or abnormal enhancement except postsurgical changes. Overall, surgery was warranted; the decision to wait or operate immediately could be argued both ways, but we felt it was in the best interest for the child to remove this benign tumor shortly after the incidental finding.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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