Choroid Plexus Papilloma of the Fourth Ventricle in a Pediatric Patient: A Case Report

Luma Qutub 1, Bandar Y. Sulaimani 2, Khalid T. Alghamdi 3, Ahmed A. Mulla 4, Hussam Kutub 5

1. College of Medicine, Batargi Medical College, Jeddah, SAU 2. Medicine, College of Medicine, King Saud Bin Abdulaziz University for Health Sciences, Jeddah, SAU 3. Medicine, College of Medicine, King Saud Bin Abdulaziz University for Health Sciences, King Abdullah International Medical Research Centre, Jeddah, SAU 4. Neurosurgery, Ministry of National Guards Health Affairs, Jeddah, SAU 5. Consultant Neurosurgery, King Abdulaziz Medical City, Ministry of National Guards Health Affairs, Jeddah, SAU

Corresponding author: Hussam Kutub, hussamkutub@yahoo.com

Abstract

Choroid plexus papilloma (CPP) is a rare intraventricular tumor. The common locations of the tumor vary based on the age of the patient. It usually occurs in the supratentorial region in children, however in adult patients, these tumors commonly present in the infratentorial region. We are presenting a rare case of a pediatric patient with a two month history of decreased activity and loss of interest in his surroundings and gait imbalance. He underwent a suboccipital craniotomy and excision of a CPP in the fourth ventricle. In conclusion, CCP should be considered as part of the differential diagnosis of intracranial tumors when the clinical presentation and investigations are suggestive regardless of the location to avoid misdiagnosing it when it occurs in an uncommon location.

Introduction

Choroid plexus papilloma (CPP) is one of the rare intraventricular tumors that accounts for 0.4 to 1% of brain tumors in adults and 1.5 to 6% in the pediatric population [1]. It is a tumor of neuroectodermal origin, which can be cured by a total tumor resection [2]. In adults, it commonly occurs in the infratentorial region, and the most common site is the fourth ventricle. However, in the pediatric population, it commonly occurs in the supratentorial region with the most common site being the lateral ventricle [3]. In addition to that, even though it can appear at any age, 70% of the cases in the pediatric population occur in children younger than two years old [1,4,5]. CCP is a benign disease in 80% of patients with a good prognosis [6]. In this study, we present a rare case of a child with choroid plexus papilloma in the fourth ventricle.

Case Presentation

A seven-year-old boy presented with a two months history of decreased activity and loss of interest in his surroundings which was followed by a developing gait imbalance. On examination, the patient was alert and awake with no cranial nerve palsies, but he had dysdiadochokinesia and ataxia. A brain computed tomography (CT) and magnetic resonance imaging (MRI) showed a fourth ventricular enhancing lesion and hydrocephalus suggestive of a choroid plexus papilloma (Figure 1). The patient was taken to the operative room for a standard sub-occipital craniotomy with excision of the fourth ventricular lesion (Figure 2). The tumor was well demarcated arose from the choroid plexus of the fourth ventricle. A successful total excision was performed and no cerebrospinal fluid diversion. Post-operative imaging documented total excision of the lesion and resolution of the hydrocephalus. Clinically, the patient demonstrated significant improvement in mood and communication. Upon discharge, the patient was conscious, alert, communicating and was able to sit independently and to walk with help.
Discussion

The World Health Organization describes the classification of choroid plexus tumors as follows: low grade, atypical, and carcinoma, which is determined by histopathological examination. Choroid plexus papillomas have less than two mitotic figures per 10 high-power fields (HPFs), atypical ones have two to five per 10 HPFs, and carcinomas have more than five mitotic figures per 10 HPFs [7]. Benign choroid plexus tumors have a good prognosis after resection [2].

The clinical presentation of CPPs is mainly related to the raised intracranial pressure caused by the hydrocephalus which can cause headache with nausea or vomiting, reduced mentation and lateral gaze.
palsies. Subarachnoid hemorrhage may occur in some patients mainly due to bleeding from the tumor [8]. In our case, for instance, the patient presented with decreased activity, loss of interest in his surroundings, dysdiadochokinesia, and ataxia.

The differential diagnoses for choroid plexus papilloma include intraventricular tumors such as papillary ependymoma, medulloblastoma, and pilocytic astrocytoma among other intraventricular tumors [9]. Gross total resection is the treatment of choice. Hydrocephalus management is an important part of the treatment plan, which may require placing a ventriculoperitoneal (VP) shunt [10]. In our case, the patient did not require a VP shunt after the surgery.

Conclusions
Choroid plexus papilloma is a rare intracranial benign tumor that can cause hydrocephalus and increased intracranial pressure. In the pediatric population, the fourth ventricle is considered a rare location for CCPs. It is important to include CCP in the differential diagnosis of intracranial tumors in any location when the clinical presentation and imaging are supportive to avoid misdiagnosis if it occurred in a rare location. The treatment for CCP is complete surgical resection, which has an excellent prognosis and survival rate. Patients typically do not require any adjuvant treatment except in complicated or atypical cases.

Additional Information

Disclosures

Human subjects: Consent was obtained or waived by all participants in this study. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

References

1. Boyd MC, Steinbok P: Choroid plexus tumors: problems in diagnosis and management . J Neurosurg. 1987, 66:800-5. 10.3171/jns.1987.66.6.080
2. Ogiwara H, Dipatri AJ Jr, Alden TD, Bowman RM, Tomita T: Choroid plexus tumors in pediatric patients. Br J Neurosurg. 2012, 26:32-7. 10.3109/02688697.2011.601820
3. Ellenberger RG, Winston KR, Kupsky WJ: Tumors of the choroid plexus in children . Neurosurgery. 1989, 25:327-35. 10.1227/00006123-198909000-00001
4. Tacconi L, Delfini R, Cantore G: Choroid plexus papillomas: consideration of a surgical series of 33 cases . Acta Neurochir (Wien). 1996, 138:802-10. 10.1007/BF01411257
5. McEvoy AW, Galloway M, Revesz T, Kitchen ND: Metastatic choroid plexus papilloma: a case report . J Neurooncol. 2002, 56:241-6. 10.1023/a:1015044012574
6. Muñoz Montoya IE, Maldonado Moran MA, Santamaría Rodriguez P, Toro Lopez S, Perez Cataño CS, Lacque Suarez JC: Choroid plexus papilloma of the fourth ventricle: a pediatric patient . Asian J Neurosurg. 2019, 14:585-8. 10.4103/ajns.AJNS_501_18
7. Ruggeri L, Alberio N, Alessandrello R, et al.: Rapid malignant progression of an intraparenchymal choroid plexus papilloma. Surg Neurol Int. 2018, 9:131. 10.4103/sni.sni.434_17
8. Zhou WJ, Wang X, Peng JY, et al.: Clinical features and prognostic risk factors of choroid plexus tumors in children. Chin Med J (Engl). 2018, 131:2938-46. 10.4103/0366-6999.247195
9. Muly S, Liu S, Lee R, Nicolaou S, Rojas R, Khosa F: MRI of intracranial intraventricular lesions. Clin Imaging. 2018, 52:226-39. 10.1016/j.clinimag.2018.07.021
10. Konar SK, Kandregula S, Beniwal M, et al.: Management of choroid plexus tumours: a comprehensive study from a tertiary hospital. Clin Neurol Neurosurg. 2021, 201:106454. 10.1016/j.clineuro.2020.106454