Case Report

Third ventricle choroid plexus papilloma: 2 cases

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A B S T R A C T

The third ventricle is an uncommon location for choroid plexus papillomas. In adults, these tumors most commonly occur in the fourth ventricle. In children, they are more commonly found in the lateral ventricles. When these lesions are discovered in the third ventricle, they are often posteriorly located. Hydrocephalus and macrocephaly are typical sentinel findings. We present 2 cases of this uncommon presentation of third ventricular choroid plexus papilloma.

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Introduction

Choroid plexus papillomas are uncommon, representing less than 1% of all intracranial tumors [1]. These tumors have an unusual age distribution in which they are typically found in either lateral ventricle in children, but the fourth ventricle is a more common location for adults with this tumor [2]. Choroid plexus papillomas arise from the epithelium of the choroid plexus and may produce CSF which can result in hydrocephalus. Another source of hydrocephalus is obstruction of the interventricular CSF flow pathways.

Choroid plexus papillomas of the third ventricle are exceedingly rare with only 19 cases reported as of 1983 [3] and scant reports of this entity in the interim. Reported here are 2 separate cases of third ventricle choroid plexus papillomas with disparate clinical presentations.

Case 1

A 3-year-old female presented to the emergency department after falling out of bed. She struck her head on the floor and...
later developed headache and emesis 2 hours after the fall. A CT of the head was performed (Fig. 1A) which revealed a well-defined mass centered in the anterior third ventricle. There was associated ventriculomegaly without frank periventricular edema. No intracranial hemorrhage or skull fracture was revealed.

The child was admitted to the pediatric intensive care unit where an MRI with contrast was requested the following day. MRI showed a predominantly FLAIR hyperintense lobulated mass in the third ventricle with central linear FLAIR hypointensity (Fig. 1B). Again, no periventricular edema was perceptible. Contrast-enhanced T1-weighted imaging (Fig. 1C) revealed a frond-like character to the periphery of the tumor with robust enhancement throughout most of the tumor. The cerebral aqueduct was not obstructed. Differential considerations included choroid plexus papilloma, atypical choroid plexus papilloma, and choroid plexus carcinoma. There was no evidence of metastatic disease in the neural axis. The tumor was resected with confirmation of choroid plexus papilloma on histology. The patient recovered without recurrent disease.

Case 2

A 3-month-old male presented to the emergency department with altered mental status. CT of the head was requested which demonstrated a small mass which was epicentered in the posterior third ventricle (Fig. 2A). There was obstructive hydrocephalus with the mass extending into the cerebral aqueduct. Follow-up MRI showed enhancement of the mass (Fig. 2B) with characteristic frond-like enhancement (Fig. 2C). As in the first case, differential considerations included choroid plexus papilloma, atypical choroid plexus papilloma, and choroid plexus carcinoma. No metastatic lesions were seen in the brain or spine. The mass was resected without complication.

Discussion

In children, choroid plexus papillomas make up 2%-4% of brain tumors with 10%-20% occurring in the first year of life.
Fig. 2 – (A). Sagittal CT of the head shows a mass projecting into the posterior aspect of the third ventricle (arrow). The third ventricle is enlarged (asterisk).
(B). Axial contrast-enhanced T1-weighted image of the brain demonstrates a lobulated frond-like mass occupying the posterior third ventricle (arrow).
(C). Sagittal heavily T2-weighted image of the brain reveals a mass (arrow) extending into the cerebral aqueduct.

[4]. Presented here is a case of third ventricle choroid plexus papilloma discovered incidentally in a 3-year-old as well as a symptomatic third ventricle choroid plexus papilloma in a 3-month old. Interestingly, the tumor in the 3-year-old was located in the anterior aspect of the third ventricle, and despite its size and mass effect, there was no imaging demonstration of transependymal flow of CSF. This may be explained by the chronicity of the lesion.

Choroid plexus papillomas are WHO grade I lesions. The major competing differential considerations with this mass are the atypical choroid plexus papilloma and the choroid plexus carcinoma which are WHO grades II and III, respectively. The atypical choroid plexus papilloma is difficult to distinguish from the choroid plexus papilloma on imaging. Choroid plexus carcinomas commonly manifest in the trigone of one of the lateral ventricular atria and often invade the surrounding brain parenchyma. Imaging of the spine is necessary with choroid plexus carcinoma, as metastatic disease is more likely with this lesion.

In addition to the overproduction of CSF, these tumors are difficult to manage because of their high vascularity. Despite the challenge of intraoperative bleeding, endoscopic removal of this tumor has been reported [5]. Both children presented here recovered without demonstration of recurrence or metastasis.

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