Treatment of third ventricular choroid plexus papilloma in an infant with embolization alone

Case report

JOSEPH J. WIND, M.D.,1 RANDY S. BELL, M.D.,2,3 WILLIAM O. BANK, M.D.,3 AND JOHN S. MYEROS, M.D.4

1Department of Neurologic Surgery, The George Washington University; 2Department of Neurologic Surgery, Walter Reed Army Medical Center; 3Department of Interventional Neuroradiology, Washington Hospital Center; and 4Division of Neurologic Surgery, Children’s National Medical Center, Washington, DC

The authors present the case of a 3-month-old boy with a third ventricular tumor consistent with a choroid plexus papilloma. This child presented with macrocephaly, irritability, inability to roll over, and vomiting. He was found to have an enlarged head circumference, a full and tense fontanel, splayed sutures, and forced downward gaze. Imaging revealed severe ventriculomegaly and a brightly enhancing third ventricular lesion consistent with papilloma. Treatment planning included placement of a ventriculoperitoneal shunt to treat hydrocephalus and to allow the child to grow prior to resection. Due to the vascular nature of these tumors and the age of this child, the tumor was embolized with a plan for eventual resection; however, embolization resulted in involution and total regression of the tumor. There is no residual disease at last follow-up of 16 months. In this specific scenario of a choroid plexus papilloma in an infant, when operative intervention may be technically difficult and associated with significant morbidity, embolization with close observation may be a valid treatment option. If used, the patient would need to be closely followed for evidence of residual or recurrent disease, which would require operative intervention.

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KEY WORDS • choroid plexus papilloma • papilloma • embolization • third ventricle

Choroid plexus papillomas are benign tumors found most frequently in the pediatric population. They present challenges to operative management in terms of the young age at typical presentation, propensity for CSF overproduction, and significant vascularity. Due to this vascularity, endovascular embolization in the preoperative setting has been described as a method for decreasing surgical blood loss during the resection of these tumors. We present a unique case of a third ventricular choroid plexus papilloma associated with CSF overproduction, which was treated with endovascular embolization alone.

Case Report

History and Examination. This 3-month-old boy presented to medical attention with macrocephaly, irritability, vomiting, and inability to roll over. On physical examination, he was found to have a head circumference above the 99th percentile, a full and tense fontanel, splayed cranial sutures, and forced downward gaze.

Radiographic studies including noncontrast CT and contrasted MR imaging demonstrated severe ventriculomegaly. The MR imaging sequences demonstrated a lesion within the third ventricle, which avidly and homogeneously enhanced on postcontrast T1-weighted sequences (Fig. 1). Based on the radiographic appearance of this lesion, a presumptive diagnosis of choroid plexus papilloma was made.

Treatment. Given the age of the patient and the location of the tumor, a ventriculoperitoneal shunt was placed with the goal of treating hydrocephalus and allowing the patient to grow to a point at which resection would be safer and more feasible. However, the patient developed CSF leakage from the cranial shunt incision site 1 month after insertion; the shunt was removed and an EVD was placed. The EVD documented outputs of greater than 1000 ml of
The EVD was subsequently converted to a ventriculoatrial shunt with a programmable valve.

At this point, it was determined that endovascular embolization of the tumor might offer some benefit in decreasing tumor CSF production, allowing the child to grow and undergo resection in the future.

Standard diagnostic cerebral angiography was performed, at which point the vascular supply of the tumor was determined to be derived from an enlarged posterior medial choroidal branch arising form the right posterior cerebral artery. A Marathon Microcatheter (ev3 Endovascular, Inc.) and Mirage 0.008-in guidewire (ev3 Endovascular, Inc.) were used to selectively catheterize the right posterior cerebral artery via the left vertebral artery. A microcatheter injection was performed, followed by selection of the posterior medial choroidal branch (Fig. 2). The guidewire was removed, and 0.3 ml of Onyx-18 (ev3 Endovascular, Inc.) was injected over the course of 12 minutes, demonstrating excellent tumor penetration (Fig. 3).

The patient tolerated the procedure well. Subsequent cranial imaging demonstrated ventricular collapse. The settings on the programmable ventriculoatrial shunt valve were adjusted to accommodate the apparent decreasing CSF production following embolization. The patient was discharged from the hospital without any neurological deficits.

Posttreatment Course. Serial MR images were obtained to follow the tumor size. These images demonstrated a steady regression of the tumor, with no visible lesion present on an MR imaging obtained 7 months after embolization. Subsequent MR imaging studies have demonstrated no evidence of residual tumor at the last follow-up of 16 months (Fig. 4). The patient has had normal neurological function and development during this period.

Discussion

Choroid plexus papillomas are relatively rare intracranial tumors, representing less than 1% of intracranial neoplasms overall. They are more frequently encountered in children than in adults and account for up to 12% of intracranial neoplasms in children younger than 2 years of age. Choroid plexus papillomas are most frequently encountered in the lateral ventricles in children, whereas they occur within the fourth ventricle more commonly in adults.14

Choroid plexus papillomas are histologically benign neoplasms derived from neuroectoderm, assigned a WHO Grade I; Grade II designation is reserved for atypical CPPs. Resection of these tumors is often curative, with little chance of recurrence following gross-total resection.12,16 These tumors present special management challenges due to several unique characteristics, including the potential for CSF overproduction and the inherent vascularity of these lesions.

Overproduction of CSF can lead to extremely large daily production of CSF—reported as high as 2000 ml per day—and subsequent hydrocephalus.1,3,11 Studies have demonstrated the expression of a protein aquaporin-1 in normal choroid plexus and CPPs, and it may be involved in CSF overproduction.4 Electron microscopy performed on CPPs has also confirmed ultrastructural similarities between normal choroid plexus and these tumors, making them suitable for CSF overproduction.4

The vascularity of these lesions is often quite robust and varies somewhat with the intraventricular location of the tumor; they can often be supplied by a rich anastomotic bed of choroidal blood vessels. Due to the rich vascular supply of these tumors, endovascular embolization as a preoperative adjunct has been used in an attempt to reduce blood loss intraoperatively.
These challenges are further amplified in the setting of an infant, where surgical accessibility is made more difficult by the small size of the child and intolerance for significant blood loss. Mortality rates due to excessive blood loss have been reported as high as 15% in the treatment of these tumors, with relatively high rates of perioperative morbidity as well. Preoperative endovascular embolization has been only somewhat successful in the pediatric population; difficulties have been attributed to technical challenges with cannulating the posterior choroidal arteries. Preoperative endovascular embolization has been only somewhat successful in the pediatric population; difficulties have been attributed to technical challenges with cannulating the posterior choroidal arteries. The advance of microcatheters and microguidewires has allowed access to smaller and more distal blood vessels within the cerebrovascular system. This, coupled with new embolic agents such as Onyx, has offered endovascular neurosurgeons and interventional neuroradiologists further tools for embolization procedures in the treatment of vascular lesions and tumors.

It is important to note that the use of preoperative embolization in this case was intended to decrease blood supply to the tumor and therefore decrease CSF overproduction, so that the patient’s shunt could be reinserted. Resection was planned once the patient had grown larger, making surgery safer and more feasible. The regression of the tumor was a serendipitous finding.

Despite CPPs being histologically benign lesions, there are rare reports of progression of previously designated WHO Grade I or II CPPs progressing to choroid plexus carcinomas. The initial pathologies in these reported cases were all somewhat atypical, often with focal areas of increased mitotic activity. If embolization is to be attempted as an initial treatment for these tumors, the rare possibility of progression of these tumors must be considered and discussed with the patient’s family, and perhaps periembolization endoscopic biopsy should be undertaken to ensure accurate pathological diagnosis.

Close radiographic follow-up with serial MR imaging is warranted to assess the regression of these tumors and to observe for any recurrence or residual disease. Any recurrent or residual disease would warrant resection.

Conclusions

Choroid plexus papillomas in infants present significant challenges to the treating physicians. Endovascular embolization of these tumors is a well-recognized adjuvant to make subsequent surgery safer and more feasible. In the specific setting of a CPP in an infant where operative intervention may be technically difficult and associated with significant morbidity, embolization with close observation may be a treatment option. If used, the patient will require close follow-up for evidence of residual or recurrent disease, which would require operative intervention.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper. Author contributions to the study and manuscript preparation include the following. Conception and design: Myseros, Bell, Bank. Acquisition of data: all authors. Analysis and interpretation of data: Myseros, Wind, Bell. Drafting the article: Myseros, Wind, Bell. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors.

References


Address correspondence to: John S. Myseros, M.D., Division of Neurological Surgery, Children’s National Medical Center, 111 Michigan Avenue NW, Washington, DC. email: jmyseros@cnmc.org.