Extraventricular choroid plexus papilloma in the brainstem

Case report

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Choroid plexus papilloma (CPP) is extremely rare in the brainstem. The authors report the case of a 10-year-old boy with a lesion in the pons that was misdiagnosed as a glioma preoperatively. The boy underwent partial resection of the lesion, which was diagnosed as a CPP based on histopathological findings. The authors review the MRI findings in this case and conclude that the presence of a well-defined boundary and no obvious cerebral edema are valuable features for distinguishing brainstem CPP from glioma. Although previous reports of parenchymal CPPs have described enhancement on contrast-enhanced T1-weighted MR images, the lesion in this case did not demonstrate significant enhancement. The authors note that the diagnosis of extraventricular CPP cannot be ruled out in a case of brainstem tumor without marked enhancement.

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KEY WORDS • choroid plexus papilloma • brainstem • MRI • Virchow-Robin space • oncology

Choroid plexus papilloma (CPP) is a rare neuroectodermal tumor derived from the choroid plexus epithelial cells, accounting for 0.4%–0.6% of all primary brain tumors. The most frequent sites are the lateral ventricle trigone in children and the fourth ventricle in adults. Extraventricular CPP, which may occur in the subarachnoid space or brain parenchyma, is very rare. Parenchymal CPP may easily be misdiagnosed, and brainstem CPP is extremely rare. Here, we report on a young boy with a pathologically confirmed brainstem CPP.

Case Report

History and Presentation. A 10-year-old boy was admitted to our hospital with a 2-month history of persistent headache and nausea. There was no evidence of or complaint about movement disorder or paresthesia.

Preoperative Imaging Studies. Magnetic resonance imaging showed a mass, about 2.5 × 2.6 × 2.5 cm in size, located anterior to and slightly to the right of the pons (Fig. 1), without any connection to the fourth ventricle. The lesion was uniformly hypointense on T1-weighted images and hyperintense on T2-weighted images, without any evidence of calcification or cystic characteristics. The mass showed no significant enhancement on T1-weighted images obtained after administration of a contrast agent. No space-occupying lesion was found in the ventricular system. The mass was clearly demarcated from the surrounding pontine parenchyma, but was compressing it, causing a thinning of the pontine cortex and flattening of the fourth ventricle and a thinning of the cortex of the pons. There was no obvious peritumoral cerebral edema or occlusion of the right CPA cistern. The radiologists considered low-grade glioma to be the most likely diagnosis.

Operation and Postoperative Course. The pontine tumor was resected through a retrosigmoid approach. Intraoperatively, the tumor was noted to be grayish red with a jelly-like texture and clear boundary. The mass was anterior to and slightly to the right of the pons, surrounded by parenchyma, and protruding into the CPA. Partial resection was performed.

The postoperative pathological examination confirmed the lesion as CPP (Fig. 2). The patient’s symptoms were alleviated by the surgery, and a CT scan performed 3 days postoperatively showed reduction in tumor size and mass effect and restoration of the morphology the fourth ventricle (Fig. 3). The child was discharged on the 5th day after surgery.

Discussion

Choroid plexus papilloma is a rare tumor that develop-
ops from choroid plexus epithelial cells, accounting for 0.4%–0.6% of all primary brain tumors.6,13 It typically occurs in the ventricular system. The lateral ventricle trigone is the most common location in children, whereas the fourth ventricle is more likely to be involved in adults; occurrence in the third ventricle is uncommon. CPP in the extraventricular regions, including the subarachnoid space and brain parenchyma, is very rare. In the posterior fossa, the CPA is the area most susceptible to the development of subarachnoid CPPs,7,9 while the cerebellum is where parenchymal CPP is most likely.17 Extraventricular CPP in the brainstem is extremely rare, and we know of only one previously been reported case.12

The typical imaging appearance of an intraventricular CPP is that of a lobulated mass with a well-defined circumscription and occasional punctate foci of calcification, often associated with expansion of the ventricular system due to communicating hydrocephalus. The lesions are hypointense on T1-weighted MR images, are hyperintense on T2-weighted images, and show marked enhancement on postcontrast T1-weighted images.3,8,17 Extraventricular CPPs generally have similar imaging features to intraventricular ones, including a clear boundary and obvious enhancement on postcontrast T1-weighted MR images.14,17 When the tumor is associated with necrosis or cysts, uneven ring-like enhancement will be evident.

Due to the atypical site of extraventricular CPP, it is generally misdiagnosed as another type of tumor based on preoperative imaging. If the lesion’s density or signal intensity is not quite the same as that of a typical CPP, misdiagnosis is even more likely. In our case, the lesion’s MRI features had some similarities to those of other intraparenchymal CPPs reported in the literature,14,17 including well-defined circumscription, hypointensity on T1-weighted images, and hyperintensity on T2-weighted images. However, the mass showed no enhancement on...
postcontrast T1-weighted images, which is more typical of low-grade glioma. In addition, unlike the 2 cases of parenchymal CPP described by Zhang et al., which caused obvious peritumoral edema, there was no significant cerebral edema in our case. The atypical location of our patient’s tumor and the lack of enhancement on MRI were the main reasons for the initial misdiagnosis, and these atypical characteristics deserve the attention of the clinicians in radiological differential diagnosis.

Two hypotheses have been proposed regarding the pathogenesis of extraventricular CPP. 1) Intracisternal CPP may develop from the implantation or dissemination of a primary intraventricular CPP via the subarachnoid cerebrospinal fluid pathways, or it may originate in a small choroid plexus tuft protruding into the subarachnoid space. 2) Cerebral parenchymal CPP may arise from primitive ectopic choroid plexus epithelial cells in the cerebral parenchyma or may be explained by migration of choroid plexus epithelial cells during brain development. We propose that epithelial cells shed from intraventricular choroid plexus into the Virchow-Robin spaces may seed into the parenchyma via cerebrospinal fluid circulation and eventually form a CPP, although this hypothesis is unconfirmed. As for the present case, we believe that the tumor was likely generated from shed epithelial cells of the fourth ventricle choroid plexus seeding in the Virchow-Robin spaces of pontine perforating arteries and pushing adjacent brainstem along with its growth. Perhaps this growth pattern could explain why this particular lesion demonstrated a clear boundary and no obvious peritumoral edema on MRI. As to the lack of obvious postcontrast enhancement on MRI in our case, we are not quite sure, but it may be related to the status of intratumoral blood supply or degree of blood-brain barrier damage.

The imaging differentiation of a primary pontine CPP from a CPP in the CPA cistern protruding into the pons is very important in forming a surgical plan. Although it is sometimes hard to distinguish these lesions, we suggest that the following MRI features may be helpful. If the tumor is located in the brainstem, the normal brain tissue around it is often compressed on axial, coronal, and sagittal imaging. In addition, lateral shift of the brainstem caused by the mass is not obvious, and the CPA cistern is always partly evident, though narrowed. These features were clearly evident in our case.

Because of its atypical location and imaging features, extraventricular CPP may easily be misdiagnosed. We believe that on MRI, the well-defined circumscription and lack of marked cerebral edema may be useful signs to distinguish brainstem CPP from brainstem astrocytoma, which is characterized by ill-defined margins and obvious peritumoral cerebral edema. Moreover, we believe that the presence of absence of enhancement should not be the main basis for differentiating CPP from other tumors. Even though a lesion does not demonstrate enhancement, the diagnosis of extraventricular CPP still could not be ruled out. Currently, resection with precise navigation is the most effective treatment method.

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: You. Acquisition of data: Xiao. Analysis and interpretation of data: Xu. Drafting the article: Xiao. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: You.

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