Intra-fourth-ventricular choroid plexus papilloma miming ependymoma

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Abstract

Choroid plexus papilloma (CPP), a low incidence central nervous system (CNS) tumor, typically develops as an intraventricular neoplasm arising from the epithelium of the choroid plexus. Infratentorial CPP is predominantly clustered in adults with roughly 70% in the fourth ventricle, while supratentorial CPP commonly found in the lateral ventricles, is the most frequent location in children. The clinical and imaging features of CPP are not typical and may induce the misdiagnosis as other types of primary brain tumors. In this paper, we described a fourth-ventricular CPP that was misdiagnosed as ependymoma despite the manipulation of groundbreaking magnetic resonance imaging (MRI) sequences. These findings indicated that CPP should be considered when performing the differential diagnosis of intraventricular neoplasms. Clin Ter 2021; 172 (2):99-103. doi: 10.7417/CT.2021.2292

Keywords: choroid plexus papilloma, ependymoma, fourth ventricle tumor, children

Introduction

Choroid plexus papilloma (CPP) generally presents as an intraventricular neoplasm that originates from the choroid plexus epithelium and accounts for less than 4% of all intracranial neoplasms in children and fewer than 0.5% of neoplasms in adults (1). CPP is localized infratentorially in adults, with approximately 70% positioned in the fourth ventricle, whereas, in children, the most common site is supratentorial, located in the lateral ventricles (2). Although CPP may occur at any age, 70% of all cases have been identified in patients under the age of 2 (1-4).

Ependymoma refers to a glial cell tumor that commonly develops in cells that line the ventricular system or the central canal, and less often, ependymomas are located outside of the central nervous system (CNS), or intraparen-
Fig. 1. A hyperintense solid tumor, located in the fourth ventricle and extending through left Luschka foramen, on axial T2-weighted imaging.

Fig. 2. Axial apparent diffusion coefficient of the tumor and the parenchyma.
Fig. 3. The detailed perfusion parameters for the tumor (blue ROI) and the parenchyma (orange ROI).

Fig. 4. Histopathological results show that the tumor was composed of papillae, lined by cuboidal to columnar choroid plexus epithelium, with a central fibrovascular core. Nuclear pleomorphism was not observed (H & E, x 40).
Discussion

Choroid plexus tumors are categorized as CPP (grade I), atypical CPP (grade II), and choroid plexus carcinoma (grade III), according to the 2016 World Health Organization (WHO) classification guidelines (7). Microscopic examination reveals that CPP features papillary fronds, lined by bland columnar epithelium. Generally, CPP does not feature mitotic activity, nuclear pleomorphism, or necrosis (4). Ependymomas are classified as subependymoma and myxopapillary ependymoma (grade I), classic ependymoma (grade II), and anaplastic ependymoma (grade III), according to the 2016 World Health Organization (WHO) classification guidelines (7). Among these types, classic ependymoma is the most common.

Unlike most other primary brain neoplasms, which are more common in the infratentorial region in children and the supratentorial compartment in adults, this trend is inverted for CPPs, with 70% of CPPs in adults situated in the fourth ventricle, whereas in children, CPPs are usually located in the lateral ventricles (2). Hence, pediatric CPP in the fourth ventricle, as observed in our case, is quite uncommon. Due to ventricular system obstruction, CPPs almost always cause hydrocephalus, inducing symptoms that include headache, nausea, and vomiting. On imaging, CPP typically appears as a well-circumscribed, lobulated mass, which is hyperintense compared to the adjacent normal-appearing parenchyma. CPPS generally enhance homogeneously and vividly, exhibiting an irregular, frond-like pattern, resulting in a cauliflower-like appearance. The presence of strikingly heterogeneous contrast enhancement or a markedly necrotic appearance indicates choroid plexus carcinoma (4). Speckled ossification is observed inside of the tumor in approximately 25% of CPPs (4). In our case, the cauliflower-like appearance on the T1-perfusion map was not typical (8-11).

The three most well-known and common posterior cranial fossa tumors in children are medulloblastoma, pilocytic astrocytoma, and ependymoma (12,13). Approximately 60% of ependymomas derive from the fourth ventricle, whereas the remainder are supratentorially located, and up to 50% of these are extraventricular and intraparenchymal (5-7,12,13). Ependymomas are often heterogeneous masses, with numerous necrotic regions, calcification, cystic alterations, and hemorrhage. Hydrocephalus presents generally, in most cases of ependymomas. Posterior fossa ependymomas are inclined to spread via the Luschka and Magendie foramina; thus, they are also known as “plastic ependymomas”. This is a typical hallmark that can be effectively observed on both computed tomography (CT) and MRI (12,13).

CPP, in the present report, appeared as a solid mass, situated in the fourth ventricle, and extended via the left Luschka foramen, which is a very common behavior of plastic ependymomas. Even though advanced MRI sequences, such as DWI and T1-perfusion were exploited, this tumor was misdiagnosed as ependymoma, a more common, fourth-ventricular brain tumor found in pediatric patients. Therefore, the imaging characteristics that can be used to differentiate between the CPP and other brain tumors are not specific enough to separate these two tumor types effectively.

The treatment of choice for both CPP and ependymoma is gross surgical tumor excision. However, for cases in which the tumor extends into the Luschka and Magendie foramina or cerebellopontine angle, surgical procedures can be complicated, the complete eradication of tumor tissue might not be possible without disrupting adjacent eloquent structures (12-14).

Conclusion

This report described an atypical CPP, which resulted in the misdiagnosis of ependymoma, despite the employment of advanced MRI protocols. Further studies remain vital to identify imaging features capable of discriminating CPP from other brain neoplasms, which will improve the precision and effectiveness of diagnostic and prognostic outcomes.

Ethical statement

Institutional review board of Children’s hospital 2 approved this prospective study (Ref: 352/NĐ2-CDT). Informed consent of legal guardians of patient was obtained.

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ICMJE Statement and Conflict of interest

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Author’s contribution

TAT and PAT contributed equally to this article therefore considered as co-first authors. NMD and TAT gave a substantial contribution in acquisition, analysis, and data interpretation. NMD and PAT prepared, drafted, and revised manuscript critically for important intellectual content. Each author gave the final approval of the version to be published and agreed to be accountable for all aspects of the work, ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

References