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Case Report

A case report of atypical choroid plexus papilloma in the cervicothoracic spinal cord [☆]

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ABSTRACT

Choroid plexus papillomas (CPPs) are uncommon, benign intracranial tumors that can occur in both children and adults. In adults, CPPs are typically identified in the fourth ventricle, whereas in children, they most commonly occur in the lateral ventricle. CPPs that arise from the extraventricular system are extremely rare and difficult to diagnose. We report a case of extraventricular, atypical CPP located in the cervicothoracic spinal cord of a 2-year-old girl.

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Introduction

Choroid plexus papillomas (CPPs) are benign tumors classified as World Health Organization (WHO) grade I malignancies [1] and account for 0.4%–1% of all intracranial tumors [2,3]. CPPs can be detected in all age groups, but the incidence increases among younger ages, with a median age of 3.5 years at diagnosis [2,4]. CPPs are primary brain tumors that arise from the epithelial cells of the choroid plexus and may occur at any

location containing plexus. In children, CPPs are typically located in the supratentorial compartment, with the lateral ventricle being the most common site. By contrast, in adults, the preferred site is the fourth ventricle [3,5]. The extraventricular detection of a primary CPP is extremely rare, and several sites have been described in the literature, including the cerebello-pontine angle [6], suprasellar region [7], intraparenchymal areas [8,9], and sacral nerve roots [10]. Here, we describe a case of extraventricular CPP arising from the cervicothoracic spinal cord.

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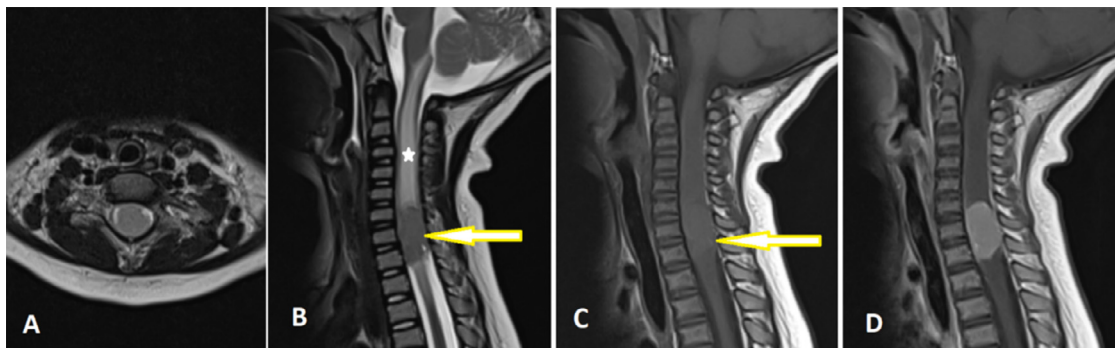


Fig. 1 – (A) Axial T2-weighted (T2W) image of the cervicothoracic spinal cord shows an intramedullary solid mass. (B) Sagittal T2W image revealed the tumor was located at the C7–D2 level (arrow) with syringohydromyelia (star) and was well-defined and slightly hyperintense compared with the spinal cord parenchymal. (C) Sagittal T1-weighted (T1W) image indicated that the lesion (arrow) was isointense compared with the spinal cord parenchyma. (D) Axial contrast-enhanced T1W image demonstrated homogenous enhancement of the tumor.

Case report

The patient was a 20-month-old girl who experienced no significant medical problems during the neonatal period, with normal psychomotor development prior to presentation. At 20 months, her parents noticed abnormal symptoms, such as weakness in the limbs and difficulty turning her head. The symptoms progressed quite rapidly over the course of approximately one month; therefore, she was admitted to the hospital.

Magnetic resonance imaging (MRI) showed an intramedullary, oval-shaped, well-defined solid mass at the C7–D2 levels, and syringohydromyelia was observed on T2-weighted imaging (T2W; Fig. 1). The tumor was isointense on T1-weighted imaging (T1W) and slightly hyperintense on T2W compared with the spinal cord parenchyma, with homogenous enhancement following gadolinium administration (Fig. 1). The location and general appearance of this tumor suggested an initial diagnosis of ependymoma or astrocytoma. The patient also underwent contrast-enhanced MRI of both the brain and the total spine, but no other lesions were detected.

The patient underwent a surgical biopsy, and histologic results confirmed atypical CPP. However, no other lesions were identified, either in the cerebral ventricles or in the spinal cord. Thus, the final diagnosis was ectopic atypical CPP in the cervicothoracic spinal cord. Due to this location, we were unable to remove the tumor, and the patient received adjuvant radiotherapy. The patient died as a result of respiratory distress after 5 months.

Discussion

CPPs are encountered in all age groups but most commonly occur in children [4], with 45% of cases detected within the first year of life and 74% within the first decade [11]. Most CPPs are located in the ventricular system, and Laurence re-

ported that 50% of CPPs occur in the lateral ventricles, 37% in the fourth ventricle, 9% in the third ventricle, and the remainder are typically intracerebral [11]. CPPs that arise outside of the ventricular system are very rare and may occur due to the enlargement of an intraventricular papilloma or drop metastasis along the cerebrospinal fluid pathways from a primary intracranial CPP or develop from ectopic foci of the choroid plexus [12,13]. Sporadic literature reviews describing primary intraspinal cord CPPs have reported occurrence at the sacral nerve roots, sacral canal, or cauda equina [10,14,15]. Because these CPP locations are extremely rare, they are often initially approached as common primary tumors of the spinal cord. Boldorini et al. [10] reported a case of primary CPP located at the sacral nerve roots, with an initial differential diagnosis of ependymoma or schwannoma of the terminal filum. DeMarchi et al. [14] described a case of CPP arising from cauda equina, with a differential diagnosis that included meningioma, neurofibromatosis, schwannoma, and ependymoma.

On MRI, the typical imaging findings of intraventricular CPPs include well-defined, lobulated masses that are homogeneously isointense or slightly hypointense on T1W imaging and hyperintense on T2W imaging [5,16]. After contrast injection, the lesions are often marked enhanced and tend to be homogeneous. In addition, hydrocephalus is frequently detected [5,16]. Shi et al. [17], in addition to the typical imaging findings for extraventricular CPPs, described atypical imaging findings that included rounded or oval shapes that appear hyperintense on T1W imaging, hypointense on T2W imaging, or mixed hyperintense on both T1W and T2W, with poor contrast enhancement and the absence of hydrocephalus.

In this case, an intramedullary tumor located in the cervicothoracic spinal cord was detected in a 20-month-old girl. On MRI, the tumor had some imaging features consistent with CPPs, including isointensity on T1W imaging, hyperintensity on T2W imaging, and homogenous enhancement after contrast administration. By contrast, the oval shape and location were atypical characteristics for CPPs. These imaging features and location make the tumor indistinguishable from ependymoma and astrocytoma, which are the most common

intramedullary tumors that occur in children. Moreover, because extraventricular CPPs are extremely rare, CPP was not considered in the initial diagnosis of this case. The patient underwent brain MRI, which revealed no evidence of other ventricular tumors; therefore, this case was diagnosed as an ectopic CPP.

Conclusion

In this article, we reported a case of extraventricular atypical CPP. Although many imaging features of the tumor were consistent with CPPs, its location in the cervicothoracic spinal cord is extremely rare in the literature.

Author contributions

Nguyen XH, Nguyen MD, Thieu-Thi TM, and Dong VH contributed equally to this article as co-first authors. All authors have read the manuscript and agree to the contents.

Patient consent

Informed consent for patient information to be published in this article was obtained.

Ethical statement

Appropriate written informed consent was obtained for the publication of this case report and accompanying images.

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