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Case Report

Choroid Plexus Carcinoma in a 5-Year-Old Girl: Computed Tomography and Magnetic Resonance Imaging Findings

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Abstract

Choroid plexus carcinoma is a rare, aggressive intraventricular neoplasm that predominantly affects young children. A 5-year-old girl presented with a syndrome of raised intracranial pressure. Computed tomography demonstrated a large lobulated hyperattenuating mass centered in the right lateral ventricle, associated with marked asymmetric obstructive hydrocephalus and mass effect. After contrast administration, the tumor enhanced intensely and heterogeneously. Magnetic resonance imaging showed an irregular heterogeneous intraventricular mass with internal non-enhancing areas, extensive peritumoral edema, heterogeneous low apparent diffusion coefficient foci, and avid heterogeneous gadolinium enhancement. These findings favored a malignant choroid plexus tumor. The patient underwent subtotal resection. Initial histologic assessment favored atypical choroid plexus papilloma; however, neuropathologic reassessment integrating morphology and immunohistochemistry established choroid plexus carcinoma, central nervous system World Health Organization grade 3. Tumor cells showed diffuse cytokeratin expression, S100 and vimentin positivity, negative p53 staining, and a Ki-67 proliferation index of approximately 60%. This case highlights imaging features that should raise suspicion for carcinoma rather than a lower-grade choroid plexus tumor and illustrates the value of integrated radiologic-pathologic assessment when the initial diagnosis is equivocal.

Keywords: *Choroid Plexus Carcinoma; Pediatric Brain Tumor; Intraventricular Tumor; Hydrocephalus; Magnetic Resonance Imaging*

Introduction

Choroid plexus tumors are uncommon neuroepithelial neoplasms arising from the choroid plexus epithelium and are classified as choroid plexus papilloma, atypical choroid plexus papilloma, and choroid plexus carcinoma. Choroid plexus carcinoma is a central nervous system World Health Organization grade 3 tumor and occurs mainly in infants and young children. Imaging plays a central role in suggesting malignancy, defining local extension and hydrocephalus, planning surgery, and assessing neuraxis dissemination. The objective of this report is to describe the computed tomography and magnetic resonance imaging features of a pediatric choroid plexus carcinoma and to emphasize the features that favored carcinoma despite an initially less aggressive pathologic impression [1-3].

Case Report

A 5-year-old girl was admitted with a clinical syndrome of raised intracranial pressure. Non-contrast computed tomography of the brain showed a large lobulated and heterogeneous hyperattenuating mass centered in the right lateral ventricle. The lesion produced marked asymmetric obstructive hydrocephalus, compression of the adjacent cerebral parenchyma, and substantial mass effect. Contrast-enhanced computed tomography demonstrated intense heterogeneous enhancement with internal non-enhancing areas and conspicuous intratumoral vessels (Figure 1).

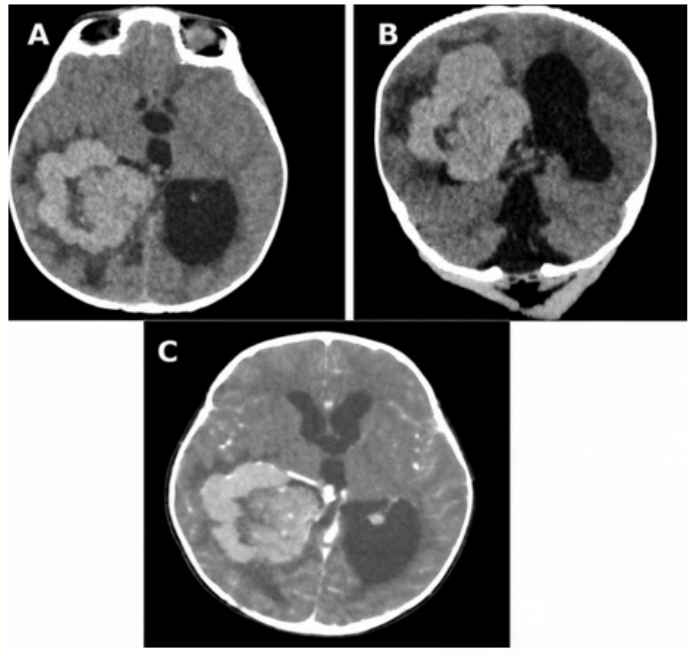

Figure 1: *Brain CT. Axial unenhanced (A) and coronal unenhanced (B) images show a large lobulated heterogeneous hyperattenuating mass centered on the right lateral ventricle, with marked asymmetric obstructive hydrocephalus and mass effect. Axial contrast-enhanced CT (C) demonstrates intense heterogeneous enhancement, internal non-enhancing areas, and prominent intratumoral vessels.*

Magnetic resonance imaging confirmed a large irregular intraventricular tumor. On fluid-attenuated inversion recovery imaging, the mass was heterogeneous and was associated with extensive adjacent parenchymal edema and severe hydrocephalus. The lesion contained solid and non-enhancing components, with heterogeneous low apparent diffusion coefficient areas in the solid tumor. Post-gadolinium T1-weighted imaging demonstrated avid heterogeneous enhancement. The irregular tumor-parenchyma interface, marked edema, necrotic-appearing areas, and heterogeneous restricted diffusion favored a malignant choroid plexus tumor rather than a conventional papilloma (Figure 2).

The patient underwent subtotal surgical resection. Initial histologic assessment favored an atypical choroid plexus papilloma. Subsequent neuropathologic reassessment, integrating morphology with immunohistochemical findings, established the diagnosis of choroid plexus carcinoma, central nervous system World Health Organization grade 3. The tumor showed diffuse cytokeratin expression, S100 and vimentin positivity, negative p53 staining, and a high Ki-67 proliferation index of approximately 60%.

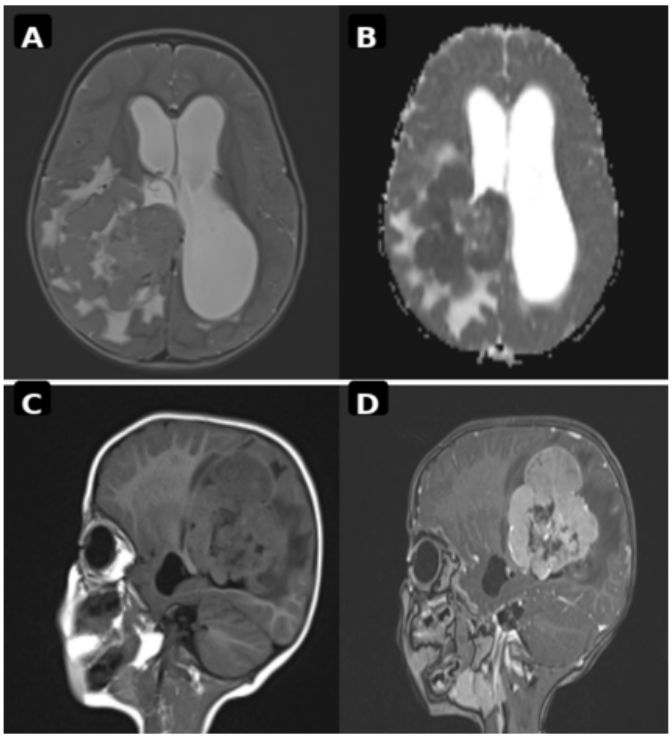

Figure 2: *Brain MRI. Axial FLAIR (A) and sagittal T1-weighted (B) images depict a large lobulated intraventricular mass with heterogeneous signal, surrounding edema, marked hydrocephalus, and mass effect. Sagittal post-gadolinium T1-weighted imaging (C) shows intense heterogeneous enhancement. The ADC map (D) demonstrates heterogeneous low-ADC areas within the solid tumor component.*

Discussion

Choroid plexus carcinoma represents the malignant end of the choroid plexus tumor spectrum. In children, these tumors most commonly arise in the lateral ventricles. Hydrocephalus is frequent and may result from cerebrospinal fluid overproduction, obstruction of cerebrospinal fluid pathways, impaired resorption, or a combination of these mechanisms. On computed tomography, choroid plexus tumors are often hyperattenuating and strongly enhancing because of their cellularity and vascularity. Magnetic resonance imaging better characterizes tumor heterogeneity, necrosis, edema, invasion, and dissemination [1,2].

The distinction between papilloma, atypical papilloma, and carcinoma cannot always be made confidently by imaging alone. Nevertheless, several features increase suspicion for carcinoma: large size, irregular or infiltrative margins, heterogeneous signal and enhancement, necrosis or hemorrhage, extensive peritumoral edema, parenchymal invasion, and cerebrospinal fluid dissemination. Conventional papillomas are more often well-circumscribed, frond-like, and relatively homogeneous, although overlap is substantial. In the present case, the irregular lobulated appearance, prominent edema, internal non-enhancing areas, and heterogeneous low apparent diffusion coefficient foci were important malignant clues and supported pathologic reassessment [1,2].

Histopathologic diagnosis is based primarily on morphology, including architectural loss, increased cellularity, brisk mitotic activity, necrosis, and brain invasion. Immunohistochemistry supports epithelial differentiation and helps exclude mimics, but no single marker replaces morphologic assessment. A high Ki-67 index is compatible with an aggressive proliferative tumor. The initial interpretation as atypical choroid plexus papilloma in this case illustrates the recognized diagnostic overlap within the choroid plexus tumor spectrum and the importance of correlating pathology with the radiologic pattern [3].

Management is multidisciplinary. Maximal safe resection is the cornerstone of treatment, and greater extent of resection is associated with improved survival. Complete resection may be limited by the tumor's marked vascularity, adherence to critical structures, or invasion of adjacent brain. Adjuvant treatment is individualized according to the patient's age, residual tumor, dissemination, molecular or genetic context, and institutional protocol. Because leptomeningeal spread may be present at diagnosis or develop later, craniospinal magnetic resonance imaging and appropriate clinical surveillance are important components of staging and follow-up [4-6].

Differential diagnosis

The principal differential diagnoses for a large pediatric intraventricular mass include lower-grade choroid plexus tumors, ependymoma, and embryonal tumors. The site of origin, tumor margins, degree of heterogeneity, edema, invasion, and enhancement pattern are useful for narrowing the diagnosis, but histopathologic confirmation remains necessary.

Diagnosis	Typical imaging pattern	Features favoring or arguing against the diagnosis in this case
Choroid plexus carcinoma	Large irregular intraventricular mass; heterogeneous enhancement; necrosis or hemorrhage; edema, invasion, and possible dissemination.	Favored by irregular margins, marked edema, internal non-enhancing areas, heterogeneous low ADC, and pathologic confirmation.
Atypical choroid plexus papilloma	Lobulated plexus-centered mass with avid enhancement; imaging overlap with both papilloma and carcinoma.	Initial histologic consideration, but aggressive imaging features and high proliferative index favored carcinoma.
Choroid plexus papilloma	Well-circumscribed frond-like or cauliflower-like mass with intense, often relatively homogeneous enhancement; hydrocephalus is common.	Extensive edema, necrotic-appearing areas, irregular interface, and high Ki-67 were atypical for a conventional papilloma.
Ependymoma	Ventricular wall-based heterogeneous mass with cysts, calcification, and variable enhancement; often arises in the fourth ventricle in children.	The lesion was centered on the lateral ventricular choroid plexus and showed marked vascular enhancement.
Atypical teratoid/rhabdoid or other embryonal tumor	Heterogeneous pediatric mass with necrosis, hemorrhage, and restricted diffusion; may be intraventricular or parenchymal.	Considered because of restricted diffusion and heterogeneity, but plexus-centered origin and epithelial choroid plexus pathology supported carcinoma.

Table 1: *Imaging differential diagnosis of a pediatric intraventricular mass.*

Conclusion

In a young child, a large irregular plexus-centered intraventricular mass with heterogeneous enhancement, necrotic-appearing areas, extensive edema, and restricted diffusion should raise strong suspicion for choroid plexus carcinoma. Radiologic-pathologic correlation is especially valuable when the initial histologic diagnosis is equivocal.

Teaching Points

- Marked peritumoral edema, irregular invasive-appearing margins, necrosis, and heterogeneous diffusion restriction favor choroid plexus carcinoma over a conventional papilloma.
- Choroid plexus tumors are highly vascular; computed tomography and magnetic resonance imaging are essential for defining vascularity, hydrocephalus, local extension, and surgical risk.
- Discordance between aggressive imaging and an initially lower-grade pathologic diagnosis should prompt integrated radiologic-pathologic review.

MCQs

1. Which imaging feature most strongly favors choroid plexus carcinoma over a conventional choroid plexus papilloma?

A. Smooth frond-like contour

B. Relatively homogeneous enhancement

C. Extensive edema with an irregular invasive-appearing margin

D. Absence of hydrocephalus

Answer key: C

2. In young children, choroid plexus carcinoma most commonly arises in which location?

A. Lateral ventricle

B. Fourth ventricle

C. Cerebellopontine angle

D. Pineal region

Answer key: A

3. Choroid plexus carcinoma is classified as which central nervous system World Health Organization grade?

A. Grade 1

B. Grade 2

C. Grade 3

D. Grade 4

Answer key: C.

Ethical Approval

Institutional Review Board approval was not required or was waived for this single anonymized case report, according to local institutional policy.

Declaration of Patient Consent

The authors certify that written informed consent for publication of the anonymized clinical information and images was obtained from the patient's parent or legal guardian.

Conflicts of Interest

There are no conflicts of interest.

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Generative artificial intelligence assisted with language editing. The authors verified all content, and no artificial intelligence tool was used to create, alter, or interpret the medical images.

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