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RADIOLOGIC DIFFERENTIAL OF A THIRD-VENTRICULAR MASS IN A SEVEN-MONTH-OLD INFANT: CHOROID PLEXUS TUMOR VERSUS CRIBRIFORM NEUROEPITHELIAL TUMOR

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AUTHOR AND AFFILIATION

Lina Lasri[1], Samia Obilat[1], Alia Yassine Kassab[1], Lina Belkouchi[1], Siham El Haddad[1], Nazik Allali[1], Latifa Chat[1]

[1] Department of Pediatric Radiology, Children's Hospital of Rabat, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco

Corresponding author: Lina Lasri

ABSTRACT

Intraventricular tumors in infants constitute a narrow but clinically important differential diagnosis. We report a seven-month-old boy who presented with progressive macrocephaly, a bulging anterior fontanelle, and generalized hypotonia. Brain magnetic resonance imaging showed marked supratentorial ventriculomegaly and a well-circumscribed, lobulated mass centered on the third ventricle. The lesion was isointense to gray matter on T1-weighted imaging, intermediate in signal on T2-weighted imaging, did not restrict diffusion, and enhanced avidly and heterogeneously after gadolinium administration. Susceptibility-sensitive imaging showed internal hemorrhagic foci. Proton magnetic resonance spectroscopy demonstrated elevated choline with markedly reduced N-acetylaspartate and creatine. A choroid plexus tumor was favored radiologically, with carcinoma considered because of the hemorrhage and heterogeneous enhancement; cribriform neuroepithelial tumor was the principal alternative diagnosis. Histopathologic confirmation and follow-up data were unavailable. This case illustrates how multiparametric magnetic resonance imaging can narrow the differential diagnosis of an infantile intraventricular mass while emphasizing that imaging cannot distinguish these entities definitively without tissue-based assessment.

KEYWORDS

choroid plexus tumor; cribriform neuroepithelial tumor; third ventricle; infant; magnetic resonance spectroscopy

MAIN ARTICLE

INTRODUCTION

Intraventricular masses in infancy include choroid plexus tumors, ependymal tumors, atypical teratoid/rhabdoid tumor, and other rare embryonal neoplasms. Age, ventricular location, morphology, diffusion characteristics, enhancement, susceptibility, and metabolic profile can substantially narrow this differential, but definitive classification increasingly depends on histologic and molecular findings [1,2].

Choroid plexus tumors are uncommon overall but occur predominantly in young children. They comprise choroid plexus papilloma, atypical choroid plexus papilloma, and choroid plexus carcinoma. Pediatric lesions most often arise in a lateral ventricle; third-ventricular origin is distinctly uncommon [2-4]. Cribriform neuroepithelial tumor (CRINET), a provisional entity in the 2021 World Health Organization classification of central nervous system tumors, is a rare SMARCB1-deficient intraventricular tumor that can mimic a choroid plexus neoplasm [2]. We present an imaging-based diagnostic approach to a third-ventricular mass in a seven-month-old infant.

CASE PRESENTATION

A seven-month-old boy with no relevant medical history was admitted for progressive enlargement of the head. His occipitofrontal circumference was 50 cm, approximately three standard deviations above the expected value for age. Examination showed a tense, bulging anterior fontanelle and generalized hypotonia, suggesting raised intracranial pressure. Brain magnetic resonance imaging was performed.

The examination demonstrated marked supratentorial ventriculomegaly and a midline mass centered on the third ventricle. The lesion was well defined and roughly ovoid, with microlobulated, papillary contours. It was isointense to gray matter on T1-weighted images and intermediate in signal intensity on T2-weighted images. No diffusion restriction was reported. After gadolinium administration, the mass enhanced avidly and heterogeneously. Gradient-echo susceptibility imaging showed internal foci of signal loss interpreted as hemorrhagic change.

Single-voxel proton magnetic resonance spectroscopy demonstrated elevated choline and markedly reduced N-acetylaspartate at short echo time. At long echo time, choline remained dominant, with very low N-acetylaspartate and creatine, resulting in an increased choline-to-creatine ratio. These findings supported a high-turnover neoplastic process but were not tumor-

specific. The leading radiologic diagnosis was a choroid plexus tumor, with carcinoma considered because of the hemorrhagic component and heterogeneous enhancement. CRINET was retained as the principal differential diagnosis. Tissue diagnosis was recommended; however, histopathologic, therapeutic, and follow-up information was not available for this report.

DISCUSSION

Radiologic reasoning

The combination of infancy, an intraventricular location, a papillary or frond-like surface, and avid enhancement strongly suggests a choroid plexus tumor [1,3,4]. The third ventricle is an unusual location, although pediatric cases have been reported [3,5]. Hydrocephalus may result from obstruction of cerebrospinal fluid pathways and, in some choroid plexus tumors, increased cerebrospinal fluid production.

Imaging cannot reliably grade a choroid plexus tumor. Features that increase concern for carcinoma include irregular or invasive margins, heterogeneous signal and enhancement, necrosis, hemorrhage, adjacent edema, and leptomeningeal dissemination [3,4]. In this patient, hemorrhagic change and heterogeneous enhancement were concerning, but the lesion was well circumscribed, no invasion or dissemination was documented, and diffusion restriction was absent. Consequently, carcinoma can be proposed as a radiologic possibility but cannot be asserted without histopathology.

Contribution and limitations of magnetic resonance spectroscopy

Pediatric choroid plexus tumors can show high choline with very low or absent N-acetylaspartate and creatine. Higher choline and lactate have been described in carcinoma, whereas a prominent myo-inositol peak may favor papilloma [6,7]. In the present case, elevated choline and reduced N-acetylaspartate and creatine supported a neoplasm with increased membrane turnover. These findings do not independently establish malignancy and overlap with other pediatric brain tumors; therefore, the spectroscopy result should be integrated with conventional imaging rather than used as a stand-alone discriminator [6,7].

CRINET as the principal differential diagnosis

CRINET occurs in young children and typically arises within or near the ventricular system, including the third and fourth ventricles. Histologically, it consists of non-rhabdoid primitive cells arranged in cribriform and trabecular patterns, with loss of nuclear INI1 expression due to SMARCB1 alteration [8-10]. Molecular profiling has shown similarity to the ATRT-TYR

subgroup, although reported CRINET outcomes are comparatively favorable [9]. Because its imaging appearance may overlap with that of a choroid plexus tumor, the distinction requires histopathology, INI1/SMARCB1 assessment, and, when available, molecular profiling.

Need for integrated diagnosis

The 2021 World Health Organization framework emphasizes integrated histologic and molecular diagnosis [2]. For a suspected choroid plexus carcinoma, tissue evaluation establishes the tumor type and grade, while assessment of TP53 is clinically important because of the association with germline TP53 alterations and Li-Fraumeni syndrome [11]. Conversely, complete loss of nuclear INI1 expression supports a SMARCB1-deficient tumor such as CRINET in the appropriate morphologic setting [8-10]. Imaging is therefore essential for localization, operative planning, and staging, but it cannot replace tissue diagnosis in this differential.

Limitations

This report is limited by the absence of histopathologic confirmation, treatment details, and clinical or imaging follow-up. The susceptibility and spectroscopy findings were available from the original radiology report, but the corresponding source images were not available for inclusion in the figure. The manuscript therefore presents a radiologic differential diagnosis rather than a confirmed tumor diagnosis.

CONCLUSION

In an infant with macrocephaly and raised intracranial pressure, a papillary, avidly enhancing third-ventricular mass should prompt consideration of a choroid plexus tumor. Hemorrhage and heterogeneous enhancement may raise concern for carcinoma, but they are not diagnostic, particularly when invasion and diffusion restriction are absent. CRINET is a rare but important mimic in this age group and location. Histopathologic and molecular evaluation remains mandatory for definitive classification.

Learning Points

• A papillary, strongly enhancing intraventricular mass in an infant most strongly suggests a choroid plexus tumor.

• Hemorrhage and heterogeneous enhancement may increase suspicion for carcinoma but cannot establish tumor grade.

• CRINET is a rare SMARCB1-deficient ventricular mimic that requires tissue-based diagnosis.

• Spectroscopy supports tumor characterization but must not be interpreted as a histologic diagnosis.

FIGURES

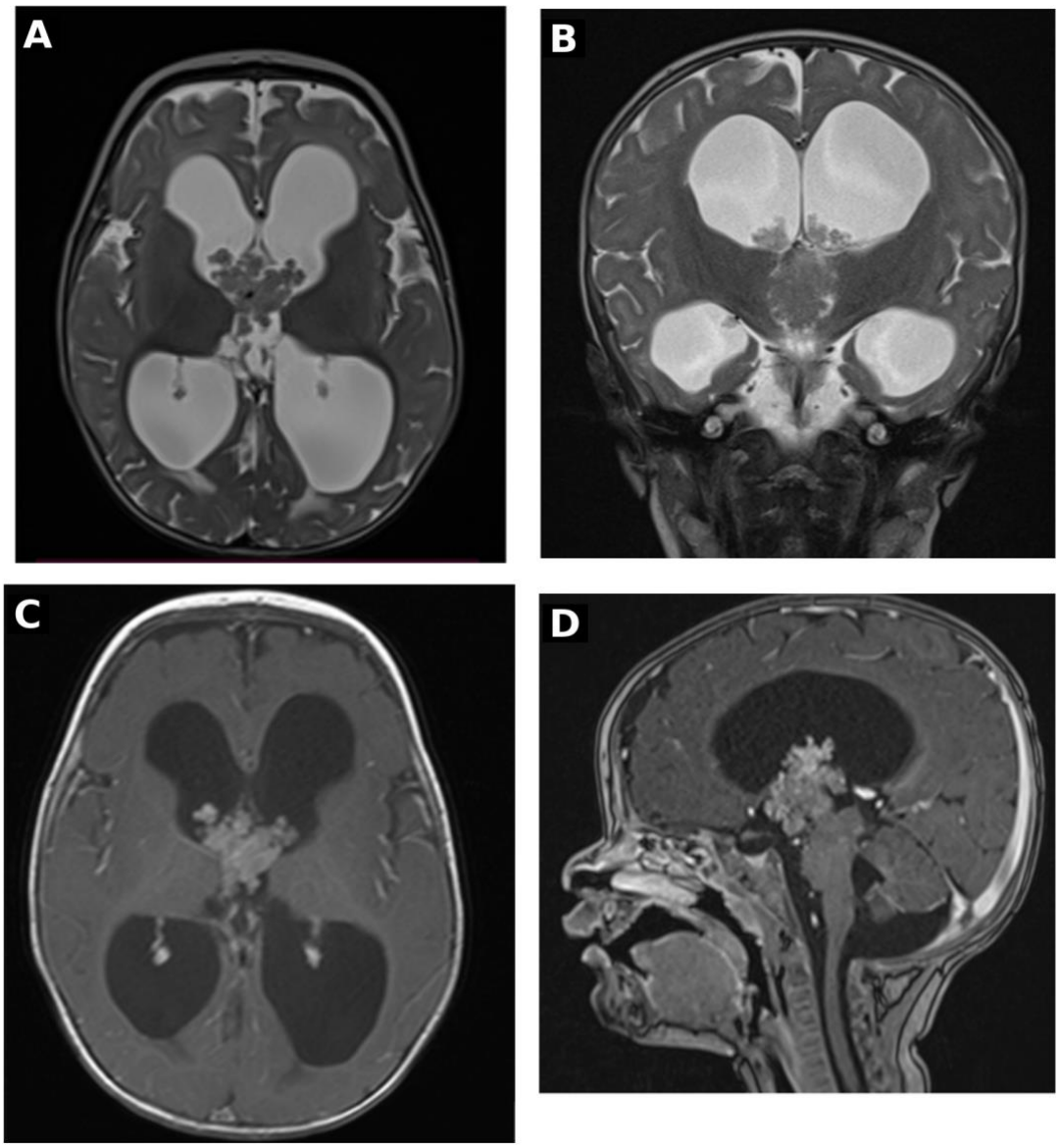

Figure 1. Magnetic resonance imaging of the third-ventricular mass. Axial (A) and coronal (B) T2-weighted images show a lobulated intraventricular mass centered on the third ventricle, associated with marked supratentorial ventriculomegaly. Axial (C) and sagittal (D) post-gadolinium T1-weighted images demonstrate avid heterogeneous enhancement and a papillary, frond-like configuration.

DECLARATIONS

Consent. Written informed consent for publication of this case and the anonymized images was obtained from the child's legal guardians.

Conflicts of interest. The authors declare no conflicts of interest.

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