Radiology Case Reports 19 (2024) 6112–6116

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Obstructive hydrocephalus due to choroid plexus carcinoma of third ventricle in pediatric: A rare case report ☆

Ferdina Widokartika Ashri, MD[a,*], *Harry Galuh Nugraha, MD*[a], *Raisa Mahmudah, MD*[a], *Birgitta Maria Dewayani, MD*[b]

[a] *Department of Radiology, Faculty of Medicine, Universitas Padjadjaran, Dr. Hasan Sadikin General Hospital, Bandung, Indonesia.*

[b] *Department of Anatomical Pathology, Faculty of Medicine, Universitas Padjadjaran, Dr. Hasan Sadikin General Hospital, Bandung, Indonesia*

ARTICLE INFO

Article history:
Received 13 August 2024
Revised 22 August 2024
Accepted 23 August 2024

Keywords:
Choroid plexus carcinoma
Choroid plexus of the third ventricle
Obstructive hydrocephalus
Pediatric brain tumor

ABSTRACT

Choroid plexus carcinoma (CPC) is an uncommon tumor that accounts for less than 1% of all pediatric brain tumors. CPC usually originates in the lateral ventricle, followed by the fourth ventricle; the incidence in the third ventricle is only 5% of all CPC cases (children and adults). We report an extremely rare tumor arising from the choroid plexus of the third ventricle in a 6-year-old child with progressive headache, macrocephaly, left hemiparesis, and sunset eyes. The imaging found a well-defined, lobulated mass with strong enhancement in the posterior part of the third ventricle, resulting in obstructive hydrocephalus. The patient underwent an endoscopic biopsy and histopathological examination, which resulted in choroid plexus carcinoma.

© 2024 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

Introduction

Choroid plexus carcinoma (CPC) is a rare neuroepithelial malignant tumor originating in the choroid plexus, classified as a WHO grade 3 tumor [1,4]. It primarily affects children, usually in their first 2 years of life, and accounts for fewer than 1% of all pediatric brain tumors [1,2,4,11]. CPC mostly begins in atria of the lateral ventricle, followed by the fourth ventricle, with only 5% of CPC cases arising in the third ventricle [2]. We present an extremely unusual tumor that emerges from the third ventricle's choroid plexus causing obstructive hydrocephalus.

Case presentation

A 6-year-old boy was referred to our emergency department with a history of progressive headaches for 2 years and left

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: dr.ferdinawa@gmail.com (F.W. Ashri).

https://doi.org/10.1016/j.radcr.2024.08.127

1930-0433/© 2024 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

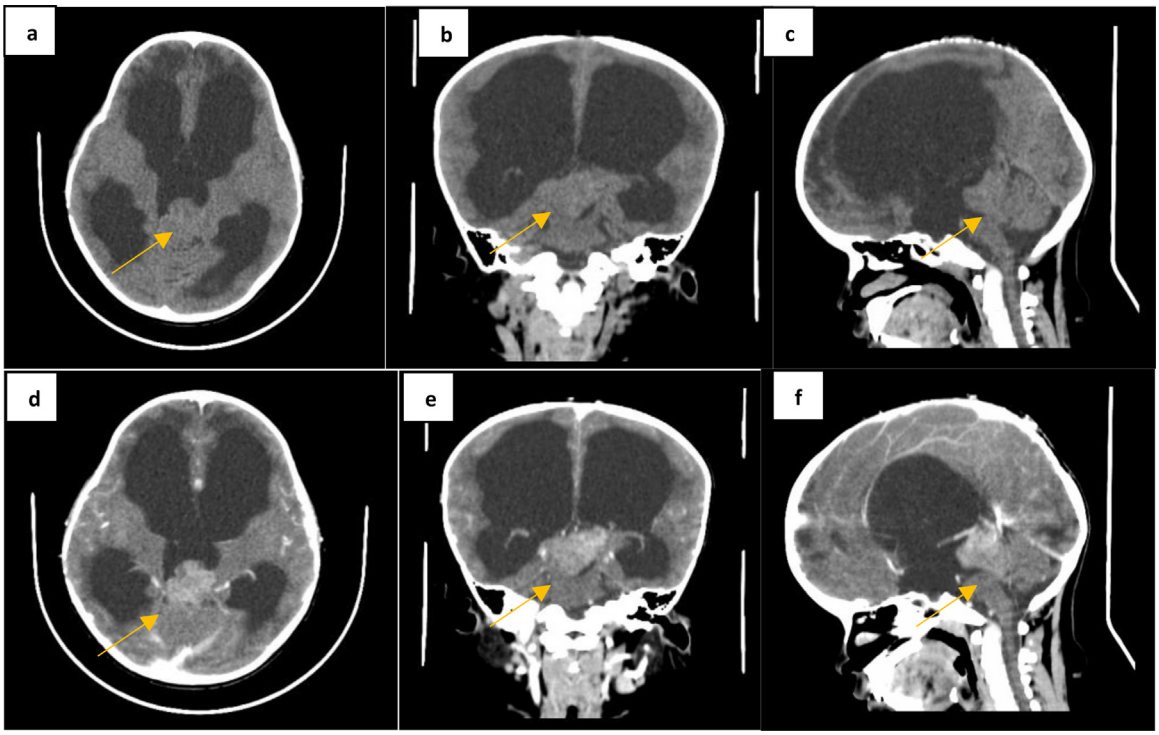

Fig. 1 – (A-C) Axial-coronal-sagittal NECT head and (D-F) CECT head showed a solid mass (yellow arrow) arising in the posterior third ventricle that obstructed the cerebral aqueduct and causing hydrocephalus.

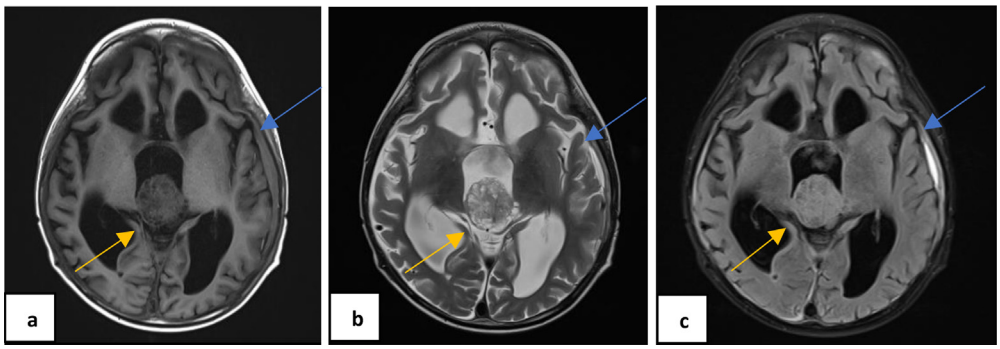

Fig. 2 – The MR Imaging showed a mass (yellow arrow) in third ventricle with hypointense signal on T1W1 (A), slight hyperintense signal on T2W1 (B), no supression on T2-FLAIR (C) and subdural hygroma (blue arrow).

hemiparesis for 1 year. He had no previous seizures, fever, or trauma. Physical examination showed that the patient was conscious (GCS E4V5M6); both pupils were bilateral normal size and reactive. He had macrocephaly (Head circumference was 57 cm, greater than >99 percentile) with an open fontanella anterior, frontal bossing, and sunset eyes. There was also left-side hemiparesis (power MRC grade 5/2) and paresis of the left III, IV, and VI cranial nerves.

In noncontrast computed tomographic (CT) scanning, there was a lobulated isodense mass in the posterior third ventricle that infiltrated the left lateral ventricle and obstructed the cerebral aqueduct, causing hydrocephalus. These solid tumor was enhancing in post contrast CT scan (Fig. 1).

Due to progressive unresponsiveness of patient during course of admission, he was initially treated with an emergent ventriculoperitoneal shunting (VPS) and endoscopic biopsy procedure after head CT scan which led to significant improvement in patient symptoms. Following MRI evaluation had done after VPS installation (Fig. 2), the lesion appeared as hypointense lobulated mass relative to white matter on both noncontrast T1 and T2, slightly hyperintense in T2 and no suppression on T2FLAIR, measuring approximately 32.4 × 30.2 × 21.1 mm in the third ventricle causing significant ventriculomegaly. There was a dead space filled with CSF at left temporal area, which formed a subdural hygroma.

This mass was located in the posterior part of the third ventricle and seen arising from the roof of the ventricle. The mass

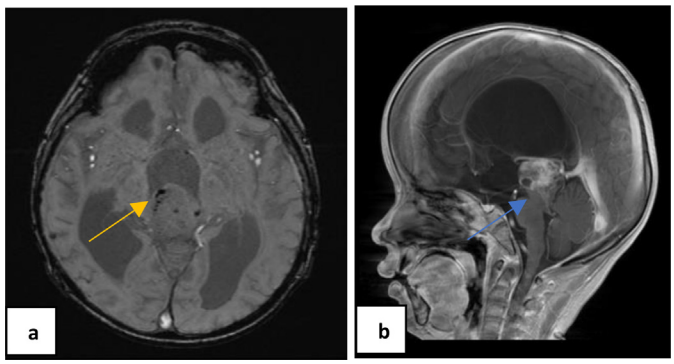

Fig. 3 – The SWI sequence (A) showed multiple signal loss (yellow arrow) explaining the hemorrhage and a heterogenous enhancement in post contrast (B) (blue arrow).

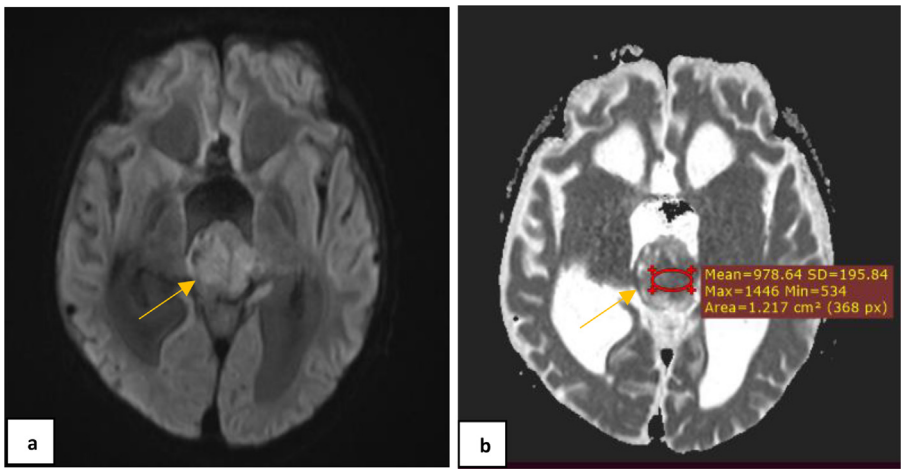

Fig. 4 – The tumor (yellow arrow) showed restricted diffusion in DWI (A)-ADC (B) with ROI taken in the solid part of the mass in ADC map revealed mean ADC value 978 x 10^{-6} mm^2/s.

showed signal loss in SWI indicating microhemorrhages, with significant heterogenous strong enhancement on contrast-enhanced T1 images. There was no invasion of the surrounding brain parenchyma (Fig. 3).

Diffusion weighted imaging (DWI) and apparent diffusion coefficient (ADC) showed restricted diffusion that explained the hypercellularity (Fig. 4) with mean ADC value 0.978 $\times$ 10^{-3} mm^2/s. The patient was planned to have chemotherapy and radioteraphy.

Pathologic examination was performed on the patient's tumor tissue from endoscopic biopsy. On macroscopic examination, the tumor appears brownish-white and rubbery. Microscopic examination (Fig. 5) showed large cells with polygonal shape, high cellularity, hyperchromatic nuclei and nuclear pleomorphism, frequent mitoses, necrosis, and microhemorrhage [7]. The pathological evaluation supported choroid plexus carcinoma at the third ventricle (WHO grade III).

Discussion

Choroid plexus tumors (CPTs) are extremely rare tumors. They account for 0.3% to 0.6% of all cerebral tumor cases [2,5,7,10]. According to the most recent WHO categorization system in 2021, CPTs are divided into 3 subtypes: choroid plexus papilloma (CPP, grade I), atypical choroid plexus papilloma (aCPP, grade II), and choroid plexus carcinoma (CPC, grade III). Approximately 80% of all CPTs were CPPs, 15% were aCPPs, and less than 5% were CPCs [5]. CPTs can be discovered in patients of all ages; about 70%-80% occur in children, especially in the first 2 years of life [1,2,10].

In this case, we reported a 6-year-old boy patient that came with macrocephaly due to hydrocephalus obstructive. After imaging, we concluded a mass within third ventricle and confirmed as plexus choroid carcinoma by histopathology examination.

CPTs develop from the choroid plexus epithelium and can appear anywhere this tissue exists [11]. The normal choroid plexus is a lobulated structure that arises from the walls of the ventricles. It has a central highly vascularized stroma surrounded by neuroectoderm-derived cells [11]. Based on the quantity of choroidal tissue present, CPTs in children are often located inside the ventricular system, with the most common site being the atrium of the lateral ventricle, followed by the fourth ventricle and the third ventricle [5,7,9,10]. In contrast, the majority of tumors in adult instances are located in the fourth ventricle [5,7,9,10]. In the lateral ventricle, the tu-

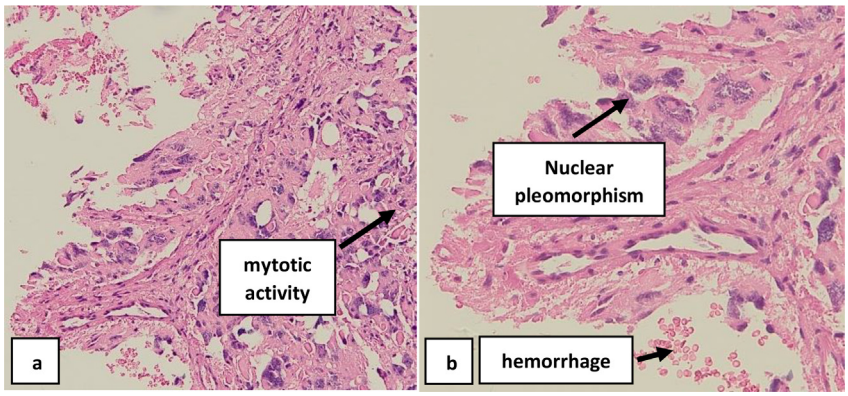

Fig. 5 – Histopathological examination showed tumor component in H-E stain; original magnification x100 (A) and original magnification x200 (B), both described hypercelullarity, nuclear pleomorphism, large cell with bizzare nucleus, mytotic activity, and hemorrhage.

mor is usually attached to the choroid plexus in the inferior portion of the trigone, to the roof of third ventricle and arise along the posterior medullary velum of the fourth ventricle [2]. The third ventricle is an infrequent location for CPTs. Additionally, extraventricular regions such the sacral canal, pineal gland, brain parenchyma, suprasellar, cerebellopontine angle, and Luschka foramen are rare sites where it can occur [5,8,10]. Tavallaii et al. [8] reported that there are 55 cases of reported CPPs and just 3 cases of CPC located in the third ventricle, which elicits the extreme rarity of third ventricle CPC.

Hereditary syndromes such as LiFraumeni syndrome or Aicardi syndrome may coexist with CPC [7,11]. The symptoms of high intracranial pressure and hydrocephalus have been associated with these malignancies [1,3]. Combinations of obstructive hydrocephalus, elevated CSF production, and reduced CSF reabsorption at the arachnoid granulations result in hydrocephalus in CPTs. [2] Hydrocephalus, which presents as macrocephaly, splaying of the cranial sutures, fontanel widening/bulging, and forced downward gaze (sometimes called sunset eyes), is a common presentation in the pediatric population [1].

The radiological diagnosis is based on a CT scan and an MRI. CPC usually show an iso to hyperdense appearance on brain CT scans as a result of micro-hemorrhages and micro-calcifications [7,8]. In 20%-25% of individuals, intralesional calcification may be seen [7].

In our case MR imaging found a solid tumor that hypointense in T1WI, slight hyperintense in T2WI, no suppression on T2FLAIR and heterogenous enhancement in post contrast, without local invasion of brain parenchyma. The MRI characteristics of CPC are heterogeneous because of hemorrhage, necrosis, edema, and intra-atumoral cyst [1,7]. These tumors frequently infiltrate the surrounding brain parenchyma and metastasize through the cerebrospinal fluid [1,7]. Regardless of pathogenic degree, the CPTs were typically slightly hypo- or isointense on T1WI, slightly hyper- or isointense on T2WI, and moderately or substantially enhanced in postcontrast imaging [5]. In CPTs, mild hypointense on T2WI was uncommon unless there were extensive, thick calcification or persistent hemorrhage regions. Separating these probabilities would require a CT or SWI sequencing scan [5]. It has been found that signal characteristics and enhancement patterns cannot differentiate between benign and malignant CPTs [1,5,7]. The imaging features of choroid plexus carcinoma are summarized in Table 1.

Table 1 – Summary of choroid plexus carcinoma imaging features [1,6,8].

Characteristic: Calcification (Rare in children, 14%-25% of all cases in age), necrosis, microhemorrhage, parenchymal invasion.	
NECT	Iso to Hyperdense
T1WI	Slightly hypointense/isointense
T2WI	Isointense/slightly hyperintense
FLAIR	No suppression
DWI-ADC	Restricted Diffusion
T1Gd	Strong and heterogeneous enhancement
MRS	Absent NAA, increase Cr, Cho, Lac

The ADC value of tumor in our case was 0.978×10^{-3} mm^2/s. A lower ADC, indicating increased mitotic activity, could be useful for identifying aCPP or CPC with a poor prognosis [13]. A mean cutoff ADC $\leq 1.397 \times 10^{-3}$ mm^2/s and a tumor volume ≥ 21.05 mL predicted significantly poorer survival ($P = .001$, and $P = .031$, respectively) [13].

Horská et al. investigated pediatric intraventricular lesions using MR spectroscopy. Significant amounts of choline-containing substances are present in papilloma and choroid plexus cancer, while creatine and the neuronal/axonal marker N-acetyl aspartate are absent. Conversely, the levels of lactate and choline in the choroid plexus carcinoma were greater than those in the choroid plexus papilloma. In these circumstances, MRS may provide additional diagnostic information, although it is neither precise nor sensitive [12]. Angiographically, these tumors are hypervascular, with numerous small, tortuous vessels seen in the arterial phase and a homogenous blush in the venous phase [2].

CPP (choroid plexus papilloma) and atypical CPP (atypical choroid plexus papilloma) are the main differential diagnoses [1]. All 3 of the primary choroid plexus cancers have overlapping imaging characteristics [1,5]. CSF spread occurs in both

benign and malignant forms. CPP seldom invades the brain; hence, frank parenchymal invasion with concomitant edema indicates CPC [1]. Other brain tumors that can be considered in the differential diagnosis are astrocytoma, ependymoma, primitive neuroectodermal tumor, atypical teratoid/ rhabdoid tumor, tumors of the pineal origin, papillary meningioma, cerebellar medulloblastoma and metastatic papillary tumor, such as gastrointestinal tract, kidney, bladder, pancreas and thyroid gland [1,2,7].

CPCs exhibit malignant histological characteristics such as high cellular density, pleomorphism, reduced papillary pattern, necrosis, and invasion of surrounding parenchyma [7]. Microscopically, CPPs are composed of well-structured fibrovascular papillary formations lined by a single layer of columnar or cuboidal epithelium without malignant features [7]. In contrast, CPCs are recognized with the presence of at least 4 of 5 malignant features such as increased cellularity, high mitotic activity (more than 5 in 10 HPF), multiple areas of necrosis, highly pleomorphic nuclei and blurring of papillary architecture [8].

CPCs had 5-year survival rates of approximately 26% [14]. Craniospinal dissemination in CPC may be as high as 12%-30% [11]. Surgical resection is seen to be the most effective treatment for CPC [3]. Patients with CPC who receive solely surgical treatment have extremely poor outcomes: the disease advances quickly and patients frequently pass away within a year [3]. Nevertheless, the amount of surgical resection continues to be the most critical determinant in determining long-term survival. Radiation therapy administered early may increase survival [3]. Unfortunately, due to the early age of the patients and the size of the field that needs to be treated, radiation is not an option in most situations [3]. While chemotherapy helps prolong life, it cannot stop recurrence [3].

Conclusion

The choroid plexus tumors in third ventricle are rare and must be differentiated from another tumor from the surrounding area like the pineal gland region. However, awareness of the imaging features of different neoplasms on CT and MRI can help narrow the differential diagnosis.

Patient consent

I confirm that written informed consent for the publication of this case report has been obtained from the patient's legal guardian.

REFERENCES

[1] Osborn AG, Hedlund GL, Salzman KL. Osborn's Brain Imaging Pathology and Anatomy. Second Edition. Salt Lake City, UT: Elsevier, Inc; 2017.

[2] Coates TL, Hinshaw DB, Peckman N, Thompson JR, Hasso AN, Holshouser BA, et al. Pediatric choroid plexus neoplasms: MR, CT, and pathologic correlation. Radiology 1989;173(1):81–8. doi:10.1148/radiology.173.1.2781035.

[3] Mishra A, Srivastava C, Singh SK, Chandra A. Ojha BK Choroid plexus carcinoma: Case report and review of literature. J Pediatr Neurosci 2012;7(1):71–3. doi:10.4103/1817-1745.97633.

[4] Ogiwara H, Dipatri AJ Jr, Alden TD, Bowman RM, Tomita Tadanori. Choroid plexus tumors in pediatric patients. Br J Neurosurg 2012;26(1):32–7.

[5] Lin H, Leng X, Qin CH, Du YX, Wang WS, Qiu SJ. Choroid plexus tumours on MRI: similarities and distinctions in different grades. Cancer Imaging 2019;19:17.

[6] Triulzi FM. Neuroradiology of Brain Tumors. Practical Guide Based on the 5th Edition of WHO Classification. Switzerland AG: Springer Nature; 2023.

[7] Crea A, Bianco A, Cossandi C, Forgnone S, Fornaro R, Crobeddu E, et al. Choroid Plexus Carcinoma in Adults: Literature Review and First Report of a Location into the Third Ventricle. World Neurosurg 2020;133:302–7. doi:10.1016/j.wneu.2019.10.051.

[8] Tavallaii A, Keykhosravi E, Rezaee H, Khamoushi M. An extremely rare case of choroid plexus carcinoma in the third ventricle of an infant: case report and review of the literature. Interdisciplin Neurosurg 2020;22:100873 ISSN 2214-7519. doi:10.1016/j.inat.2020.100873.

[9] Ogiwara H. Choroid plexus tumors in pediatric patients. Br J Neurosurg 2012;26(1):32–7 © 2012 The Neurosurgical Foundation ISSN: 0268-8697 print /ISSN 1360-046X online. doi:10.3109/02688697.2011.601820.

[10] Bohara M, Hirabaru M, Fujio S, Higashi M, Yonezawa H, Karki P, et al. Choroid Plexus Tumors: Experience of 10 Cases with Special References to Adult Cases. Neurol Med Chir (Tokyo) 2015;55(12):891–900. doi:10.2176/nmc.oa.2015-0126.

[11] DM Cannon, P Mohindra, V Gondi, TJ Kruser, KR Kozak Choroid plexus tumor epidemiology and outcomes: implications for surgical and radiotherapeutic management

[12] Horská A, Ulug AM, Melhem ER, Filippi CG, Burger PC, Edgar MA, et al. Proton magnetic resonance spectroscopy of choroid plexus tumors in children. J Magn Reson Imaging 2001;14(1):78–82. doi:10.1002/jmri.1154.

[13] Sasaki T, Kim J, Moritani T, Capizzano AA, Sato SP, Sato Y, et al. Roles of the apparent diffusion coefficient and tumor volume in predicting tumor grade in patients with choroid plexus tumors. Neuroradiology 2018;60(5):479–86. doi:10.1007/s00234-018-2008-2.

[14] Berger C, Thiesse P, Lellouch-Tubiana A, Kalifa C, Pierre-Kahn A, Kim HY, et al. Bouffet E: choroid plexus carcinomas in childhood: clinical features and prognostic factors. Neurosurgery 1998;42:470–5.