

Radiological imaging of choroid plexus tumors: Systematic approach toward diagnosing a choroid plexus tumor

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ABSTRACT

Choroid plexus tumors (CPTs) are rare intraventricular tumors comprising approximately 1% of all brain tumors. The common locations are the lateral ventricle (most common location in children), the third and fourth ventricles (most common location in adults), and cerebellopontine angle. Here, we present the case of a 10-month-old child with complaints of fever (99.6F), abnormal eye movements, and bilateral papilledema. Ultrasound cranium of the child revealed a well-defined hyperechoic lesion adjacent to the trigone and occipital horn of the right lateral ventricle. No vascularity could be appreciated on color Doppler. Non-contrast computed tomography of the brain showed a well-defined lobulated mass lesion epicentered at the choroid plexus of the right lateral ventricle which was hyperdense relative to the brain parenchyma with specks of calcification. Contrast-enhanced magnetic resonance imaging showed a solid intensely enhancing lobulated mass lesion with frond-like morphology originating from the choroid plexus of the occipital horn of the right lateral ventricle. Surgical excision of the CPT was done under aseptic conditions. We try to reinforce the ultrasound (USG), CT, and MRI findings of a CPT which ultimately came out to be a choroid plexus papilloma (CPP) on histopathological examination.

Key words: Choroid plexus, Papilloma, Tumors

Choroid plexus tumors (CPTs) are rare intraventricular tumors. They account for approximately 1% of all brain tumors [1]. These are often misdiagnosed by the reporting personnel and the diagnosis is usually confirmed on histopathological examination. CPTs originate from the neuroectoderm of the choroid plexus cells that secrete the cerebrospinal fluid (CSF) [2]. The common locations are the lateral ventricle, the third and fourth ventricles, and cerebellopontine angle. Other described rare locations are the posterior fossa and suprasellar region. In adults, they are commonly located in the infratentorial compartment while in children, they are supratentorial in location. The most common location in children is the lateral ventricles. On the other hand, they are most commonly located in the fourth ventricle in adults. Incidence is more common in children below the age of 2 years [2].

Here, we present the case of a 10-month-old child with complaints of fever, abnormal eye movements, and bilateral papilledema. The main aim of this case report is to give a step-by-step approach to rule out the differential diagnoses and reach a final diagnosis of a CPT.

CASE REPORT


A 10-month-old child presented to the pediatrics department with complaints of headache, fever, and abnormal eye movements for the past 2 months. Parents of the child gave a history of head trauma 17 days back. There was no history of loss of consciousness.

On examination, the temperature of the child was 99.6F. He was conscious but irritable. No abnormal body movements were noted. Fundus examination revealed bilateral papilledema.

Hematological and urine examinations of the child were unremarkable. The child was advised USG of the cranium to rule out any intraventricular bleed. Proper informed consent was taken from the child's father before every investigation. USG of the cranium (Fig. 1) revealed a well-defined hyperechoic lesion adjacent to the trigone and occipital horn of the right lateral ventricle. There was no mass effect. No vascularity could be appreciated on color Doppler evaluation. Differential diagnoses of periventricular hyperechoic lesion on USG cranium are hematoma in the germinal matrix (perinatal period only), mass lesion, and periventricular leukomalacia. Since the patient gave a history of fall, the differential diagnoses given were intraventricular hematoma and an intraventricular mass lesion. The child was advised of further investigations.

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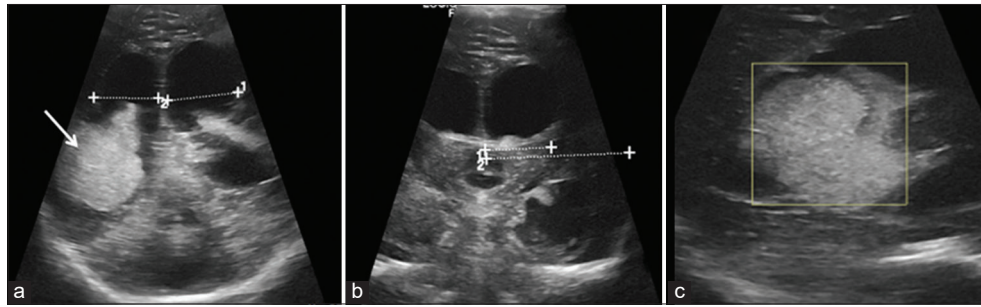


Figure 1: Ultrasound images showing (a) hyperechoic lesion (arrow) noted adjacent the occipital horn of the right lateral ventricle. Grossly dilated lateral ventricles. (b) Ratio of the diameter of the lateral ventricle with the diameter of the brain was $>60\%$ suggestive of hydrocephalus. (c) No vascularity noted on color Doppler. Few echogenic foci noted in the lesion

Non-contrast computed tomography of the brain was done (Fig. 2). CT scan of the brain revealed a well-defined, lobulated mass lesion epicentered at the choroid plexus of the right lateral ventricle which was hyperdense relative to the brain parenchyma with specks of calcification. The diagnosis of an intraventricular mass was confirmed. In view of the specific location and hyperdense mass, a differential diagnosis of intraventricular meningioma and a CPT was given. Contrast-enhanced magnetic resonance imaging of the brain was done the next day (Fig. 3). MRI showed a lobulated, solid mass lesion originating from the choroid plexus of the occipital horn of the right lateral ventricle which was isointense to gray matter on T1WI and hyperintense on T2/FLAIR images. It showed specks of blooming on gradient recalled echo sequences and intense post-contrast enhancement. The margins of the mass were well defined and showed a frond-like appearance. Marked hydrocephalus (out of proportion to the mass) was seen in the bilateral lateral ventricle, third ventricle, and fourth ventricle with adjacent periventricular ooze which was more around the trigone of the right lateral ventricle. The diagnosis given was a CPP with a choroid plexus carcinoma (CPC) as a differential. The MRI features suggesting CPP are a solid tumor with no necrotic areas, lobulated appearance, well-defined margins, no significant invasion into the adjacent brain parenchyma, and a frond-like morphology with intense homogenous enhancement.

Surgical excision of the CPT was done under aseptic conditions and a part of the tumor was sent for histopathological examination. To prevent bleeding, gentle coagulation of the tumor under constant irrigation was done. Total excision of the tumor was achieved. In our case, the cortical incision was made posterior to the vein of Labbe. This provided the shortest access to the trigone. Adjuvant chemotherapy and radiotherapy were not given as it is not indicated for uncomplicated CPP.

Post-operative histopathological examination revealed a typical papillary architecture on hematoxylin and eosin stain suggestive of CPP (Fig. 4). The child had an uneventful recovery without any neurological deficits. Papilledema also subsided a few days after surgery.

DISCUSSION

According to the latest World Health Organization (WHO) classification system [3], CPTs can be classified into three subtypes:

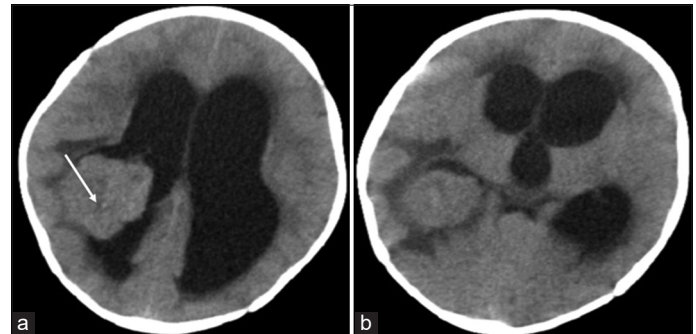


Figure 2: Non-contrast computed tomography of the brain revealed a well-defined, lobulated mass lesion epicentered at the choroid plexus of the right lateral ventricle which was hyperdense relative to the brain parenchyma with specks of calcification. The mass lesion showed calcific foci (arrow) within it. Note the hydrocephalus which is out of proportion to the mass

CPP (CPP Grade I), atypical CPP (aCPP Grade II), and CPC. CPTs show calcification in about 30% of cases and show intense post-contrast enhancement. Most of the CPT are benign (CPP) and have an indolent progression. Shi *et al.* described atypical CPPs as those showing a few histological features of malignancy such as increased mitotic activity or an increase in the layers of epithelial cells. The clear diagnostic criteria of these tumors have not been established. In contrast to benign CPP, they tend to recur and cause meningeal or intracerebral metastasis [4]. In children less than 2 years of age, CPC is common [5]. Larger tumor volume, irregular internal morphology, presence of necrosis, marked peritumoral edema, heterogeneous contrast enhancement, and invasion into surrounding brain parenchyma could be signs of a CPC. However, these cannot be differentiated on imaging alone [6].

According to the previous literature, rare locations of CPPs are in the cerebellar hemispheres [7] and suprasellar region [8]. Kimura *et al.* described a tumor in the suprasellar region as a meningioma which later came out to be a CPP on histopathological examination. Papillomas can have smooth, lobulated, or irregular margins with calcification in 24% of patients. All CPPs enhance with intravenous contrast and the enhancement is typically intense. On the other hand, meningiomas tend to have smooth margins (69%) and a higher calcification rate (46%). Variable sized areas of internal signal void interpreted as regional blood flow, calcification, or old hemorrhage were seen in all cases of CPP [8].

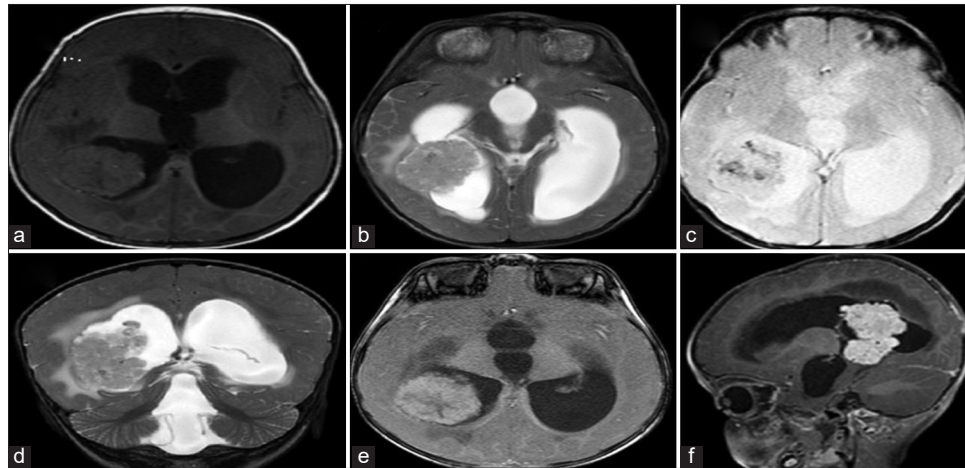


Figure 3: MRI images. (a) On axial T1W1, the mass is isointense to gray matter; (b) on axial T2W1, the mass is hyperintense relative to gray matter; (c) on axial GRE images, it shows specks of blooming suggestive of calcification; (d) on coronal T2W1, note the CSF ooze adjacent the ventricle; (e) on axial post-contrast T1W1, the mass shows intense homogenous enhancement; (f) 3D reconstruction shows the lesion to originate from the ventricles

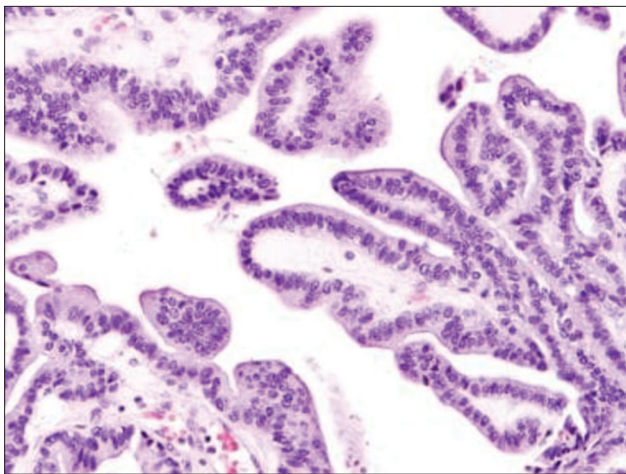


Figure 4: Histopathology showed typical papillary architecture of a choroid plexus papilloma

Wagle *et al.* described the CT and MRI appearance of CPP as a lobulated mass with an intense homogenous enhancement with signal void areas. They postulated that CPPs cause hydrocephalus by affecting CSF dynamics, both by obstruction and by excessive production of CSF [9]. Guermazi *et al.* reviewed the CT and MRI findings of a wide spectrum of lesions that affect the choroid plexus such as neoplasms (papilloma, leukemia, meningioma, lymphoma, and metastases), infections (bacterial, fungal, and viral), cysts, hemorrhage, congenital abnormalities (Sturge-Weber syndrome, Klippel-Trenaunay-Weber syndrome, and vascular malformations), and non-infectious inflammatory disorders (xanthogranulomas, inflammatory pseudotumor, neurosarcoidosis, rheumatoid nodule, and villous hypertrophy). Few of the findings of choroid plexus involvement are specific for a particular pathological process. The absence of any blood-brain barrier and the differential permeability of the choroidal epithelium explain the diffusion of contrast agents into the extracellular space of the choroid plexus which results in enhancement of the choroid on both CT and MR images. On the other hand, no enhancement is observed in the CSF after

contrast medium administration. Patients with CPCs frequently have focal neurological deficits, extensive parenchymal invasion, and peritumoral vasogenic edema which are suggestive of a malignant lesion [10]. Dhillon *et al.* demonstrated that CPTs show frequent mitotic figures (>5/10 HPF), increased cellularity, nuclear pleomorphism, blurring of papillary pattern, and necrosis on histopathological examination [11]. Molecular genetic changes and several genetic loci in CPTs were also studied [12].

Pencalet *et al.* proved that total surgical excision is curative in cases of papillomas. For carcinomas, the most effective treatment remains total surgical excision. However, adjuvant treatment in the form of chemotherapy in younger patients (<3 years of age) and chemoradiotherapy in older children reduces the risk of recurrence [13].

CONCLUSION

Since CPTs are rare tumors, a systematic approach is needed to reach a diagnosis of a CPT. Clinically correlating the history with characteristic findings of hydrocephalus out of proportion to the size of lesion, calcifications and intense post-contrast enhancement in a mass lesion epicentered in the ventricles in a child can lead us to the diagnosis of a choroid plexus neoplasm. Differentiation between CPP and CPC can be made reliably in many cases based on imaging alone with features such as enhancement pattern, volume of tumor, involvement of brain parenchyma, and presence of necrosis.

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