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Clinicoradiological Diagnosis of Silent Choroid Plexus Papilloma in a Child

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Abstract

Background: Choroid plexus papillomas (CPPs) are benign slow-growing tumors with the most common site as the choroid plexus of the lateral ventricle in children. These are highly vascular solid tumors which have classical diagnosis in contrast-enhanced computerized tomography (CECT) and contrast-enhanced magnetic resonance imaging (CEMRI). These require early management because of their secondary symptoms related to hydrocephalus and others due to tumoral compression. **Case Report:** We present a 10-month-old male child with abnormal eye movements and fever following head trauma seventeen days before. Ultrasound of the brain revealed the hyperechoic mass in the right lateral ventricle. CECT and CEMRI diagnosed as a case of choroid plexus papilloma with gross hydrocephalus. This was a silent CPP which came to limelight following head trauma. Ventriculoperitoneal shunt was placed for the decompression of the ventricular system and was advised for subsequent surgical management. **Conclusion:** Atypical presentations of CPP can confuse in making the diagnosis and this delay can be harmful to the patient. Early diagnosis of CPP is very important as these are slow-growing tumors which can lead to various complications if late diagnosis is made. Cross-sectional modalities such as CECT and CEMRI are the most valuable diagnostic modality tools for the early diagnosis.

Keywords: Contrast-Enhanced Computerized Tomography, Contrast-Enhanced Magnetic Resonance Imaging, Choroid Plexus, Choroid Plexus Papilloma, Hydrocephalus

1. Introduction

Choroid plexus papillomas (CPPs) are of the neuroepithelial origin and arise from choroid plexus. These constitute 3% of all intracranial tumors of the children[1]. The most common site is ventricular system. The site of lateral ventricle is more common in children as compared to fourth ventricle in adults. Other rare sites are from third ventricle and cerebellopontine angle[2]. There are cases where the intraparenchymal location of CPP has also been described.

2. Case Report

A 10-month-old male child reported with the complaints of fever and abnormal eye movements of 2 days duration followed head trauma 17 days back. The child was perfectly alright previous to the head trauma. On examination, the child was conscious without any neurological deficit. Fundus examination had shown papilledema. Cranial ultrasound revealed a hyperechoic mass measuring 4.8 cm × 3.6 cm in the right ventricle just abutting the interventricular choroid plexus. There was no vascularity in color flow imaging. Both the lateral ventricles show moderate dilatation. The 3rd and 4th ventricles were also dilated. NCCT of head was done which revealed lobulated slightly hyperdense mass in the right lateral ventricle with adjacent compression of the brain parenchyma (Figure 1 a and b). Magnetic resonance imaging (MRI) had shown an isointense lobulated mass on TIWI and slightly hyperintense on T2WI sequences. FLAIR sequence had revealed surrounding edema.

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Post-contrast TIWI with fat suppression had shown avidly enhancing mass in the right lateral ventricle with encroaching on surrounding regions (Figure 2a-c, and Figure 3 a-c).

3. Discussion

CPP arises from choroid epithelium and these are very slow-growing tumors and may be diagnosed quite late as happened in our present case.

The choroid plexus first appears at the roof of the fourth ventricle after closure of neural tube at six weeks of gestation. At 20 weeks of gestation, this gets appearance of adult choroid plexus. CP is covered with ependymal lining and this forms the cerebrospinal fluid (CSF). Hence, this becomes the pathway for many diseases to cross through on either side[3]. Sometimes, choroid plexus cysts cause confusion in the diagnosis during pregnancy but majority resolve in the later stage. If these persist, then the follow-up is required to differentiate from other cystic type of pathologies[4]. CPP may not cause symptoms for a long time as there is potential space of expansion of the tumor in the ventricle. The obstruction can be caused after it blocks the pathway of CSF. The tumor compresses on the ventricular wall and eventually the brain parenchyma. CPP can show excessive calcification and even ossification which can mask the enhancement pattern. The common symptomatology includes signs of raised intracranial tension which includes headache, nausea and vomiting, drowsiness, papilledema, visual disturbances, and involvement of cranial III and VI nerves causing ocular palsies. Hydrocephalus (tension-obstructive type) and macrocephalus are also the common features. Psychosis and seizure presentation may also be there. These are most commonly found in the lateral ventricle (supratentorial) and the age of occurrence in the first and second decade. The trigone area is the most common site of involvement. About 85% of CPP occur below 5 years of age in adults fourth ventricle (infratentorial) is preferred site in addition to the rare locations such as third ventricle, CP angle, suprasellar region, pineal region, frontal lobe, and cerebellum[5,6]. These are solid vascular tumors and show frond-like vivid enhancement pattern. Macroscopically CPP looks like cauliflower type of mass. CPPs are papillary structures with fibrovascular core lined by columnar or cuboidal epithelium with vesicular nuclei

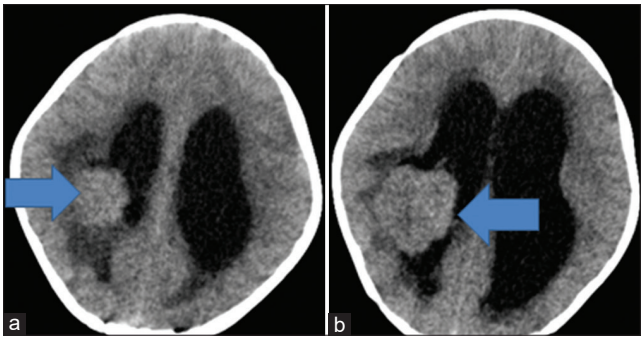

Figure 1: NCCT head. (a) Axial section at higher level shows a hyperdense lobulated mass projecting in in the right lateral ventricle (blue arrow). (b) Axial section at trigone level depicting the mass causing compression of the adjacent brain parenchyma (blue arrow).

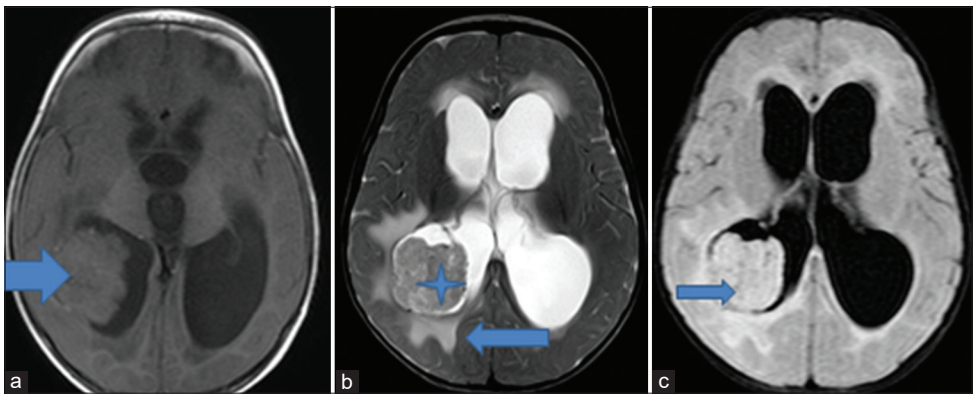

Figure 2: Axial section of MR images. (a) TIWI shows an isointense lobulated mass in the posterior horn of the right lateral ventricle extending to the trigone (blue arrow). (b) T2WI shows the same mass with the surrounding edema. (c) Fluid-attenuated inversion recovery (FLAIR) sequence shows the mass (blue star) compressing the brain parenchyma with periventricular ooze (blue arrow).

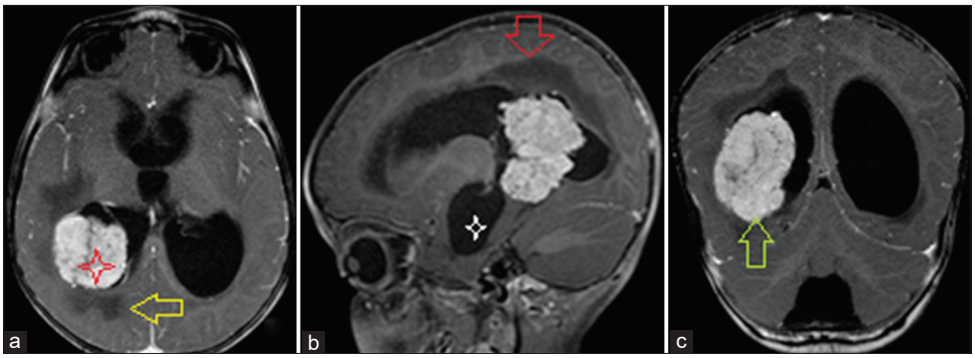

Figure 3: Contrast-enhanced TIWI with fat suppression. (a) Axial section shows avid enhancing mass (red star) with surrounding edema (yellow arrow). (b) Sagittal section of the same mass lesion shows dilated ventricular system (white star) with compression of brain parenchyma (red arrow). (c) Coronal section avid enhancing choroid plexus papillomas almost occupying the potential space of the right lateral ventricle (green arrow).

and look like choroid plexus[7,8]. There are three grades of these type of tumors as per the WHO grading (Table 1). These are benign tumors falling in the category of the WHO grade I[9].

There is known association with von Hippel-Lindau disease, Aicardi syndrome, and Li-Fraumeni syndrome[10]. There have been cases of infantile myofibromatosis which manifest as choroid plexus mass and the spontaneous regression takes place[11]. Hydrocephalus is the most common feature in these cases and this is because of overproduction and decreased absorption of CSF. About 25% masses show speckled calcification. Contrast-enhanced computerized tomography shows lobulated frond-like mass with vivid enhancement. This gives cauliflower appearance. These are isodense masses but heterogenousity in character and enhancement pattern points toward the carcinoma. MRI also shows avid enhancement pattern. TIWI shows isointense and T2WI shows hyperintense in nature. Flow voids can also be seen in the lesions. Spectroscopy shows increased choline and decreased NAA (N-acetylaspartate) peaks. Angiography can further demonstrate the blush and the enlarged feeding choroidal arteries with shunting. Angioinfarction is always helpful before surgical maneuver to avoid blood loss. Surgical total excision is the aim of management and is successful in most of the cases. CSF seeding is not the feature in these low Grade I tumors as compared to the higher grade. Coagulation of the CPP always helps in regression of the tumor and the pedicle should be dealt carefully to avoid post-operative ventricular hemorrhage. The intracranial drain is placed to monitor the bleeding if there[12].

Table 1: The WHO grading of CPP

Grading	Diagnostic criteria
Grade I	< Two mitoses are present per 10 HPF
Grade II	Atypical CPP
Grade III	Choroid plexus carcinoma

CPP: Choroid plexus papillomas

4. Conclusion

CPPs in lateral ventricles are though common in early childhood but the early diagnosis is very important. These tumors are silent and slow growing and lead to hydrocephalus and compression of the surrounding regions leading to complications. Clinical symptomatology with cross-sectional imaging can overcome all the complications in early management.

5. Acknowledgment

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6. Conflicts of Interest

None declared.

7. Source of Support

Nil.

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