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An unusual case of choroid plexus papilloma in infancy. Diagnostic and management challenges

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ABSTRACT

Choroid plexus papillomas (CPPs) are rare tumours having bimodal distribution. Pediatric CPPs are commonly present in the supratentorial compartment and most commonly located in the lateral ventricle and usually present at 16-18 months. In the present case authors illustrate an atypical case of CPP in a 6-month-old infant who was admitted to our hospital after a trivial fall with no features of hydrocephalus or raised intracranial pressure. Quick surgery keeping in mind the Maximal allowable blood loss (MABL) and low physiological reserve of infants with quick decompression of cysts and devascularizing the tumor from all around and removing the tumor in toto plays a key role in operating the pediatric tumor. MRI features play a key role in preoperative diagnosis; nevertheless, it is difficult to distinguish atypical CPT from other lesions. Surgical resection is the cornerstone of this treatment.

Keywords
choroid plexus papilloma,
lateral ventricle,
maximal allowable blood loss

INTRODUCTION

Choroid plexus tumors (CPTs) infrequently occur with an intracranial derivation from the epithelium of the ventricular system's choroid plexus and are of neuroectodermal origin[1]. In adults, CPTs represent 0.3%–0.6% of all brain tumors, whereas in children, they comprise between 2% and 5%[3]. The World Health Organization (WHO) categorizes CPTs as choroid plexus papillomas (CPPs, WHO I), atypical choroid plexus papillomas (ACPPs, WHO II), and choroid plexus carcinomas (CPC WHO III)[6].

CPTs normally occur in areas with a choroid plexus, usually in the ventricular system [2,3]. These tumors are more common in supratentorial locations, including the lateral ventricles among children and the fourth ventricle in the elderly [7,8]. The usual presentation is associated with increased intracranial pressure due to hydrocephalus secondary to CSF overproduction and/or obstruction of CSF flow. There are several causes of hydrocephalus, including direct mechanical obstruction of the flow of cerebrospinal fluid (CSF), hemorrhagic blockage of arachnoid granulations, and excessive production of CSF [4,5].

The median age of diagnosis is 18 months in the pediatric population [14]. In 1927, Walter Dandy managed to perform the initial successful removal of pediatric CPT from the left lateral ventricle of a

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14-year-old girl, whereas Van Wagenen did the same in an infant aged 3 months[15]. Surgical extirpation is the best therapeutic option and significantly curtails the possibility of regrowth if gross total resection (GTR) is performed [16]. A majority of children who undergo treatment for these tumors are often very young; hence, their physiological reserve and maximal allowable blood loss (MABL) are low; hence, the need for meticulous surgery, especially from experienced neurosurgeons, becomes paramount. Profuse bleeding during surgery may be life-threatening among children with plexus tumors because CPTs are highly vascular and most children treated for plexus tumors are very young with low minimal allowable blood loss (MABL). Hence, it is crucial that feeding arteries are identified soon enough to reduce blood loss [17].

CASE REPORT

History and physical examination

This 6-month-old male infant, born out of a non-consanguineous marriage, presented to our institute with a minor head injury after falling off the bed and accidently hitting his head. He was taken to a nearby hospital for evaluation. The prenatal and antenatal periods were uneventful, and the child had a normal vaginal delivery with a normal cry immediately after birth. Before admission, the child had a history of episodes of excessive inconsolable crying for the past 2 months.The child was taking normal feeds and had attained normal developmental milestones as per age. At the time of examination, the child was active and playful. The anterior fontanelle was open and lax, and the head circumference (HC) was 44 cm (50th centile). There was scalp swelling in the left parietal region with no skin discoloration or underlying fracture. He underwent computerized tomography (CT) scan of the brain (Fig 1) and was referred to our center because of a brain tumor. There were no features suggestive of increased intracranial pressure (ICP) such as papilledema.

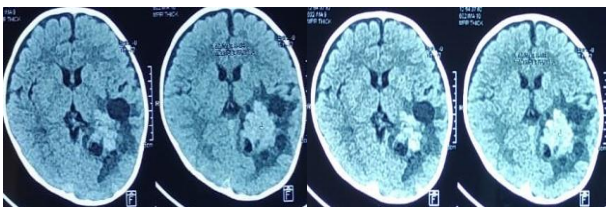

Fig 1: Pre-operative CT brain of the infant showing a lobulated hyperdense lesion in left parietal lobe, solid cystic in consiste and solid part enhancing on contrast with no evidence of hydrocephalus or mass effect .

Imaging findings

CT scan showed a large lobulated, hyperdense solid cystic lesion located in the left parieto-occipital region, with the solid part of the tumor enhancing on contrast (Fig. 1). On MRI, the lobulated solid cystic lesion of size approximately 55mm x 45 mm x 50 mm, located in the left parieto-occipital region. The lesion solid part was isointense on both T1 and T2. The cystic part of the tumor was hypointense on T1 and hyperintense on T2 and bright contrast enhancement of the solid part was observed. There was no evidence of hydrocephalus, mass effect, or peri-lesional edema.

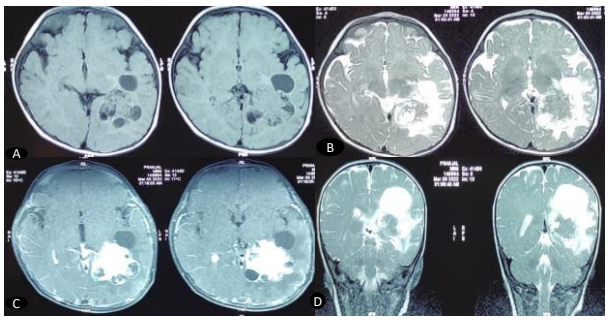

Fig. 2. Pre-operative T1 axial (A), T2 axial (B), T1 axial contrast (C) T2 Coronal brain MR imaging of infant showing lobulated, solid cystic lesion in left parietal lobe with solid part brilliant contrast enhancement. No evidence of hydrocephalus, mass effect, perilesional edema, mass effect

Management and surgical technique

Our working diagnosis was ependymoma, and other differential diagnosis was high grade glioma and choroid plexus papilloma (CPP). The absence of hydrocephalus blinded us to CPP diagnosis. We planned for surgical excision. The patient was given general anesthesia and positioned prone, and left-sided parieto-occipital osteoplastic craniotomy was performed. The brain was lax on removing the bone flap. Subsequently, the duramater was opened and transcortical-transventricular approach was taken to the left occipital horn. The tumor was soft to firm in consistency, yellowish-white in color, and appeared cauliflower-like. Considering that pediatric patients have very low MABL and these tumors are pose significant bleeding risks, microscopic circumferential dissection, coagulation, and disconnection of tumor vascular supply from the lateral ventricle was performed.

The feeders from the choroidal arteries were identified under a high-magnification microscope and coagulated. The stalks of the attachments were coagulated and cut using bipolar electrocautery. Gross total excision of the tumor was achieved, and

the lesion was excised in-toto (Fig 3). Intraoperative blood loss was 200 cc, which did not exceed the calculated MABL of 250 cc, and no blood transfusion was required. The choroid plexus of the left lateral ventricle was clearly visible and coagulated. Ventricular drain placed and maintained for 3 days in the left lateral ventricle.

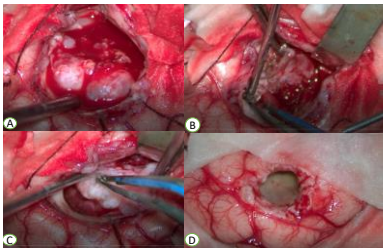

Fig. 3. Intra-operative images. The patient was positioned prone

(A) tumour was identified at a depth of 3.5 cm posterior to trigone of left lateral ventricle

(B) tumour was firm, moderately vascular, yellowish white in colour.

(C) Excised tumor segments had cauliflower appearance indicating choroid plexus origin of the tumor

(D)). Hemostasis achieved after gross total excision of the tumour.

Post operative course

Post-operative period was uneventful. Post-operative CT scan of the brain showed complete excision of the lesion (Fig 4). Patient was extubated next day. The baby was discharged on post-operative day 7 and he was taking oral feeds. The infant was doing well at the last follow-up at 6 months post-operation. A post-operative Magnetic Resonance Imaging (MRI) brain revealed no evidence of residual lesion.

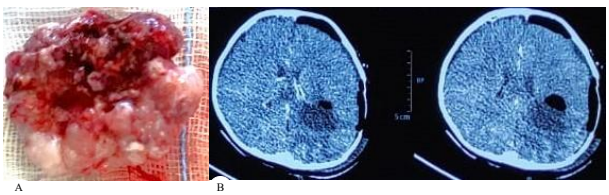

Fig 4 Tumor segments had cauliflower appearance indicating choroid plexus origin of the tumor(A) Post operative CT scan showing gross total excision of the tumor

Discussion

CPP accounts for 2-4% of all pediatric brain tumors and is of neuroectodermal origin [14,16]. In the pediatric population, CPTs usually occur in the lateral ventricles. They commonly appear as papillary or lobulated intraventricular lesions and are usually associated with hydrocephalus[8,9]. The usual presentation of CPTs in the pediatric population is an enlarged head size associated with clinical presentation suggestive of increased intracranial pressure. In our case, the child had no clinical features suggestive of raised ICP (anterior frontanalle was normal and absent papilledema) and presented with mild head injury after falling from bed. After imaging evaluation, there was a diagnostic dilemma between ependymoma and choroid plexus papilloma. CPT is usually associated with hydrocephalus in 40% of patients [12,14], and some articles reported that moderate or severe hydrocephalus was seen in almost all cases[2,6,8,14,17] which was absent in this case. Note that hydrocephalus was not associated in this study, which is one of the most significant differences between it and typical CPT. The absence of hydrocephalus as a marker poses a diagnostic dilemma for radiologists and neurosurgeons.

These tumors can be challenging to treat because they most often present in childhood and are highly vascular. During surgery due to low MABL, excessive blood loss can become life threatening. Physiological reserve in infants is very low; hence, attention should be paid to blood loss right from the skin incision and subcutaneous tissue dissection because by the time we reach the tumor, blood loss should not limit us in excision of the tumor.

Experienced neurosurgeons who have a lot of experience in operating pediatric cases are required along with an expert team of neuroanaesthetists who are experts in managing pediatric neurosurgery cases.In the pediatric age group and especially infants, one major challenge is the risk of hypothermia, which has to be tackled from the beginning by the neuroanaesthesia team . in planning surgery, the goal should be quick timed decompression of the cyst. the approach is to remove the tumor in toto by devascularizing the tumor from all around. The cyst proves to be beneficial and helps in the surgery as quick decompression of the tumor helps in reaching and excision of the tumor. Early identification of feeding arteries is therefore important to reduce blood loss. Gross total resection (GTR) has become the standard of care for avoiding tumor recurrence.

Conclusion

In contrast to typical lesions, hydrocephalus was not found in our case of CPP, which posed a major diagnostic challenge. A team of experienced pediatric neurosurgeons and a neuroanaesthesia team experienced in managing pediatric neurosurgery cases is required to operate a pediatric choroid plexus tumor. Quick surgery keeping in mind the MABL and low physiological reserve of infants with quick decompression of cyst and devascularizing the tumor from all around and removing the tumor in toto plays a key role in

operating the pediatric tumor . MRI features play a key role in preoperative diagnosis; nevertheless, it is difficult to distinguish atypical CPT from other lesions. Our study showed that atypical CPT cases were treatable by radical excision.Surgical resection is the cornerstone of this treatment.

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