Atypical Choroid Plexus Papilloma of the Third Ventricle in a 3-Month-Old Infant

Ebtesam Abdulla, BSc, MD* Nabeel Hameed, MD, AFSA** Asma AlQaseer, MB BCh BAO*** Nisha Chandran, MD ****

Choroid plexus papilloma (CPP) is a rare benign tumor arising from the lining of the choroid plexus. Pediatric patients usually present with signs of hydrocephalus that necessitate immediate intervention.

A three-month-old female infant presented with increasing head size. Initial imaging showed a third ventricular mass lesion with non-communicating hydrocephalus. The patient underwent surgery and the lesion was completely excised with no complications. Histopathology examination confirmed the diagnosis of the WHO grade II atypical choroid plexus papilloma. This is a rare case of a child with an atypical CPP of the third ventricle highlighting the investigations, diagnosis, and surgical challenges.

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Choroid plexus tumors are rare neuroepithelial intraventricular tumors, representing less than 1% of all intracranial tumors and less than 4% of brain tumors in children[1-3]. Choroid plexus papillomas (CPPs) are benign tumors originating from the choroid plexus. They are usually found in the fourth ventricle in adults and lateral ventricle in infants and children[3-6]. The third ventricle is a very rare location, only a few cases reported in the literature. The challenge with these lesions is related to the propensity of the cerebrospinal fluid and the extreme vascularity which can lead to significant blood loss during surgery, necessitating an incomplete resection of these tumors[3,7,8].

The aim of this report is to present a rare case of an atypical CPP located in the third ventricle which was completely excised.

THE CASE

A three-month-old female infant presented with one-month history of progressively increasing head size noticed by her parents. The patient's parents have also noticed an upward gaze and muscle rigidity one week prior to the presentation. The patient was born via normal spontaneous-vaginal delivery. The head circumference was normal at birth. On examination, the patient was alert and conscious, with no other neurological deficits. The patient's head size measured 48 cm, which is larger than normal for her age. The examination also revealed lax fontanelles, positive sunset sign bilaterally, and engorged scalp veins. All laboratory investigations were within normal limits.

CT of the brain showed frond-like soft tissue within the third ventricle with an extension into the left lateral ventricle with maximum dimensions of 4.5×2.7×3.0 cm. Splaying of the fornix was also seen with the tumor extending into the interpeduncular cistern. MRI of the lesion showed hypo-intense signals in the T1-weighted sequence and iso-intense signal in T2- weighted sequence with vivid, homogenous enhancement in post-contrast sequence. Additionally, marked obstructive hydrocephalus and dilation of the lateral ventricle were seen. There was no evidence of structural or neuronal migration abnormalities, see figure 1. Magnetic resonance angiography and magnetic resonance venography were normal without abnormal mass vascularity or collateral vessels. MRI of the spinal cord showed no abnormal cord lesions or leptomeningeal enhancement. Based on the radiographic appearance of this lesion, a diagnosis of choroid plexus papilloma was highly suspected.

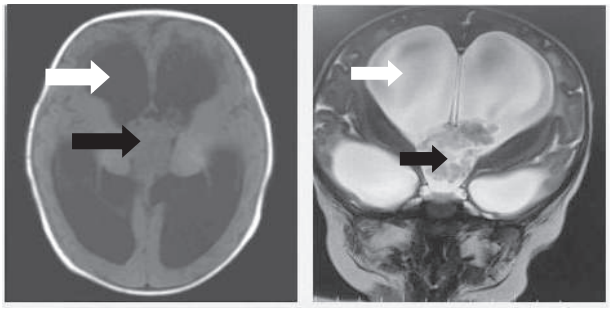

Figure 1: Preoperative T1 and T2 Weighted MRI of the Brain Showing Third Ventricle Mass (Black Arrow) with Marked Hydrocephalus (White Arrow)

* Neurosurgery Resident
Department of Neurosurgery
** Consultant Neurosurgeon
Department of Neurosurgery
*** Surgical Resident
Department of Surgery
**** Specialist Histopathologist
Department of Pathology
Salmaniya Medical Complex
Kingdom of Bahrain
E-mail: Dr.Ebtesam@hotmail.com, axoncell@gmail.com, asma.alqaseer@gmail.com, nishanijesh@gmail.com

The patient underwent trans-callosal excision of the tumor with the placement of an external ventricular drain, see figure 2. A mass resection was achieved, with no intraoperative or postoperative complications. A postoperative CT scan of the brain showed no evidence of residual tumor within the third ventricle with regression of the hydrocephalus, see figure 3. The patient had an uneventful recovery and was discharged. The histopathology specimen was consistent with the diagnosis of an atypical choroid plexus papilloma (WHO grade II), see figure 4.

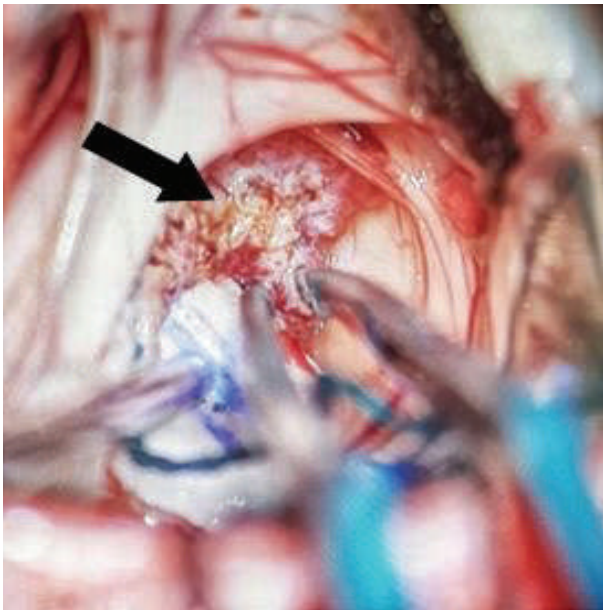

Figure 2: **Intraoperative Image of the Lesion Showing a Cauliflower-like Vascular Mass in the Third Ventricle with an Attachment to the Choroid Plexus (Black Arrow)**

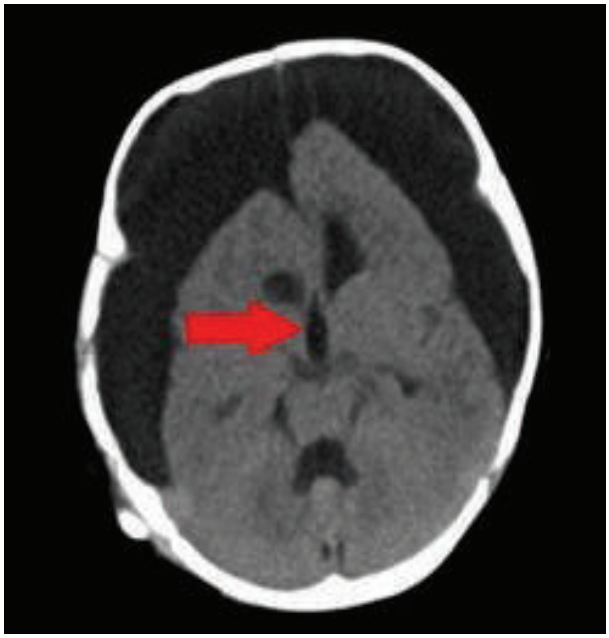

Figure 3: Postoperative Image of CT Brain Showing Total Excision of the Third Ventricular Mass and Regression of the Hydrocephalus (Red Arrow)

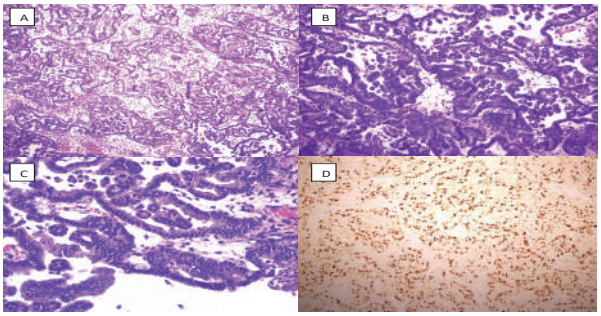

Figure 4: Sections Showing Atypical Choroid Plexus Papilloma with Prominent Papillary Architecture and Lack of Sheeting 5x and 10x (A and B). Active Mitoses are Noted in High Power 40x (C). The Proliferation Index is 40% Assessed by Ki-67 Immunohistochemical Stain (D)

DISCUSSION

CPP is a rare benign neuroepithelial tumor, accounting for less than 1% of all intracranial neoplasms[1]. CPP is seen primarily within the ventricular system; it rarely extends to the extra-ventricular region. In the pediatric population, 80% of CPPs arise in the lateral ventricle and 16% in the fourth ventricle[7]. The third ventricle is a rare location of CPP and accounts for less than 4%[7]. It is more frequent during the first year of life, and the mean age at diagnosis is 0.7 years[7,9,10].

The diagnosis of CPPs in children is challenging as they have a slower onset of symptoms. The clinical features and presence of neurological deficits depend on the patient's age and site of the lesion. The most common presentation in children is hydrocephalus and features of raised intracranial pressure[3,8,9,11]. This is due to several mechanisms which include overproduction of cerebrospinal fluid (CSF) by the tumor cells, obstruction of the ventricular system, dysfunction in reabsorption system, arachnoid adhesions or elevated CSF protein concentrations[3,7]. Infants with CPPs may present with an increasing head size, lethargy, decreased activity, and poor psychomotor milestones. As the child grows older, visual impairment and gait disturbances may be noted due to the raised intracranial pressure and/or the direct pressure caused by the tumor[3,4].

MRI of the brain is the best modality to visualize CPPs[3]. On the MRI, the lesion will appear well-defined, iso-intense or hypo-intense in T1 and iso-intense or hyper-intense in T2-weighted sequence. Hydrocephalus is a common radiographic finding, and calcification is seen in up to 25% of cases[7-9]. Due to the vascular nature of the tumor, angiography can be performed pre-operatively to safely plan the surgery[7,8].

The final diagnosis of choroid plexus tumors is made following surgical resection of the lesion and histopathology analysis[5]. Choroid plexus tumors classified according to the WHO into 3 grades. Grade I is choroid plexus papilloma, grade II is atypical choroid plexus papilloma, and grade III is choroid plexus carcinoma[7,8,10,12]. Atypical CPP are intermediate lesions, and were added to the classification in 2007 primarily due to its mitotic activity[8,10]. Recent studies have suggested that increased mitotic activity is associated with a higher recurrence rate in pediatric patients[12,13].

Surgical resection of CPP is the treatment of choice[3,5,6,8]. Due to the rich vascular supply of these tumors, gentle coagulation of the tumor under constant irrigation is preferred to prevent life-threatening intraoperative bleeding[3,11]. In addition, adequate control of the vascular pedicle supplying the tumor is essential to prevent ventricular hemorrhage and brain edema[3,11]. Hydrocephalus is generally relieved following the excision of the tumor; however, in some cases, persisting hydrocephalus can occur due to arachnoidal adhesions at the level of the subarachnoid spaces and arachnoidal granulations leading to communicating hydrocephalus. In such cases, a permanent CSF diversion procedure may be required[7,11]. Complete surgical removal of CPP leads to excellent prognosis with 5-year survival rate of up to 100%[2-4,9].

CONCLUSION

Atypical choroid plexus papilloma located in the third ventricle is a rare diagnosis in the pediatric population. Surgical resection is the mainstay treatment and is associated with excellent outcomes.

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