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Case Report

Extraventricular Intraparenchymal Choroid Plexus Carcinoma in a Pediatric Patient: A Case Report

Mohamed Shamsaldin[1], Adnan Ghanem[2], Noofel Al-Ashhab[1], Sakhr Al-Faqih[1] Abdulghani Al-Aswadi [3], Mojahed Me'asar[1]

1Department of Neurosurgery, 21 September University for Medical and Applied Sciences, Yemen

2Faculty of Medicine and Health Sciences, Thamar University, Yemen

3Faculty of Medicine and Health Sciences, Sana'a University, Yemen

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Abstract

Background: This article reports a case of intraparenchymal, supratentorial, extraventricular choroid plexus carcinoma (CPC) in a pediatric patient.

Case Report: A 10-year-old girl presented with a 4-month history of chronic progressive headache, left hemiparesis, urine incontinence and blurry vision that deteriorated later to a complete blindness. A brain magnetic resonance imaging (MRI) demonstrated an intra-axial combined cystic and solid mass in the right frontal lobe with calcified foci. A gross total resection was performed followed by chemotherapy and Radiation therapy. The histopathological examination revealed the diagnosis of CPC. On serial MRI, a small newly developed focal enhanced recurrent lesion in the right frontal lobe, anteromedially to the previous surgical bed was identified after one year of follow-up.

Conclusion: This case reports the importance of early surgical management and continuous monitoring of extra- ventricular CPC by serial MRI to treat any recurrent lesion very early.

Keywords: choroid plexus carcinoma, pediatric, extraventricular, intraparenchymal.

Mohamed Shamsaldin[1], Adnan Ghanem[2], Noofel Al-Ashhab[1], Sakhr Al-Faqih[1] , Abdulghani Al-Aswadi [3] , Mojahed Me'asar[1]

Introduction

Choroid plexus tumors (CPTs) are rare brain tumors that represents 2–5% of all pediatric brain cancers [1]. The annual incidence of all CPTs is approximately 0.3 per million individuals [2]. CPTs are histologically classified by the World Health Organization (WHO) into choroid plexus papilloma (CPP; WHO grade I), atypical choroid plexus papilloma (WHO grade II), and choroid plexus carcinoma (CPC; WHO grade III) [3]. CPC is an extremely rare malignant intracranial tumors [4]. CPCs mainly occur among children younger than 2 years. The median age is 1 year at diagnosis in comparison with CPP [5], 80% of all CPCs found in children. It represents about (20−40%) of all CPTs in pediatric patients [6]. CPC mainly arises within ventricles [7]. In extremely rare cases, they arise in locations outside of the ventricles, presumably from an ectopic choroid tissue [7]. For example, they may arise in cerebellum[8], cerebellopontine angle [9] or intraparenchymal brain tissue [10] [11]. Reviewing the related literatures about the origin of CPC reveals that only three previous studies reported an extraventricular Intraparenchymal origin for CPC in pediatric. To the best of the researchers' knowledge, this case is the fourth reported study among pediatric worldwide, and the first reported one in the Middle East.

Case Report

Clinical History: A 10-year-old Yemeni girl presented with a 4-month history of chronic progressive headache, which was associated with multiple attacks of vomiting. A 3-month period later to the onset of headaches, she started complaining of left hemiparesis and blurry vision on both eyes that markedly worsened over 1 month to a complete loss of vision. Then, she developed urine incontinence and became less playful. Associated symptoms included general malaise, and fatigue. No history of fever, chills, seizures, memory deficits, or behavioral changes was reported by her relatives. No history of medical illness or malignancies among her family was reported. Her family denied any history of congenital diseases or neoplasms.

Physical Examination: The girl was lethargic and disoriented. Her GCS when first seen was 13/15. She responded slowly when she was asked about her name. Her vital signs were within normal ranges. She was breathing spontaneously without any difficulties (Pso2= 95% on ambient air). Her heart rate was 88 bpm, temperature was 37C°, blood pressure was 90/60. She had a urinary catheter that was inserted in ER. The patient had left-sided body weakness. Her left side motor power was 3/5. She has normal full power in right side of body. She had a clear and fluent speech. Her cranial nerve examination demonstrated a total loss of the visual acuity with no light perception. Her fundoscopic examination revealed bilateral papilledema that was more on her right fundus. Other cranial nerves examination was within normal. Her sensory and cerebellar examinations were within normal. Meningeal irritation signs were normal. Other regional physical examinations were unremarkable.

laboratory: Findings of routine laboratory studies, including a complete blood count with differential and a basic chemistry panel, were within normal limits apart from low serum sodium.

Imaging Studies: Firstly, a contrast enhanced brain CT scan was performed, which demonstrated an intra-axial heterogenous enhanced mass measuring about (5x5x4cm) within the right frontal lobe with solid and cystic components with surrounding extensive hypodense vasogenic edema and midline shifting **(Figure. 1).**

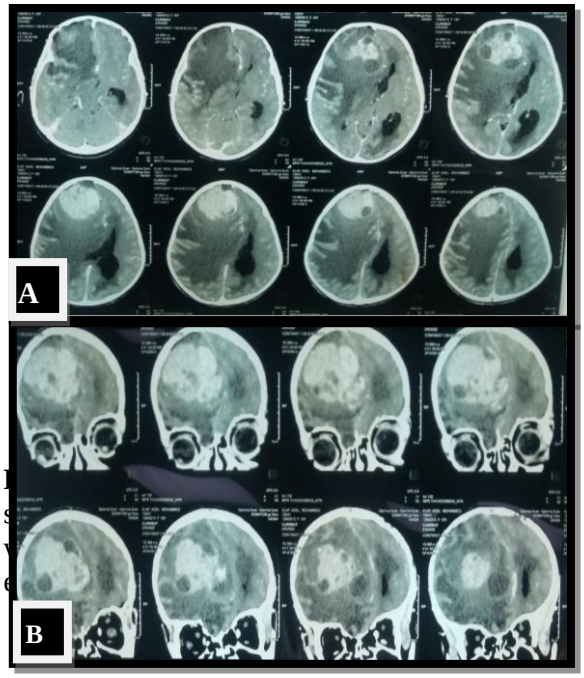

Figure 1: Contrasted Brain CT scan shows an intra-axial heterogenous mass within the right frontal lobe with surrounding extensive vasogenic edema and midline shift. **A:** Axial cuts, **B:** Coronal cuts.

Two days later, a brain magnetic resonance imaging (MRI) was performed. It demonstrated an intra-axial hyperintense mass within the right frontal lobe with a mixture of cystic and solid components and calcified foci, with severe suurounding vasogenic edema and midline shift about 1.6 cm to the left side and secondary non communicating hydrocephalus **(Figure 2)**

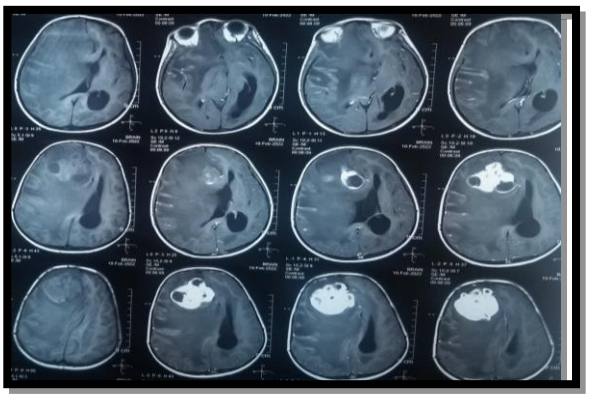

Figure 2: Brain MRI T1 (axial cuts) showing a mass within the right frontal lobe with surrounding extensive vasogenic edema and midline shift

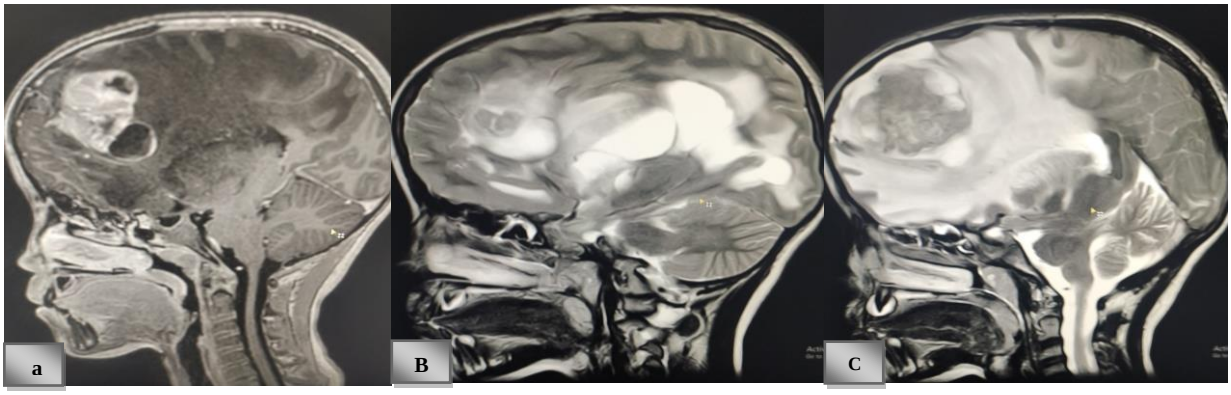

Figure 3: Preoperative contrasted Brain MRI (sagittal cuts) show a large heterogenous mass within the right frontal lobe (**A)** with extensive edema around the mass, (hyperintense in **T2** (**B&C**)

Management Plan

Initially, it was suggested that the patient's brain mass is a high-grade glioma based on neuroimaging studies. As the presentation of the patient was urgent, she was admitted immediately to the neurosurgical department at the 48 Model Hospital to be stabilized and prepared for tumor excision operation. The treatment plan was discussed with the patient's family. Surgery was planned for a craniotomy and excision of the tumor for histopathology, and may be insertion of a ventriculp-perotonial shunt or an External Ventricular Drain (EVD) if needed. Steroids were prescribed before the operation to relieve the vasogenic edema.

Pre-operative work-up

Full routine preoperative investigations were requested. A preoperative medical fitness by a multidisciplinary team formed by an anesthesiologist and pediatrician for pre-anesthesia evaluation of the patient were processed.

They advised for correction of hyponatremia before operation. A written consent was

taken from the family for the craniotomy procedure. Two units of crossed-matched

packed RBCs and two units of fresh frozen plasma were prepared. A V-P shunt and EVD

devices were prepared. All operation instruments were checked in.

Operation

A C-shape skin incision in the right frontal area was extended to cross the midline over superior sagittal sinus (SSS). Craniotomy and elevation of bone flap using an electronic drill, and bone cutter. The dura was seen dusky and adherent to the skull bone. On opening of the dura, the brain was tense; an ill-defined mass with cystic and solid components was encountered immediately. Aspiration of the cystic part along with a near total excision of the mass by dissection of its margins from the brain tissue were made by cutting and coagulating of the marginal attachments while enfolding the tumor into the area and decompression with minimal retraction on adjacent brain, SSS and the falx

cerebri, with taking maximal care to the wall of the frontal horn of the right lateral ventricle, without opening it. Then the duraplasty was done followed by replacement of the bone flap, and scalp closure. The procedure passed very smoothly without any complications to the patient. The patient was extubated on the operation table, she regained her consciousness in the recovery room without any neurological deteriorations. Then, she was shifted to the neurointensive care unit for 24 hours under observation.

Post-operative Period Follow-up

On day zero, her left side motor power was 3/5 on both extremities. On the next day, a slight improvement of the left upper and lower extremities power that was 4/5, no dysphasia, her visual acuity was no perception to light. The drain container contained only a few millimeters of serous fluid, so the drain was removed after 14 hours of the operation. An immediate control brain CT-scan was requested done that revealed a parenchymal defect on the right frontal lobe with minimal pneumocephalus at surgical bed without any significant hematoma. The residual tumor could not be seen clearly on the right frontal lobe. On the second post-operative day, the left side of body powered the same of the 1st day. The patient was able to formulate the speech, oriented, and her visual acuity was improved up to hand movement. She was shifted to the ward. Her daily clinical progress in the ward on next couple of days was smooth.

Another control brain CT scan performed 1 week after resection demonstrated a cavity without identifiable tumor and partial resolution of the cerebral edema. No significant changes were found in comparison to the first post-operative imaging. The patient was discharged home on Day 4 with a good general condition. Instructions were giving to her family.

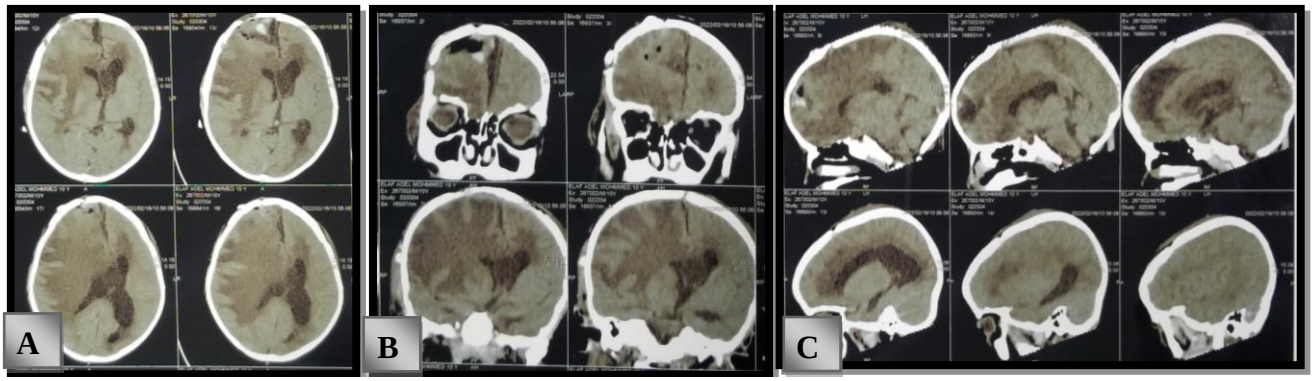

Figure 4: Control Brain CT scan on Day 1 post-operation. A: Axial cut, B: Coronal cut, C: Sagittal cut.

Pathological Examination

Figure 5: Gross picture of the tumor

Gross examination of the fresh tumor specimen was significant for multiple pieces of white-tan soft and friable tissue measuring 5 x 5 x 2 cm. Figure 5 The tissue was fixed in 10% formalin and processed for paraffin embedding. The paraffin blocks were sectioned at 4-m intervals, and these sectionswere stained with hematoxylin and eosin for microscopic examination .

Histologically, it demonstrated an ill-defined tumor proliferation composed of sheets and papillary configurations of atypical polygonal to stellate cells with patchy aggregates of large bizarre multinucleated forms. The tumor cells show abundant pale and eosinophilic to clear/foamy cytoplasm, irregular vesicular nuclei, and prominent nucleoli with frequent mitotic figures, wide areas of geographic necrosis, and few calcifications. This pathological was consistent with choroid plexus carcinoma (CPC) WHO grade III. The patient was referred to the Yemeni National Oncology Center. She started receiving an adjuvant chemo/radiotherapy treatment.

Serial MRI studies of the head and spine with gadolinium contrast administration were performed periodically in the outpatient setting. After Serial MRI control studies of the brain and spine with gadolinium contrast administration were performed periodically at the first, fifth and eighth months post surgery in the outpatient setting, both brain and spinal MRI were normal apart from previous surgical changes and eight months to the surgical resection, both brain and spinal MRI were normal apart from previous surgical changes.

One-year post tumor resection, the brain MRI showed a newly developed small focal enhanced area in the right frontal lobe, just anterior and medial to the surgical bed and measuring about 1.7x1.5x1.8 cm, which suggests a recurrent lesion.

Discussion

Choroid plexus tumors typically arise typically within ventricles. They form only a small number of brain tumors. Sometimes, they may arise from unusual extraventricular tissues as intraparenchymal cerebral tissue. Choroid plexus carcinoma is a high grade tumor which occurs predominantly in pediatric. Extraventricular CPC is extremely rare. Only a few number of pediatric patients with extraventricular CPCs were reported [10,11]. The first two cases reported a CPC, presenting an intraparenchymal CPC, were occurred in young girls, both of them were 6-year-old and within the frontal lobe [10,12]. The first CPC case located in the left frontal lobe, reported in 2001 by Carter et al. [10]. The second case situated in the right frontal lobe, reported in 2009 by Stevens et al. [12]. Another case was reported in 2021 in a 15-year-old teenager with an intraparenchymal CPC located mainly within the left frontal lobe [11].

Our case occurred in a 10-year-old girl presenting as intraparenchymal mass within

the right frontal lobe. The exact mechanism for the origin of this extraventricular tumor is still unknown. Some publications suggested theories and possible causes on how this tumor arise from unusual extraventricular location. The existence of primitive ectopic secretory choroid plexus epithelium in extraventricular brain tissues was suggested as a possible cause [13,14]. It may also arise from ependymal tissue that was separated during later stages of brain development [15].

The diagnosis of this case was not very easy because the radiological picture and the location of the mass made us suggest that the diagnosis is most likely to be an astrocytoma or high grade glioma with a wide variety of differential diagnosis for histopathology. After confirmation of the diagnosis by histopathology, the patient had to be taken to Oncology center to start chemotherapy and radiation therapy. In the beginning, the patient showed a very good outcome. However, with follow up by serial MRI, the patient had recurrence, which is something expected as it occurred in most brain lesions. In comparison, a previously reported case of extraventricular CPC showed similar recurrence 3 months post operation [10]. In some studies of usual CPTs, it was found that CPC is 20 times more likely to recur after treatment than other CPTs [16].

We faced multiple difficulties during the follow-up of this case. On the one hand, the family of the patient is of a low class and live in a far rural area located at the site of conflict and war. There are no near neuroimaging facilities around their residence. The cost of serrial neuroimaging beside the cost of traveling to our institution both made the regular follow-up very difficult. On the other hand, there was a period of time in which we lost the communication with the family and came lately with a recurrent lesion. Moreover, this type of cases needs to be monitored very closely with serial MRI to treat any recurrent lesion very early.

Conclusions

This study is made on a case of extraventricular, intraparenchymal CPC, presented as an emergency case with a huge mass, midline shift and hydrocephalus. Proper diagnostic facilities like histopthology plays an essential role in establishing the diagnosis of CPC. Furthermore, a complete to near complete excision of the CPC in extraventricularlocation followed by adjuvant Chemotherapy shows promising results. MDT is an important step to maximize the benefits for the patient and enhance the outcome. The regular follow up with serial MRI can identify the recurrence early as CPC has higher rate of recurrence. The unique presentation of this rare extraventricular CPC provides insight for the diagnosis and treatment of other rare instances of CPTs. More studies about this cancer with unusual locations are very important to be published to improve the outcome and to know the various presentations and complications of every case.

Informed Consent

A written informed consent was obtained from the patient's family members for publication of this case report and

accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request**.**

Conflict of interest statement

The authors declare that the article content was composed in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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