Case Report

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Choroid Plexus Carcinoma in Early Childhood: A Fatal Consequence in a Case Exhibiting Leptomeningeal Dissemination

Pathology Section

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ABSTRACT

Choroid Plexus Carcinoma (CPC) is a rare, extremely aggressive childhood intraventricular malignancy which makes up less than 1% of paediatric brain neoplasms. It has the features of fast growth with high mitotic activity and a susceptibility for Cerebrospinal Fluid (CSF) spread. The prognosis is poor, despite the early recognition and early response. Hereby, the authors present a case of a two year and five month-old female child with progressive macrocephaly, left-sided weakness, dysphagia and loss of voice. Neuroimaging showed an intraventricular mass with leptomeningeal spread, provisionally diagnosed as choroid plexus neoplasm. An emergency craniotomy, tumour resection and External Ventricular Drain (EVD) placement were done. A friable, greyish, highly vascular mass was excised intraoperatively. Histopathology revealed crowded papillae merging into sheets, high cellularity, necrosis, nuclear pleomorphism and brisk mitotic activity. Immunohistochemistry showed cytokeratin and Ki-67 positivity with Glial Fibrillary Acidic Protein (GFAP) negativity and confirmed CPC (WHO Grade III). The child underwent extensive excision but had a rapidly progressive postoperative decline and died from the illness. This case highlights the aggressive nature of CPC in children, underlines the essential role of histopathology and immunohistochemistry in diagnosis and highlights the dismal prognosis even with an urgent surgical management. Documenting such cases contributes to the limited literature and underscores the need for improved treatment strategies.

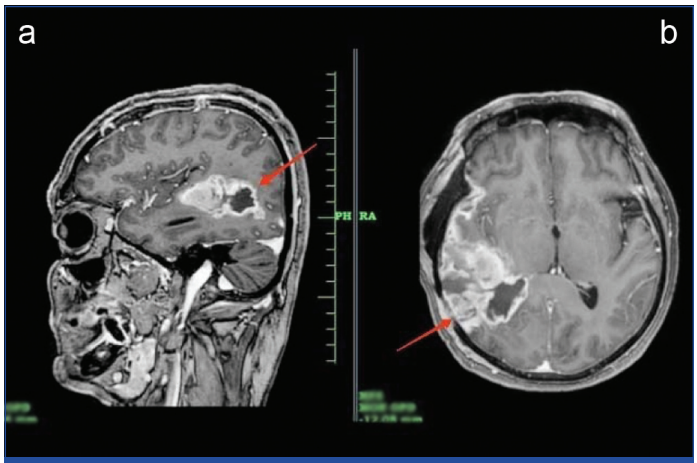
Keywords: Cerebrospinal fluid dissemination, High-grade intraventricular tumour, Hydrocephalus, Ki-67 proliferative index, Paediatric neuro-oncology

CASE REPORT

A two-year-five-month-old girl presented to the hospital with a two month history of progressive neurological symptoms. Before admission, her parents had registered a gradual decrease in movement. One month later, a progressive growth in head size and weakness of the left upper extremity was observed. At that point she had been treated symptomatically with oral analgesics for irritability, antipyretics for intermittent fever and nutritional supplements. Features of raised intracranial pressure were treated by acetazolamide and the child's condition was closely observed. Even with these measures, her situation increasingly deteriorated and she developed left-sided hemiparesis. Eight days before admission, she began to have difficulty in swallowing and loss of her voice, which were indications of continued neurologic decline. On admission, there was significant neurological distress in the child. Her Glasgow Coma Scale (GCS) measured E2V3M4, as the child had shallow eye-opening, unintelligible verbal responses and withdrawal to painful stimuli. She was drowsy, irritable and unresponsive to verbal commands. Motor examination showed left-sided hemiparesis with decreased tone and power, exaggerated deep tendon reflexes and extensor plantar response. Cranial nerve examination revealed bulbar dysfunction with dysphagia and aphonia. Progressive macrocephaly with a tense anterior fontanelle, dilated scalp veins, intermittent vomiting and irritability were consistent with elevated intracranial pressure.

Laboratory tests also showed mild anaemia (Haemoglobin- 9.8 g/dL) and increased Lactate Dehydrogenase (LDH- 480 U/L), suggestive of high tumour turnover. Serum tumour markers, including Alpha-Fetoprotein (AFP) and beta-human Chorionic Gonadotropin (β-hCG) were within normal limits, ruling out germ cell tumours. Preoperative lumbar puncture Cerebrospinal Fluid (CSF) analysis revealed elevated protein (128 mg/dL), reduced glucose (40 mg/dL) and mild lymphocytic pleocytosis (18 cells/mm^3). No organisms were detected on Gram stain or culture and infectious meningitis was ruled out.

The MRI of the brain showed a massive intraventricular mass with heterogeneous enhancement, irregular margins and prominent vascularity, consistent with hydrocephalus. There were also signs of leptomeningeal spread, suggesting aggressive biological behaviour. Radiological differential diagnoses included choroid plexus neoplasm (papilloma or carcinoma), ependymoma, central neurocytoma, subependymal giant cell astrocytoma, medulloblastoma with atypical ventricular location and meningioma. Rare intraventricular metastases due to systemic malignancies like neuroblastoma, Wilms' tumour or leukaemia were also entertained [Table/Fig-1].

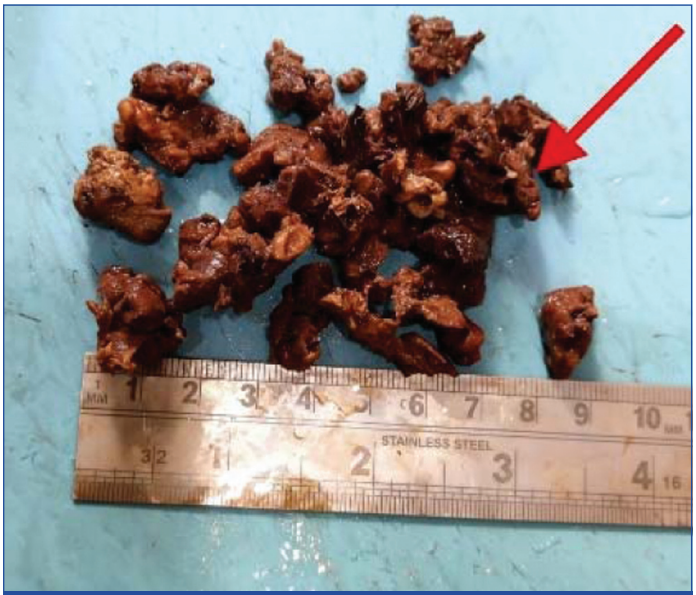


[Table/Fig-1]: MRI brain demonstrating a large intraventricular mass: a) Sagittal view showing a heterogeneously enhancing lesion with irregular margins occupying the ventricular cavity; b) Axial view revealing associated hydrocephalus and leptomeningeal spread.

As of the onset of the neurological deterioration an emergency craniotomy with tumour excision and placement of an EVD was conducted. Intraoperatively, the mass filled the ventricular cavity and appeared greyish, soft, friable and highly vascular. During dissection, massive haemorrhage took place, requiring strict haemostatic

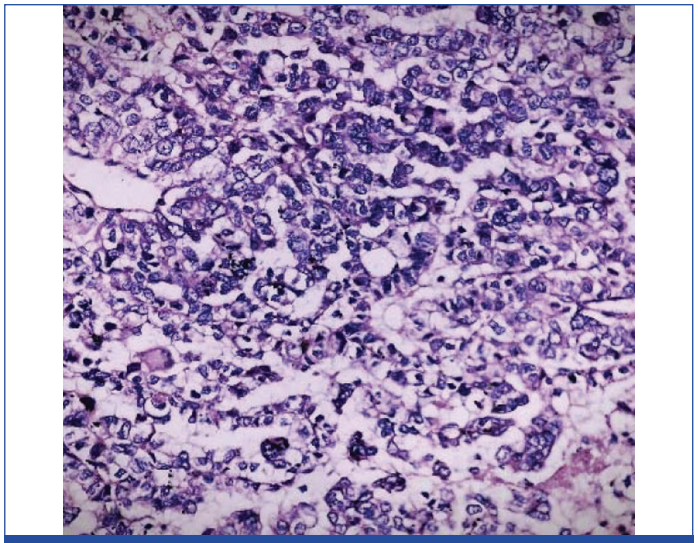
control. Despite these technical issues, gross total excision had been accomplished and the specimen was sent for histopathological analysis. The EVD was externalised for postoperative CSF drainage. Upon postoperative drainage, the clear, xanthochromic CSF showed elevated protein levels and reduced glucose levels consistent with malignancy. The average daily CSF output was 80-120 mL. For the management of intracranial pressure and for regular monitoring of CSF dynamics the EVD was kept for seven days. Over this time, the child developed intermittent fever and irritability, suggesting ventriculitis, but the repeated CSF cultures were sterile. Mild electrolyte abnormalities were observed including hyponatraemia that was corrected with supportive treatment. Gradual clamping of the drain was attempted on the fifth postoperative day in order to assess tolerance however the child was neurologically unwell, which led to continued drainage. Eventually, the catheter was removed after seven days after CSF output decreased and the intracranial pressure stabilised temporarily.

The excised specimen was examined grossly, that showed multiple irregular greyish-brown tissue fragments, aggregating in size of 7×5×1.5 cm, with individual fragments measuring between 0.5 and 2 cm in diameter [Table/Fig-2]. Microscopic observation revealed that papillary areas are densely packed and branched to form sheets of neoplastic cells with extensive areas of necrosis. Tumour was very cellular with cells grouped in complicated papillary and solid form. The neoplastic cells were cuboidal and columnar with indistinct cell borders, scant to moderate cytoplasm and hyperchromatic nuclei demonstrating pleomorphism. Nuclear characteristics were irregularly shaped, coarse chromatin and with occasional prominent nucleoli. Mitotic activity was high with an average 15-20 mitoses per 10 high-power fields and frequent atypical mitotic appearances. Several apoptotic bodies were found as well as necrotic tracts of vast proportion, indicating a strong tumour biology which would be aggressive. Significantly, the normal papillary architecture was lost with fused and distorted papillae forming solid sheets. The lack of polarity, multiple folds of tumour cell layers, aberrant nuclei and sporadic multinucleated shapes were consistent with high-grade malignancy [Table/Fig-3,4].

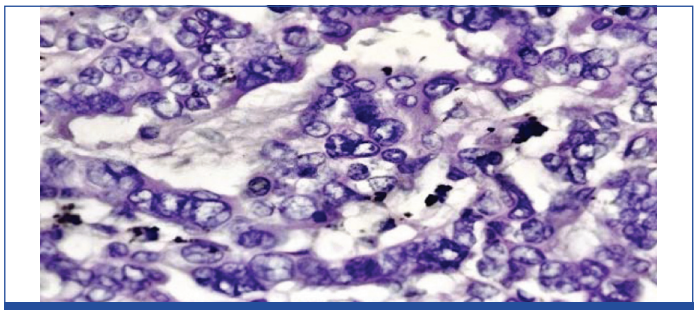


[Table/Fig-2]: Gross specimen showing multiple irregular, friable, brownish tumour fragments obtained intraoperatively.

The histopathological differential diagnosis was limited to malignant intraventricular tumours. Papillary ependymoma was considered but has perivascular pseudorosettes and GFAP positivity. Metastatic papillary carcinoma from other sites including kidney, thyroid or lung was also considered but excluded due to clinical context and immunoprofile. The differential diagnosis also included central neurocytoma with neuronal differentiation and synaptophysin positivity



[Table/Fig-3]: Section showing densely cellular sheets of tumour cells with marked nuclear pleomorphism, hyperchromasia, and loss of papillary architecture (Haematoxylin and Eosin (H&E),10X).

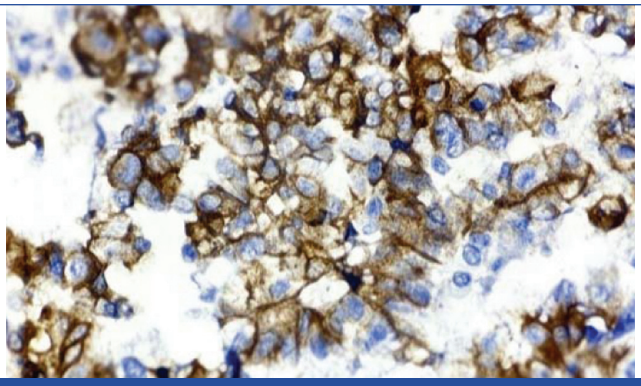


[Table/Fig-4]: Section showing disorganised architecture with densely cellular sheets of tumour cells with marked nuclear pleomorphism, hyperchromasia and high mitotic activity (H&E,40X).

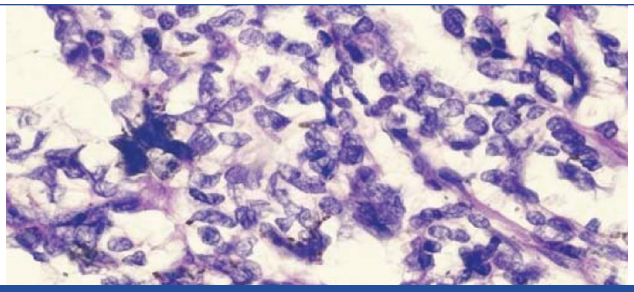
and intraventricular meningioma, typically EMA positive and with lower mitotic activity. Tumour lineage and grade were confirmed using immunohistochemistry. The tumour cells showed strong positivity for cytokeratin, confirming epithelial differentiation and GFAP negativity that excluded glial tumours such as ependymoma. Ki-67 proliferative index was markedly increased at about 60-70%, further reflecting high proliferative activity. EMA was not involved in this case, while synaptophysin was not necessary due to morphology and cytokeratin positivity. Taken together, papillary epithelial morphology, strong cytokeratin positivity, Ki-67 index scores and GFAP negativity are well documented as corroborating the diagnosis of CPC (World Health Organisation (WHO) Grade III). The type was designated according to WHO recommendations that describe the various types of tumours in choroid plexus classified by the WHO into papilloma (Grade-I), atypical papilloma (Grade-II) and carcinoma (Grade-III) [Table/fig 5-7].

In this case, the diagnosis of CPC was supported by high cellularity with loss of normal papillary architecture, nuclear pleomorphism and hyperchromasia, rapid mitotic activity, such as atypical figures, extensive necrosis and apoptosis and a Ki-67 index of 60-70%, significantly increased compared with the normal and papilloma or atypical papilloma. Markers of S-100 and Transthyretin TTR were not tested. Given its expression in a wide variety of tumours, S-100 lacks diagnostic specificity and is of limited utility in this setting. TTR, though a marker of choroid plexus epithelium, may present variable or decreased expression in high-grade CPC and is thus not a pre-requisite for the diagnosis of malignancy. Instead, the selected panel such as cytokeratin, GFAP and Ki-67 had directly addressed the major differential diagnoses and served as compelling evidence for the diagnosis.

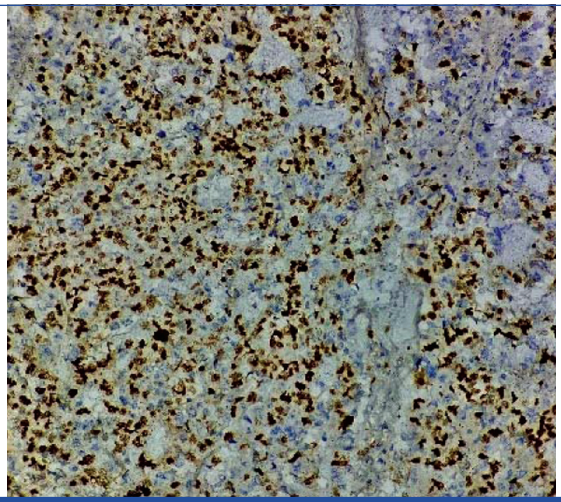
The postoperative course was stormy even after the total excision was complete. The child was irritable and febrile with variable vital parameters and episodes of tachycardia and tachypnoea which were quickly progressing into progressive hypotension.



[Table/Fig-5]: Immunohistochemistry showing strong cytoplasmic and membranous positivity for cytokeratin in tumour cells, confirming epithelial differentiation.



[Table/Fig-6]: Immunohistochemistry showing absence of Glial Fibrillary Acidic Protein (GFAP) staining in tumour cells, excluding glial differentiation.



[Table/Fig-7]: Ki-67 Immunohistochemistry showing strong nuclear positivity in tumour cells, indicating high proliferative index that underscores the aggressive biological behaviour of Choroid Plexus Carcinoma (CPC) (WHO Grade III).

On the neurological side, she remained drowsy and with poor responsiveness and reduced her GCS score from E2V3M4 on admission to E1V2M3 on the seventh postoperative day with worsening brainstem dysfunction. Since WHO grade III CPC usually needs multimodal chemotherapy with carboplatin, etoposide and vincristine, adjuvant chemotherapy was contemplated. However, the deteriorating neurological status and systemic failure of the child precluded initiating chemotherapy. Radiotherapy was discussed but was deferred because of her very young age, risk of severe neurocognitive sequelae and her unstable postoperative condition. Supporting treatment included intravenous fluids, electrolyte correction, antipyretics, broad-spectrum antibiotics, nutritional support and intensive care monitoring. Given the aggressive tumour biology, small therapeutic window and no neurological recovery, however, following discussion with the family, we pivoted the focus of treatment to palliative care. On the ninth postoperative day, the child developed sudden cardiorespiratory collapse, probably the result of elevated intracranial pressure with brainstem failure complicated by systemic instability. Despite resuscitative attempts, the patient could not be revived and was declared deceased. The postoperative survival duration was nine days.

DISCUSSION

Choroid Plexus Tumours (CPTs) are very rare intraventricular neoplasms originating from the choroid plexus epithelium and making up less than 1% of all intracranial tumours and have a high predisposition in infants and other young children [1,2]. As per the WHO, these tumours can be divided into choroid plexus papilloma (Grade 1), atypical choroid plexus papilloma (Grade 2) and CPC, Grade 3 [3]. Among these, CPC possesses the rarest and the highest aggressiveness, which is manifested by high growth rate and invasion of the parenchymal tissue with high frequency of spreading to CSF and poor survival [1,4]. Published series consistently reporting that CPCs typically present in less than three years of age children with features of elevated intracranial pressure and hydrocephalus, often with rapid neurological decompensation [2,5]. As such, our patient's age (2.5 years) and the development of progressive macrocephaly, vomiting, irritability and focal neurological deficits are strongly consistent with reported cases. Most of these published cases describe CPC as appearing within the lateral ventricles in children with large, highly vascular intraventricular masses [1,6]. In the present case, Magnetic Resonance Imaging (MRI) showed a substantial intraventricular tumour with heterogeneous enhancement and irregular margins and associated hydrocephalus. But this time this was a distinctive aggressive feature of our patient which appeared in the form of leptomeningeal spread at first presentation. Although CSF spread has been well characterised in CPC and this is considered an obvious evidence of malignant behaviour, not infrequently that has been observed at recurrence or following recurrence of the disease, rather than at diagnosis [4,5,7]. So our early leptomeningeal involvement in our case displays an unusually aggressive biological phenotype when compared to several published series. As a matter of radiologic characterisation, CPCs presented in the literature typically present with heterogeneity of enhancement, necrosis or haemorrhages and poorly defined invasive margins, whereas choroid plexus papillomas typically reflect well-circumscribed frond-like intensely enhancing lesions. Ependymomas, another key differential, typically develop from the ventricular floor, are calcified or cystic and extend through foramina, not with diffuse leptomeningeal seeding at presentation [8,9]. Typical central neurocytomas present in more older children and young adults, typically occur near the septum pellucidum and may have a less vascular, more "bubbly" or calcified appearance [8,10]. Our imaging findings, including high heterogeneity as well as irregular invasive margins, high vascularity and early spread of leptomeningeal tumour, parallel to the findings from aggressive CPCs in previous reports and help in clarifying the difference in form. Histopathologically, the reported descriptions of CPC underscore loss of papillary structure, high levels of cellularity, intense nuclear atypia, strong mitotic activity and wide necrosis. The data in our case closely correlate with these reports. Anatomy of tumour presented with diffuse disorganisation, combined with fused papillae and solid sheets, massive necrosis within the tumour and an estimated mitosis rate between 15 to 20 mitoses per 10 high-power fields, including atypical cases. Most reported cases report varying degrees of necrosis and mitotic activity but the severe necrosis and high mitotic counts in our case place it at the more aggressive end of the pathological spectrum [4,5]. The nuclear characteristics noted in the case we reported of the patient (enlarged pleomorphic nuclei, coarse chromatin, irregular nuclear structures and occasionally prominent nucleoli) also correspond to those we found in high-grade CPC, with additional atypical features such as bizarre nuclei and sporadic multinucleation, indicating especially aggressive tumour biology [1,3].

The WHO Grade III assessment for CPC is performed with particular histological criteria including loss of ordered papillary architecture, increased cellularity, markedly pleomorphic nuclear mass, rapid and atypical mitoses, necrosis and invasive growth [3]. All these

characteristics were present in this case thus meeting specified grading criteria. However, atypical papilloma (Grade-II) identified in the literature shows increased mitoses but is missing the diffuse anaplasia and extensive necrosis we observed here as well. Papilloma (Grade-I) has a wellorganised papillary structure with very little atypia and is still of similar quality [3]. Immunohistochemically, however, published CPC cases consistently exhibit cytokeratin positivity, suggesting epithelial differentiation, whilst absence of GFAP staining helps exclude glial tumours like ependymoma [1,9]. Similar immunoprofile is shown in the current case consistent with report of literature. The Ki-67 labelling index in our patient of 60-70% was significantly higher than reported values for papillomas and in atypical papillomas and exceeded those in a number of CPC series for which the index was found to be frequently 20-50% [4,5]. This highly proliferative index reflects in our patient the highly distressing clinical course and very poor prognosis. In the literature surgical series highlight gross total resection as the most important prognostic factor of CPC [5,7]. Nevertheless, even in published cases where complete excision was obtained, outcomes were limited, especially in the event of CSF dissemination or very high proliferative indices [4,7]. Just like some of the reported cases of aggressive disease, however, our patient experienced accelerated postoperative decrement despite massive total excision, highlighting that surgical management is in many cases inadequate in biologically challenging CPC. Although many published reports report limited survival benefit with adjuvant chemotherapy and in some cases, radiotherapy, our patient's rapidly deteriorating neurological and systemic status did not allow for intervention, a disparity with more stable cases reported in the literature that should have received multimodal therapy [5,7].

The overall survival in this case was death within nine days of surgery, that is inferior to most series, where while survival is limited, a combination of therapy and some forms of treatment may extend survival months to years [4,5]. This abnormal brevity of survival evidently demonstrates the sum effect of several deleterious prognostic events, as described in the literature as a severely young age, extensive leptomeningeal dissemination at presentation, widespread necrosis at the tumour growth site, profound mitotic activity and an extremely high Ki-67 index [4,5,7].

In summary, compared to published cases, there is a common age level of cancer, location, radiologic signs, histological findings of CPC of the current case, but it should be noted that it has early-stage leptomeningeal dissemination, very high proliferative index, extensive necrosis and fulminant clinical course. This comparison underlines both the unique characteristics of CPC and the wide biological spectrum of this tumour, thus emphasising that a subset of cases, including the present case, may exhibit exceptionally aggressive behaviour and poor outcomes despite optimal surgical management.

CONCLUSION(S)

The CPC is an uncommon but very aggressive paediatric intraventricular tumour with a predilection for very young children and a strong tendency for rapid progression and CSF dissemination. The case depicts an unusually fulminant pathophysiology defined by early leptomeningeal spread, extensive tumour necrosis, high mitotic activity and an exceptionally high Ki-67 proliferative index, all of which are adverse prognostic markers. The clinical course was rapidly fatal, despite gross total surgical excision, highlighting the limitations of surgery alone in biologically aggressive disease. Accurate diagnosis depends on both prudent radiological-pathological correlation and appropriate immunohistochemistry to discern CPC from other intraventricular tumours. Timely identification of high-risk pathologic characteristics is vital for effective multimodal therapy and realistic prognostic counselling. The present case contributes to the paucity of literature on CPC and highlights the urgent need for better strategies in the treatment of this devastating paediatric malignancy.

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