RESEARCH PAPER

Choroid Plexus Carcinoma - A Rare Case

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ABSTRACT

Carcinoma of the choroid plexus is an uncommon intracranial neoplasm of neuroectodermal origin with less than 2% of all glial tumours. Around 80% of choroid plexus carcinomas arise in children, in whom they constitute 15–20% of choroid plexus tumours. Up to 30% of children present with metastatic disease at diagnosis. Most common location: lateral ventricles in children and the fourth ventricle in adults [1]. Most commonly present with hydrocephalus due to CSF overproduction and flow obstruction. Due to their rarity, reports on choroid plexus carcinoma most often focus on single case or single-institution experiences with a limited number of patients.[2] Recently, we also encountered a case of right lateral ventricle choroid plexus carcinoma in a young child who was operated in good neurological grade. The clinical presentation, pathology and management are discussed here.

Keywords: Choroid plexus carcinoma, Brain tumour, Surgery, Radiotherapy

How to cite this article: Dr Raghavendra D Sagar, Dr Irappa Madabhavi, Dr Rohit Patil (2026). Choroid Plexus Carcinoma - A Rare Case. International Journal of Drug Delivery Technology;2026;16(52s): 882-886. DOI: 10.25258/ijddt.16.52s.109

INTRODUCTION

Choroid plexus carcinoma (CPC) is a rare tumour. In the total population, choroid plexus tumour represents less than 1% of all brain tumours [1]. Out of all choroid plexus tumours, only 5% is carcinoma; the diagnosis of choroid plexus carcinoma should be made with caution as it more frequently resembles a metastatic papillary tumour, such as from the kidney and thyroid [2].

Most choroid plexus tumours arise in intraventricular, but cases have been reported as arising from ectopic sites such as intracranial but extra-ventricular and in the spinal canal without an intracranial lesion. It has been suggested in a few studies that for older children and adults, a gross total resection is the best method of treatment, followed by adjuvant chemotherapy and radiotherapy [3]. However, surgical resection poses a challenge as it frequently involves an intensely vascular tumour, leading to extreme and heavy bleeding during surgery, thus affecting the patient's survival.

The grading system is classified as below:-

WHO grade I = choroid plexus papilloma, 80%, papillary formation, lack of mitosis <2 per 10 high-power field, and fibrovascular fronds.

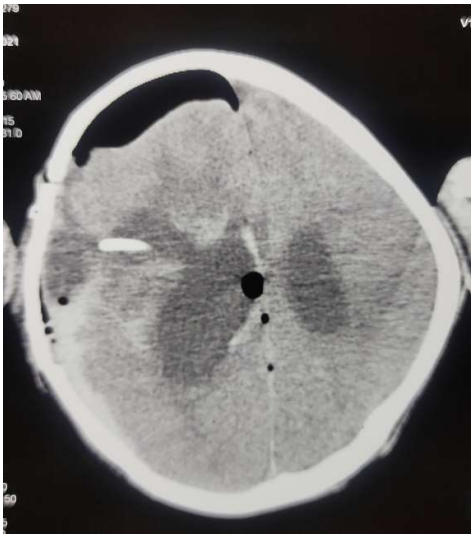
WHO grade II = Atypical choroid plexus papilloma,15%, nuclear atypia, pleomorphism, increased cellularity, necrosis, blurring of papillary pattern, frequent mitoses >2 per 10 high-power field.

WHO grade III = choroid plexus carcinoma, 5%, nuclear atypia, pleomorphism, frequent mitoses >5 per high power field, and invasion of subependymal brain tissue.

Case summary

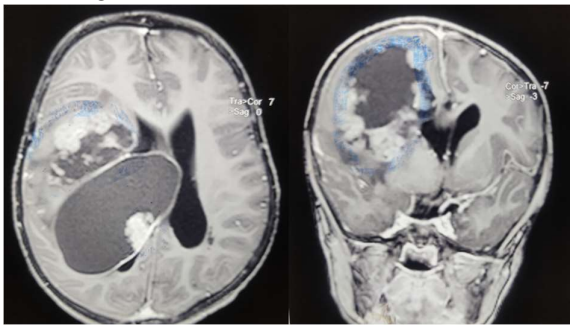
A 3-year-old male child presented to us with a history of weakness of the left side of the limbs both upper and lower limbs, since birth, and the mother complains of an increase in cranial size in the last 1 month after birth. Birth history full term normal delivery, normal milestones, and immunization complete to date. History includes chicken pox at 9 months and a past history of surgery for a brain tumour 2yrs back. Complete blood count and renal profile, liver profile, and electrolytes were within normal limits. At the time of admission, the patient was conscious, with Glasgow coma score of E4V5M6, pupils were bilateral, normal size, and reactive; bilateral papilledema was present. left-side hemiparesis (power:4/5). The rest of the neurological examination was within normal limits.

First CT Brain (08/03/2024) at 1yrs age: Large T2-weighted hyperintense signal intensity cystic lesion measuring 5.5x5.8x4.6cm in the right temporofrontoparietal lobe along the lateral ventricle with significant mass. Suggestive of? Arachnoid cyst? interhemispheric cyst? Porencephalic cyst? With a gross midline shift of 1.7cm towards the left side.



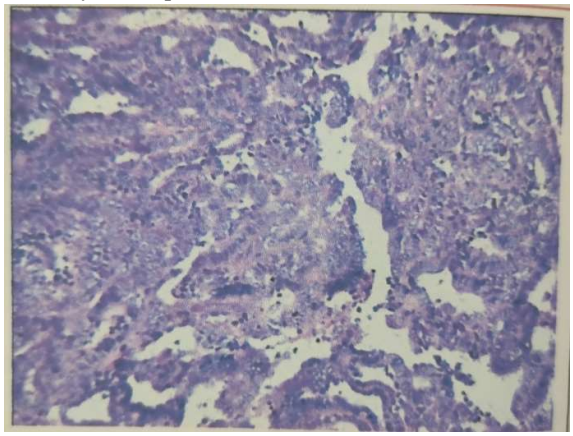
So surgery was indicated and underwent right precoronal burr hole and endoscopic fenestration of the cyst for a right sylvian fissure arachnoid cyst with mass effect. After the first surgery, the patient's condition was fine temporarily, and again after 2 years, the same complaints of increasing head size and weakness of limbs.

Present MRI Brain(18/03/2026): Right temperoparietal craniotomy, post operative cavity right frontal region measuring 2.7x3.6x2.8, medially communicating with the right lateral ventricle and subdural space, subtle diffusion restriction within and hyperintense on T1 suggestive of postoperative cavity with residual lesion along the periphery and small cystic lesion with eccentric soft tissue nodule noted within along the anterior medial aspect of postoperative cavity measuring 1.4x1.5cm.



Surgery was done again, a right frontoparietal craniotomy was made, and gross total excision of the tumour was done. The postoperative period was uneventful. Histology reported as choroid plexus carcinoma grade III and was referred to Radiation treatment.

Histopathology of Choroid plexus carcinoma. On immunohistochemistry, choroid plexus carcinoma will stain positive for cytokeratins and has variable expression for Vimentin, S100, transthyretin, and glial fibrillary acidic protein.



Haematoxylin and eosin staining showing papillary architecture with columnar cells having a high nucleocytoplasmic ratio with prominent nucleoli, fair numbers of mitosis, and areas of necrosis.

Based on the histopathology report, radiation was planned for this patient and completed radiation treatment with IMRT technique to a total dose of 5400cGy/30 fractions on Varian DMX linear accelerator at our institute.

DISCUSSION

Choroid plexus Carcinoma (CPC)is a rare malignant intracranial neoplasm. Bleggi-Torres et al. reported 15 cases of choroid plexus Carcinoma and pointed out that the main symptoms of this tumour are hydrocephalus (62.5%), intracranial hypertension (25%), and convulsion (12.5%) [2]. Neuroradiological features are nonspecific in choroid plexus Carcinoma. Some features may suggest the diagnosis, such as when the tumour invades the parenchyma or presents with metastatic nodules in the third, fourth, or lateral ventricles. The differential diagnosis, including ependymoma, primitive neuroectodermal tumour, Astrocytoma, Teratoma, and Meningioma, can have similar imaging characteristics and modern imaging cannot yet accurately define the pathologic diagnosis.[3]

Most of the time, we do not suspect this entity preoperatively, and the diagnosis is established after a histopathology report. Sometimes, the distinction between choroid plexus papilloma and CPC is not obvious even with histopathology, and in these cases, immunohistochemistry becomes important in establishing the diagnosis, as happened in our case.[4] Choroid plexus papilloma have histologic features that are very similar to those of the normal choroid plexus. The histologic criteria for malignant tumours of the choroid plexus, that is, CPCs, were first developed by Lewis in the 1960s, refined by Russell and Rubinstein two decades later, and most recently modified by the WHO. The established criteria are as follows: (1) obvious invasion of adjacent neural tissue with the infiltrating cells on a stromal base; (2) loss of regular papillary architecture; and (3) evidence of increased mitotic activity, nuclear atypia, and necrosis. [1,3]. CPCs typically stain positive for cytokeratin and display variable expression of vimentin, S100, transthyretin, and glial fibrillary acidic protein (GFAP). Positivity for S100 and transthyretin is typically less than that seen in CPP. CPCs stain positive for GFAP in approximately 20% of tumours [5]. In our case, immunohistochemical staining was positive for GFAP and cytokeratin.

Surgical resection is considered to be the most effective treatment for choroid plexus Carcinoma. The extent of surgical resection remains the most important factor in determining long-term survival in patients with choroid plexus Carcinoma, but patients treated only with surgery have had a very poor outcome as this disease progresses rapidly, and patients often die within 1 year. The early use of radiation therapy may extend survival. Unfortunately, radiotherapy is not an option in the majority of cases because of the young age of the patients and the size of the field to be irradiated. Chemotherapy contributes to long-term survival, but it cannot prevent recurrence. Current data strongly support the use of combined chemoradiation in patients older than 3 years and chemotherapy alone if the patients are younger, but the total amount of necessary adjuvant treatment and the order in which the modalities are to be used are still controversial. Unfortunately, the incidence of CPC is too low to set up a randomized study assessing radiotherapy or chemotherapy protocols for patients with CPC.
Surgical resection is the first-line treatment in all CPCs. Because of small diameters and complex neuroanatomy of the third ventricle, resection of tumours in this area are challenging and highly demands expertise [6]. Due to the high vascularity of these tumours, significant blood loss during resection should be anticipated and effectively managed, especially in the paediatric population [7]. Blood loss can be limited by initial finding and securing of arterial feeders, which are branches of adjacent choroidal arteries, and subsequent coagulation and en bloc or piecemeal removal of tumour bulk [8]. There are some reports of preoperative embolization of the feeder artery; however, this approach is challenging and has potential risks of vessel injury or stroke [9]. Neoadjuvant chemotherapy can be considered as an alternative approach that reduces tumour size and vascularity and facilitates total resection [10].

Optimal management of hydrocephalus in these patients is a matter of debate. The majority of reports suggest an emergent ventriculoperitoneal shunting (VPS) procedure in cases of neurological deterioration and definite resection of the tumour a few days later, after patient stabilization, when facilities for emergent total tumour resection are not accessible. In patients with a stable neurological situation, semi-urgent external ventricular drainage (EVD) placement and tumour resection in one session will be feasible. In these circumstances, EVD tapering and removal or change to VPS should be considered as soon as possible, given the high risk of meningitis with EVD in place [11]. Pre-operative endoscopic third ventriculostomy (ETV) is another option in patients with non-communicating hydrocephalus with different reported success rates. In three previously reported CPCs of the third ventricle, only one report mentioned preoperative VPS placement, and two others did not discuss hydrocephalus management or did not provide individual patient data about this subject. In our case, the patient showed significant neurological deterioration, including drowsiness and lack of response to stimuli upon admission and severe hydrocephalus on imaging. So, because facilities for emergent resection of the tumour were not available, we decided to emergently place a VPS which resulted in a dramatic clinical response. But, our patient experienced shunt related over-drainage in the follow-up period which is a drawback of this approach, and placement of programmable shunt systems instead of fixed pressure valves can be possibly useful to avoid this complication. As an uncommon complication, intraabdominal seeding of CPC tumour cells through ventriculoperitoneal CSF diversion is reported in the literature, and it should be kept in mind in the management of these patients [12]. But, it is a rare complication and cannot be considered as a

contraindication for VPS placement in patients with CPC.

Owing to its low incidence, guideline in managing choroid plexus carcinoma is not well established. The best treatment option is still gross total resection, with Bettegowda, et al. defining it as a reduction of more than 75% in tumour size[13]. Partial resection was defined as 25-75% reduction in tumour size. Gross total resection is achieved in 40-75% in choroid plexus carcinoma as opposed to 95% in choroid plexus papilloma. The difficulty resulting in partial resection is due to a highly vascularized tumour in deep location, making it prone to intraoperative haemorrhage. A review by Sampath et al. has seen that the mean blood loss for total resection is 540ml as compared to 890ml with partial resection. In contrast, they also found the incidence of tumour bed hematoma was higher in total resection, but the extent of excision did not significantly correlate with tumour bed hematoma. Other complications that have been more commonly reported postoperatively include subdural collection (32 to 43%), pneumocephalus (40%), focal deficit (36%), and persistent hydrocephalus requiring CSF diversion surgery more commonly in adults.

Other therapeutic options have been used as an adjunct to surgery, including chemotherapy and radiotherapy, particularly in patients with residual tumours. These adjuvant treatments, however, are not suitable for the young age group of less than 3 years old. Wolff et al have conducted a meta-analysis through a literature review of 566 choroid plexus tumour cases from 1966 to 1998 to determine the treatment modality. They concluded that surgery significantly improve prognosis and radiotherapy significantly gives better survival in choroid plexus carcinoma. Only 8 cases of 22 choroid plexus carcinoma were given chemotherapy and responded, thus the impact of this treatment option could not be sufficiently analysed to be statistically of value. Nonetheless, Berrak, et al. conducted a meta-analysis of 361 choroid plexus carcinomas and found that, out of those given chemotherapy, etoposide is the most effective agent.

Although this tumour is still associated with a poor prognosis, there has been a slight but significant increase in survival throughout the past decades. Dohrmann and Collias reported a 9-month median survival time in a review of 16 children operated on for CPC.[14] In 1992, Packer et al. reported 11 patients with CPC with a 45% event-free survival rate and a median progression-free time of 48 months [14]. Girish et al. reported median survival of 58 months for CPCs who underwent gross total excision with adjuvant therapy and of 36 months who had a subtotal resection with adjuvant therapy.[15]

The outcome of choroid plexus tumours depends on 3 factors, which are: choroid plexus carcinoma histopathology, location of tumour, and extent of resection [16]. However, Wolff et al predicted that the location of the tumour has no prognostic relevance as opposed to Berrak, et al., who found that survival is poorest in infratentorial tumour in choroid plexus carcinoma. Mean survival documented for supratentorial tumours was 26.9% at 10 years, and none for infratentorial tumours. In case of a relapse after primary treatment of choroid plexus carcinoma, it is a poor prognostic factor for survival. 5-year survival of choroid plexus carcinoma is estimated to be 25-30% in patients with gross total resection [17]. Menon, et al. calculated the survival in subtotal resection for choroid plexus carcinoma was 36 months from surgery and 58 months for gross total resection . Thus, we should at best aim for gross total resection or multiple-stage resection to prevent complications.

In our case, the patient was on follow-up after adjuvant radiotherapy for 2 months. Then, a repeat MRI scan at the end of 3rd month showed small residual disease, hence planned for ICE regimen chemotherapy with a good local control of disease so far.

CONCLUSION

Choroid plexus carcinoma is extremely rare in children, and its frequency at ectopic sites, such as in this case, is only reported in a few cases in the literature. This case poses a great challenge to us in terms of diagnosis and management. Total surgical resection was limited due to the tumour's vascularity, and adjuvant treatment was indicated following subtotal or near-total resection with a favourable oncological outcome. In the future, we hope we will have proper guidelines for the management of such cases.

Author contributions: All the authors have approved the article. Dr Raghavendra D Sagar was involved in study design and decided to submit the article for publication, Dr Rohit R Patil in the collection of data, Dr Irappa Madabhavi in the analysis and interpretation of data of writing the report.

Role of funding: No financial support for the conduct of the report was received.

Conflict of interest: The authors declare that there are no conflicts of interest related to this manuscript.

Patient Consent -yes

Acknowledgments: None.

Role of artificial intelligence in creating content: Nil

Ethical approval- Nil since it's a secondary data archived from the databases and all procedures related to data protection and privacy were carried out in compliance with relevant regulations.

Conflict of interest: Nil.

Declaration of Competing Interest:

The authors declare that they have no known competing personal relationships that could have appeared to influence the work reported in this paper.

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