International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Choroid Plexus Carcinoma- Unravelled - A Rare Case Report

Dr. Tibu John Joyse[1], Dr. Arun Kumar G[2], Dr. Kumar G V[3], Dr. Ramya[4]

[1]M.D Paediatrics, Post Graduate, Department of Paediatrics, Sri Siddhartha Medical College
[2]M.D Paediatrics, Affiliation-Assistant Professor, Department of Paediatrics, Sri Siddhartha Medical College
[3]M.D Paediatrics, Professor and Head, Department of Paediatrics, Sri Siddhartha Medical College
[4]M. D Pathology, Post Graduate, Department of Pathology, Sri Siddhartha Medical College

Abstract: ***This article details a rare case of Choroid Plexus Carcinoma CPC, a malignant CNS WHO grade III epithelial neoplasm, diagnosed in a 5-year-old boy. CPCs, accounting for 34.4 of Choroid Plexus Tumors, predominantly affect children and are associated with poor prognosis, frequently arising in the lateral ventricles. Our case report underscores the clinical presentation, diagnostic challenges, and treatment modalities of CPC, emphasizing its rarity and the critical role of surgical intervention. Despite comprehensive treatment, including surgery and adjuvant therapies, the patient succumbed three months post-operation, highlighting the aggressive nature of CPC and the urgent need for improved diagnostic and therapeutic strategies. This case contributes to the limited literature on CPC, offering insights into its management and prognosis.***

Keywords: Choroid Plexus Carcinoma, Pediatric Oncology, Surgical Treatment, Prognosis, CNS Neoplasms

1. Introduction

Choroid plexus tumours (CPT) are rare intraventricular papillary neoplasms arising from choroid plexus epithelium.[1]

Choroid plexus carcinoma (CPC) is a rare malignant epithelial neoplasm (CNS WHO grade III) accounting for 34.4% of Choroid plexus tumours.[2]A majority of CPCs (80%) occur in children and are associated with a poor prognosis. They tend to form in the posterior fossa. [2,3]

Reports on CPC are uncommon; hence they usually concentrate on single cases or experiences from a single institution with a small number of patients.[5]

We report a case of CPC diagnosed in a 5-year-old boy.

2. Case Report

A 5-year-old male child presented to our OPD with complaints of fever since 10 days, headache and vomiting since 4 days, irritability and generalized weakness and since 3 days .H/O constipation and urinary incontinence also noted since 3 days.At the time of admission, patient was conscious, with GCS of E2V2M5.

On examination; no apparent cranial nerve abnormalities except defective vision were noted. Generalized hypotonia was noted. Rest of the neurological examination was within normal limits.

Both pupils were normal in size and reactive to light and a bilateral Grade IV Papilledema present.

An initial diagnosis of atypical teratoid tumour was made.

Clinical course

- MRI brain showed features suggestive of malignant etiology- likely Atypical Teratoid/ Rhabdoid tumour/ Supratentorial Ependymoma. Right side midline shift with effaced left lateral ventricle present.
- Blood investigations revealed CBC counts and CRP levels within normal limits
- Sodium levels in the higher range of normal.
- NS1 antigen and peripheral smear examination for malarial parasite were negative.

Treatment:

- The child was treated with IVF- DNS for maintenance, Inj 3% Hypertonic Saline for the Cerebral Edema
- Inj Levetiracetam prophylactically
- Inj Furosemide and Inj Dexamethasone
- Empirical antibiotics

Patient was later transferred to NIMHANS for further surgical management

In our case, the patient was operated in low GCS, i.e., E2V2M5 and died 3 months after operation but due to loss in follow up we could not identify the exact cause of death in this case.

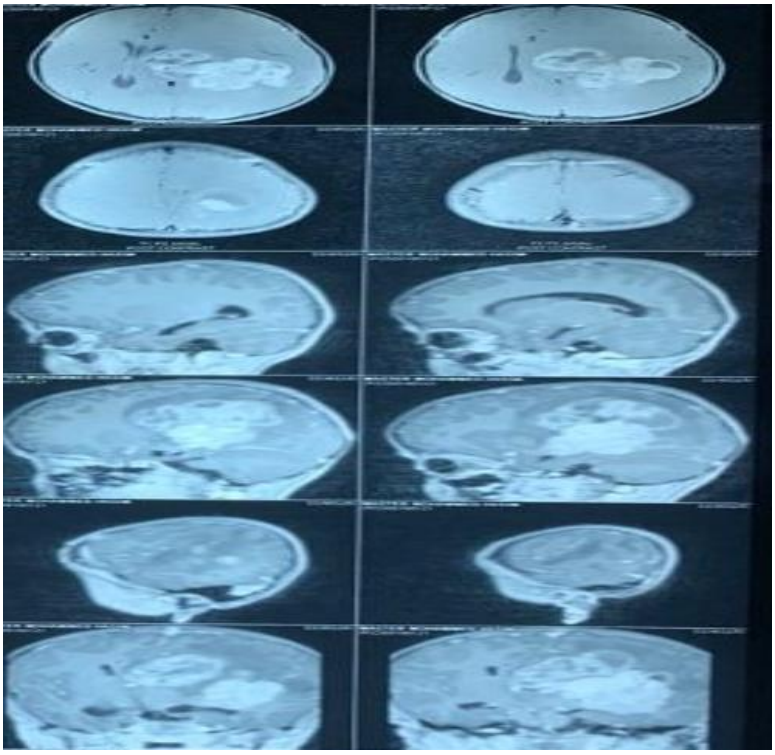

Figure 1: Areas of blooming are noted within the mass- suggestive of hemorrhage. Few areas of restricted diffusion are noted within the mass.

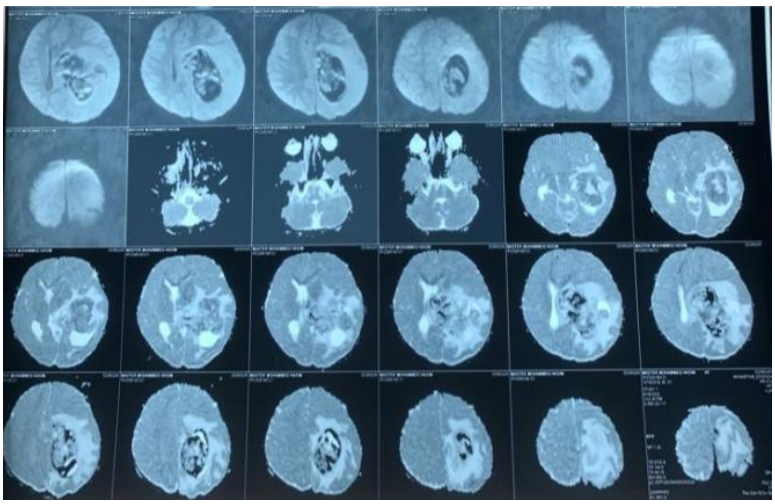

Figure 2: A large lobulated solid -cystic mass likely arising from the septum pellucidum in the midline and extending towards the left cerebral hemisphere showing heterogeneous contrast enhancement. Midline shift is noted to the right in the form of effacement of the left lateral ventricle with significant perilesional edema.

3. Discussion

CPC is a CNS WHO grade III epithelial neoplasm arising from choroid plexus epithelium.[1,2] The majority of cases involve young children, with a median presenting age of three years.[2,4]

Most cases occur in lateral ventricles followed by fourth ventricle. Rarely, CPCs can develop in the cerebellopontine angle close to the Lushka foramina; in those rare instances, they may manifest as an intraparenchymal or suprasellar mass.[6,7]CPCs present with cerebrospinal fluid (CSF) pathway obstruction causing symptoms related to hydrocephalus including, nausea, vomiting , headache and enlarged size of the head.[2,8] On MRI, they show up as massive intraventricular lesions with irregular enhancing margins, edema in the surrounding brain, hydrocephalus, and a disseminated tumor and a heterogeneous signal on T2 and T1-weighted imaging. [2]

CPCs are associated with losses of chromosome 5, 10q,18q and 22q and gains on 1, 4, 8q, 9p, 12, 14q, 20q and 21.[7] It is advised that TP53 germline mutation testing and genetic counselling be made available to all CPC patients and their families.[2] The main differential diagnosis of an intraventricular papillary tumour includes atypical teratoid/rhabdoid (AT/RT) tumours, papillary variant of ependymoma, papillary meningioma and metastatic tumour.While some choroid plexus papillomas also show nearby cerebral edema and invasion, neuroradiological characteristics in CPC are not specific. Some features, such as the tumor's invasion of the parenchyma or the presence of

metastatic nodules in the third, fourth, or lateral ventricles, may help identify the diagnosis. Despite advancements in imaging technology, the pathologic diagnosis of any of the tumors in the differential diagnosis, such as meningioma, ependymoma, teratoma, astrocytoma, and primitive neuroectodermal tumor, cannot be precisely defined by modern imaging method.[5,9]In most cases, the diagnosis is made after the histology report, and we do not suspect this entity before surgery. In some cases, even histopathology cannot definitively distinguish between CPC and choroid plexus papilloma; in these cases, immunohistochemistry plays a critical role.[3]A patient's ability to survive depends on the extent of their resection, which is the cornerstone of care. In terms of prognosis and long-term survival, gross total resection (GTR) is the most critical element. Greater success rates are observed in patients who have received GTR as opposed to those who have undergone subtotal resection.[10]After surgery, adjuvant chemotherapy can be used to decrease the tumor tissue, although there isn't much experience with this treatment. Although radiation after surgery is linked to a higher chance of survival, young children should not receive this kind of treatment.[10]CPCs are associated with a poor prognosis.[11]40–50% of patients survive after five years. Due to the tumor's tendency to spread through CSF routes and infiltrate the nearby brain tissue, GTR is challenging in every instance. The survival rate of CPCs with TP53 mutations is significantly lower.[7]

4. Conclusion

Choroid Plexus Carcinomas CPCs represent a formidable challenge in pediatric neuro-oncology due to their aggressive nature and poor prognosis. This case report of a CPC in a young child underscores the critical importance of early and accurate diagnosis, the central role of surgical intervention in management, and the need for further research into adjuvant therapies to improve outcomes. Despite advancements in neurosurgical techniques and supportive care, the survival rates for CPC patients remain low, particularly in cases where complete resection is not feasible. This case highlights the necessity for ongoing research into the genetic and molecular underpinnings of CPC to develop targeted therapies and improve prognostic outcomes. Furthermore, it underscores the importance of a multidisciplinary approach to care, encompassing surgery, potential adjuvant therapies, and supportive care to address the comprehensive needs of patients with this challenging diagnosis.

Conflict of Interest: Nil

Funding: Nil

References

[1] Jaiswal S, Vij M, Mehrotra A, Kumar B, Nair A, Jaiswal AK. Choroidplexustumors: A clinico-pathological and neuro-radiological study of23 cases. Asian J Neurosurg. 2013;8(1):29–35
[2] Louis D, Wiestler O, Cavenee W, Ohgaki H. WHO classification oftumours of the central nervous system. Lyon: International Agency for Research on Cancer; 2021. p.195-198.
[3] Singh A, Vermani S, Shruti S. Choroid plexus carcinoma: Report oftwo cases. Indian J PatholMicrobiol. 2009; 52(3):405–7.
[4] Mishra A, Srivastava C, Singh SK, Chandra A, Ojha BK. Choroidplexus carcinoma: A case report and literature review. J PediatrNeurosci. 2017;7(1):71–3
[5] Bleggi-Torres LF, Urban LA, Antoniuk A, Carboni P, Ramina R, Gugelmin ES. Choroid plexus carcinoma: Report of 15cases. *ArqNeuropsiquiatr.* 2000;58:505–11.
[6] Gopal P, Parker JR, Debski R, Parker JC. Choroid Plexus Carcinoma.ArchPathol Lab Med. 2008;132(8):1350–4.
[7] Greenfield J, Love S, Budka H, Ironside J, Perry A. Greenfield'sneuropathology. 9th ed. Boca Raton: CRC Press; 2015. p. 1709
[8] Kim T, Park MR, Hong EK, Gwak HS. Choroid Plexus Carcinoma inAdults: Two Case Reports. Brain Tumor Res Treat. 2019;7(1):48
[9] Menon G, Nair SN, Baldewa SS, Rao RB, Krishnakumar KP, Gopalakrishnan CV. Choroid plexus tumors: An institutional series of 25 patients. *Neurol India.* 2010;58:429–35.
[10] Jameel P, Varma A, Kumari P, Vagha K, Vagha J, Damke S. Choroidplexus carcinoma in an adolescent male: a case report. Journal ofMedical Case Reports. 2021;15(1)
[11] Wolff J, Sajedi M, Brant R, Coppes M, Egeler R. Choroid plexustumours. Br J Cancer. 2002;87(10):1086–91