

# Choroid Plexus Carcinoma: A Case Report

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### ABSTRACT

The tumors arising from choroid plexus epithelium are more common in the pediatric age group. Choroid plexus tumors are Choroid plexus papilloma (WHO grade I), Atypical choroid plexus papilloma (WHO grade II) and Choroid plexus carcinoma (WHO grade III). In children, choroid plexus carcinoma constitutes 15 to 20% of the choroid plexus tumors. These aggressive tumors have tendency for recurrence and metastatic dissemination along the cerebrospinal fluid (CSF) pathway. We report a case of Choroid plexus carcinoma in 1 year female child who presented with hydrocephalus.

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## Introduction

Choroid plexus tumors represent 0.3 to 0.6% of all tumors of central nervous system. These tumors were first described by Geurard in 1832<sup>[1]</sup>. They constitute nearly 5% of brain tumors occurring in childhood and 20% of the brain tumors occurring within the first year of life<sup>[2]</sup>. Rarely they occur as congenital and fetal tumors<sup>[3]</sup>. Among the choroid plexus tumors, papillomas are 5 times more common than carcinomas and most of the carcinomas occur in infants less than 3 years of age. Choroid plexus carcinomas should be distinguished from Choroid plexus papilloma by their growth pattern and histopathological features like papillary architectures covered by layers of pleomorphic choroid plexus epithelial cells, presence of necrosis and increased mitotic activity. Aggressive surgical resection with chemotherapy and with or without radiotherapy is required for survival and has very poor prognosis if the surgery is incomplete.

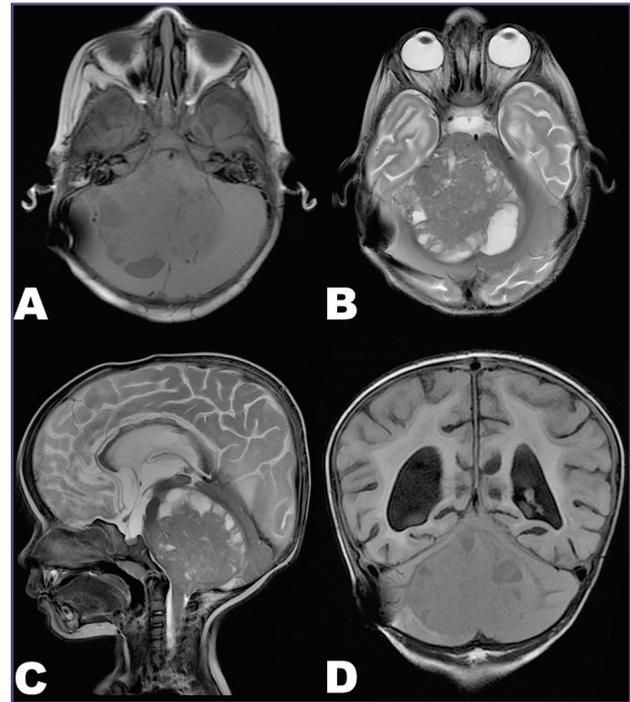
## Case report

A 1 year old girl came to the neurosurgery with history of vomit since 25 days. There was regression of motor milestones and progressive enlargement of head. On examination head circumference was 47cms. Anterior fontanella was open and bulging. Patient had dolicocephaly and right sided Bells palsy. Her heart rate was 122/min regular and respiratory rate was 28/min. On ophthalmic examinations mild papilledema with blurring of disc margin in both eyes were noted. Haematological examination showed normal hemoglobin (13.1gm/dl), raised total count (15,400/cumm) and raised ESR (29mm/hr) remaining parameters were within normal range.

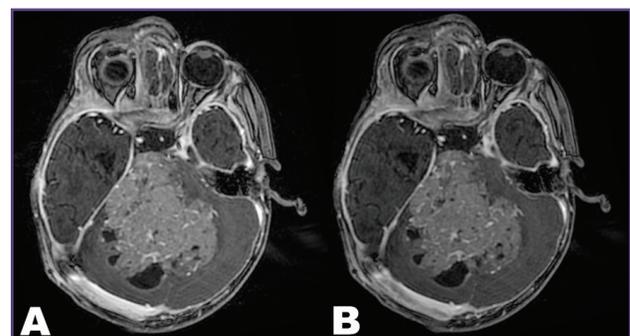
Magnetic resonance imaging (MRI) reveal Axial T1W image showing iso/hypo intense mass in the fourth ventricle, Axial T2 W image showing brain stem displacement and infiltration. There was inferior beaking of the tonsil through foramen magnum suggesting herniation and anteromedial displacement of basilar artery. Coronal FLAIR image showed mass and dilatation of ventricles (Figure 1). Axial post contrast spoiled gradient images showed intense heterogeneous enhancement of the mass (Figure 2). Based upon radiological findings, a clinical diagnosis of right cerebellar medulloblastoma was considered. A right Parieto-occipital craniotomy was done and the tumor was sent to pathology department for histopathological examination.

Grossly we received multiple gray white irregular soft tissue bits. Histopathological examination revealed tumor cells arranged in solid sheets and in papillary pattern. Papillae had delicate fibrovascular core and were lined by several layers of tumor cells. The tumor

cells had round to oval vesicular nuclei with prominent nucleoli. Most of the tumor cells had scant eosinophilic to clear cytoplasm. Atypical mitotic figures and foci of necrosis were seen. The interstitial fibrous septa showed lymphocytic infiltrate. Few foci showed normal choroid plexus lined by benign cuboidal epithelial cells (Figure 3). On immunohistochemistry tumor cells were

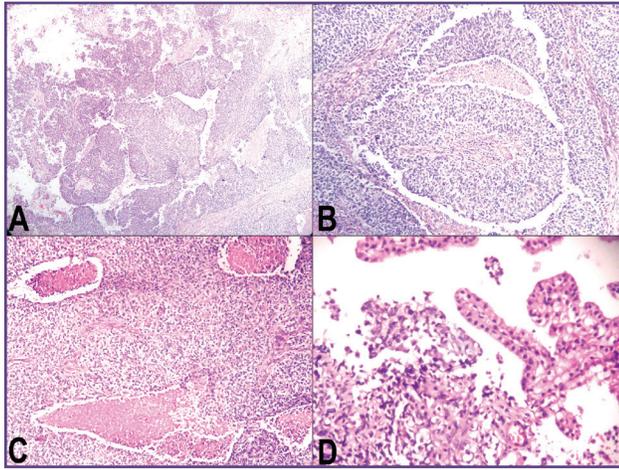


**Fig. 1:** (A) Axial T1W image showing iso/hypo intense mass in the fourth ventricle. (B) Axial T2W image showing heterogeneously hyperintense mass with peripheral cystic areas. (C) Sagittal T2W image showing brain stem displacement and infiltration. Also note the inferior beaking of the tonsil through foramen magnum suggestive of herniation and anteromedial displacement of basilar artery. (D) Coronal FLAIR image showing mass and dilatation of ventricles.

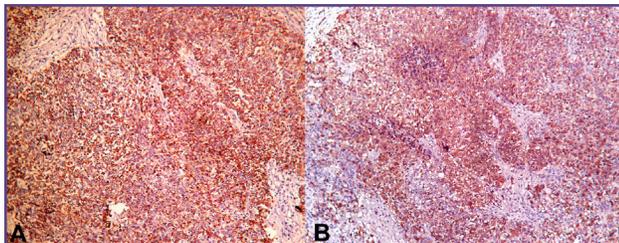


**Fig. 2:** Axial post contrast spoiled gradient images showing intense heterogeneous enhancement of the mass

positive for cytokeratin and S-100 (Figure 4). They were negative for GFAP. The tumor was diagnosed as Choroid plexus carcinoma depending upon the histopathological and immunohistochemical features.



**Fig. 3:** (A) Tumor cells arranged in papillary pattern (H&E, X50). (B) Papillae having fibrovascular core and are lined by layers of tumor cells (H&E, X100). (C) Tumor showing areas of necrosis (H&E, X100). (D) Right side showing benign choroid plexus lined by cuboidal epithelium and left side showing tumor cells (H&E, X400)



**Fig. 4:** (A) Tumor cells are cytokeratin positive (CK, X100). (B) Tumor cells are S-100 positive (S-100, X100).

## Discussion

Choroid plexus carcinoma are rare tumors occurring more often in children<sup>[4]</sup>. These tumors are of neuroectodermal origin and correspond to WHO grade III tumor. They are also considered as congenital brain tumor as few cases have been reported in premature infants<sup>[5]</sup>.

Choroid plexus tumors causes increased intracranial pressure and most of the patients present as hydrocephalus. Some of the cases present with convulsions and intracranial hypertension<sup>[6]</sup>. As the choroid plexus carcinomas invades into the brain parenchyma, the patient may have focal neurological dysfunction.

Choroid plexus carcinoma may arise denovo from the choroid plexus epithelium or may develop from the

preexisting choroid plexus papilloma. These tumors have been found to be associated with Simian virus 40 (SV40) and also dysfunction of p53 which is a tumor suppressor gene. They are also described in a association with Li-Fraumeni syndrome, where the TP53 germ line mutations are carried in families.

Radiological features of choroid plexus carcinomas are markedly enhancing intraventricular tumors invading the adjacent brain parenchyma. On non-contrast computed tomography, these tumors are heterogenous and are typically iso to hyperdense to grey matter. 20 – 45% of cases show calcifications. Contrast enhanced CT shows tumor prominently and is heterogenous with areas of necrosis and cyst formation. On Magnetic resonance imaging, T1 image is iso to hypointense and T2 image is usually iso to hypointense with hyperintense necrotic areas. Before surgery, imaging of entire neural axis should be done as the tumors have CSF seeding<sup>[7]</sup>. In our case as the choroid plexus carcinoma was not suspected clinically, entire neuraxis imaging was not done.

Choroid plexus carcinoma (WHO grade III) should be differentiated from papilloma (WHO grade I) which has papillary configuration. Papillomas mimic the structure of normal choroid plexus but have more exuberant papillae with branching. Papillae have delicate fibrovascular cores lined by single layer of columnar to cuboidal epithelium which have round to oval basally placed monomorphic nuclei and eosinophilic cytoplasm. These benign tumors do not have cellular atypia and necrosis. Atypical mitotic figures are also rarely present. Choroid plexus tumors are termed as “Atypical papilloma” when the cells lining the papillae show pleomorphism, have focal necrosis and occasional atypical mitotic figures.

In choroid plexus carcinoma, the characteristic features are blurring of papillary features, cellular atypia, invasion to adjacent brain parenchyma, areas of necrosis and increased mitotic activity. The presence of features like transitions from normal choroid plexus to abnormal confirms the origin of the lesion which was present in our case. The cellular atypia includes variation in chromatin of the tumor cells, acinar and glandular structures, solid sheets and atypical mitotic figures. Our case showed cellular atypia, blurred papillary structures lined by layers of tumor cells and areas of necrosis. Immunohistochemically, the tumor cells of choroid plexus carcinomas stain positive for cytokeratins, S-100 and Transthyretin. Approximately 20% of these tumor may express GFAP. The tumor cells are typically negative for EMA.

Differential diagnosis for these tumors include papillary variant of ependymoma, papillary meningioma, cerebellar

medulloblastoma and astrocytoma. Papillary variant of ependymoma shows tumor cells having nuclei with stippled chromatin and micronucleoli. Ependymal rosettes and perivascular pseudorosette with tumor cells having their tapered cell processes oriented towards the blood vessel should be present<sup>[8]</sup>. These tumor cells in ependymoma will be positive for GFAP on immunohistochemistry. Our case did not show rosettes or pseudorosettes and the tumor cells were negative for GFAP.

Papillary meningioma (WHO grade III) may occur in pediatric age group and can arise in the choroid plexus<sup>[9]</sup>. On immunohistochemistry, the tumor cells will be negative for cytokeratin. In our case, the tumor cells were strongly positive for cytokeratin on immunohistochemistry. Astrocytomas are GFAP positive tumors and in our case the tumor cells were GFAP negative.

Cerebellar medulloblastomas have tumor cells which are arranged in sheet like pattern. Homer-Wright Rosettes are usually found. The tumor cells have large hyperchromatic nuclei and background is highly fibrillar. The tumor cells are positive immunohistochemically for neuronal markers like NFP, Neu-N and synaptophysin and are negative for S-100 and Cytokeratin.

The mainstay of treatment is surgical resection. The extent of tumor resection is an important factor in determining the long-term survival of patients. Due to hypervascularity and poor demarcation from the adjacent brain parenchyma, the gross total removal of the tumor becomes difficult. In such cases residual tumor can be removed after radiotherapy and chemotherapy. Radiotherapy helps to increase the survival<sup>[10]</sup>. But it cannot be used in many patients due to their younger age and also the size of the field which has to be irradiated. Chemotherapy helps for the long term survival and is indicated in young children with residual tumor after surgical resection and also in patients with metastasis<sup>[11]</sup>.

## Conclusion

Choroid plexus carcinoma is rare tumor occurring in the lateral and fourth ventricles and has poor prognosis. This tumor should be considered as possibility in the intraventricular papillary neoplasm in children. Gross total resection of the tumor is treatment of choice though it is difficult due to increased vascularity and poor demarcation from the adjacent brain parenchyma. Adjuvant

chemoradiation is preferred in patients older than 3 years and chemotherapy alone in younger patients.

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None

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None

## References

1. Hyun-Seung Kang, Kyu-Chang Wang, Yeon- Mee Kim, In-One Kim, Seung-Ki Kim, Je G.Chi, Byung-Kyu Cho. Choroid plexus carcinoma in an infant. JKMS 1997;12:162-7.
2. Louis DN, Ohgaki H, Wiestler OD, Carenee WK. WHO classification of tumors of the central nervous system. Lyon: IARC;2007.
3. Raisanen J, Davis RL. Congenital brain tumors. Pathology (phila) 1993;2:103-116.
4. Mishra A, Srivastava C, Singh SK, Chandra A, Ojha BK. Choroid plexus carcinoma: case report and review of literature. J Pediatr NeuroSci. 2012;7:71-73.
5. Wakai S, Arai T, Nagai M. Congenital brain tumors. Surg Neurol 1984;21:597-609.
6. Bleggi-Torres LF, Urban LA, Antoniuk A, Carboni P, Ramina R, Gugelmin ES. Choroid plexus carcinoma: Report of 15 cases. Arq Neuropsiquiatr 2000;58:505-11.
7. Wolff JE, Sajedi M, Brant R, et al. Choroid plexus tumors. Br J Cancer 2002;87:1086-1091
8. Adesina AM. Intraoperative consultation in the diagnosis of pediatric brain tumors. Arch Pathol Lab Med. 2005;129:1653-1660.
9. Singh A, Vermani S, Sharma S, Chand K. Papillary meningioma: a rare but distinct variant of malignant meningioma. Diagn Pathol. 2007;2:3
10. Menon G, Nair SN, Baldewa SS, Rao RB, Krishna kumar KP, Gopala Krishnan CV. Choroid plexus tumors: An institutional series of 25 patients. Neurol India 2010;58:429-35.
11. Packer RJ, Perilongo G, Johnson D, Sutton LN, Vezina G, Zimmerman RA, Ryan J, Reaman G, Schut L. Choroid plexus carcinoma of childhood. Cancer 1992;69:580-5.