www.jmscr.igmpublication.org
Impact Factor 5.84
Index Copernicus Value: 83.27
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v5i7.174

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

**Original Article**

# Choroid Plexus Carcinoma of Third Ventricle – A Case Report

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**ABSTRACT**
*Choroid plexus carcinomas are extremely rare and rapidly growing intra ventricular tumors which are more frequently seen in young children. Choroid plexus tumors commonly involve lateral ventricles and fourth ventricle and are sometimes seen in third ventricle and extra ventricular location. We report a case of choroid plexus carcinoma arising from third ventricle extending into fourth ventricle in a 13 year old girl and discuss the imaging findings.*
**Keywords:** *Choroid plexus carcinoma- third ventricle- intra ventricular tumors.*

## INTRODUCTION

Choroid plexus is a mass of vascular structure present in cerebral ventricular system. It is responsible for the production and filtration of cerebrospinal fluid and acts as blood –CSF barrier. Choroid plexus is involved in many disease processes that include congenital disorders like diffuse villous hyperplasia, Sturge Weber syndrome, lipoma, choroid cysts and acquired disorders include choroid plexus papilloma and carcinoma, metastases, choroid plexitis, infantile myofibromatosis, xanthogranulomas, haemorr-hage[1]. Tumors from choroid plexus are extremely rare and form 0.4-0.6 % of all intracranial neoplasms and 2-4 % of all pediatric brain tumors[2]. The most common site is lateral ventricle (50 %), followed by fourth ventricle (40 %) and third ventricle (5 %). The extraventricular location include cerebellopontine angle, suprasellar region, pineal gland and cerebellum. About 80 % of choroid plexus tumors are benign papillomas the rest are rapidly growing aggressive carcinomas, which are more common in children. The main clinical features of these tumors are due to hydrocephalus. The other symptoms include seizures, cranial nerve palsies, and focal neurological deficits. These tumors are well defined lobulated masses with areas of haemorrhage and cystic degeneration. Parenchymal invasion and necrosis are commonly

seen in choroid plexus carcinomas.[3] Choroid plexus papilloma (CPP) can be clearly differentiated from choroid plexus carcinoma (CPC) by the presence of typical histological features. CPP shows prominent front of fibrovascular connective tissue. Hypercellularity, conspicuous mitotic activity with invasion into adjacent brain parenchyma is typical of choroid plexus carcinoma. On CT, choroid plexus tumors are is to hyper attenuating intraventricular masses and show profuse contrast enhancement. Extension into other ventricles is typical. Calcification may be seen in some cases. Hydrocephalus is more common. On MR imaging it is possible to differentiate choroid plexus papilloma from carcinoma. Choroid plexus papilloma are hypo to isointense intraventricular masses on T1 weighted images with variable signal intensities on T2 weighted images. Flow voids are common. Due to the presence of necrosis and adjacent parenchymal infiltration Choroid plexus carcinomas show more heterogenous signal intensities with cerebral edema in the adjacent white mattter. Increased metabolic activity is noted in fluorine -18 flurodeoxyglucose PET.

**CASE REPORT**

A 13 year old girl presented with progressive headache of one year duration with associated vomiting and double vision. There was no history of trauma, seizure, bowel and bladder dysfunction. History of swaying while walking to one side was present. On clinical examination the child had signs of raised intracranial tension with up gaze palsy, neck stiffness, axial ataxia, pupils were bilaterally 2mm equally reacting to light. Routine lab investigations and chest radiographs were normal. Contrast CT brain showed an ill defined lobulated heterogeneously enhancing mass 5.5 x 5.1 cms in the posterior third ventricular region. There was mild hydrocephalus with marginal grade1 perilesional edema, MR imaging with contrast revealed a poorly defined profusely enhancing heterogeneous intraventricular mass in third ventricle which is isointense to gray matter on T1 weighted and mildly hyperintense on T2 weighted and FLAIR images. The mass showed no restriction on diffusion or blooming on GRE. A few flow voids are seen with in the mass. Grade 1 perilesional edema and mild dilatation of lateral ventricles were noted. There were few cystic areas within the mass. The lesion extended inferiorly into fourth ventricle. (fig A-F) A provisional intraventricular tumor of third ventricle was made. Ventriculo peritoneal shunt followed by midline suboccipital craniectomy by transvermian approach was performed. A highly vascular and fibrous tumor was noted in the fourth ventricle extending from third ventricle with no cleavage between the tumor and ventricular ependyma. Microscopic sections from the tumor tissue showed large areas of haemorrhage and many thin walled, dilated blood vessels. The tumor cells appeared polygonal to columnar having a papillary architecture at most stratification and multilayering with sheet like arrangement at places. The nuclei showed mild pleomorphism and had speckled chromatin. Few mitoses are seen averaging 4-5/10 hpf. Few fragments of normal cerebellar tissue are also seen. On (IHC-0547-17) the tumor cells showed strong and diffuse positivity for pancytokeratin with focal positivity for vimentin and S100. GFAP and SALL-4 are negative in the tumor cells.MIB-1index was high about 17.6 %. The above features were consistent with choroid plexus carcinoma.

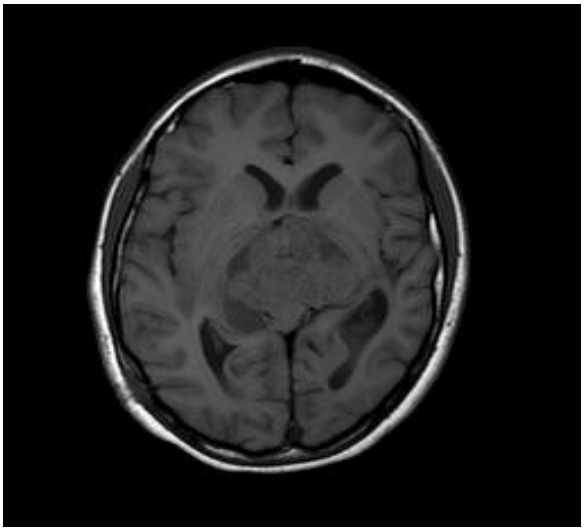

**Fig 1) A**

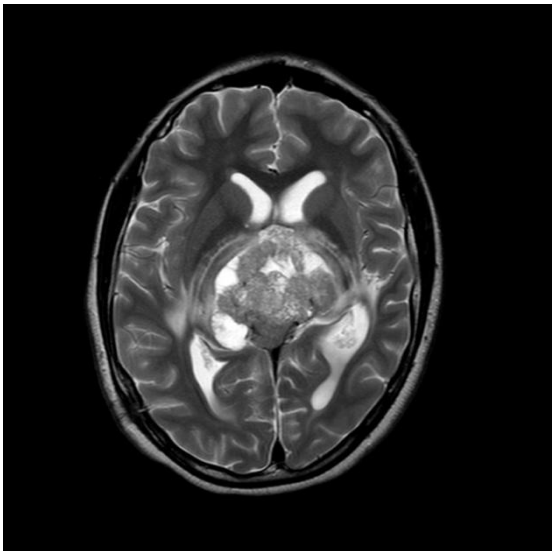

**Fig1 B)**

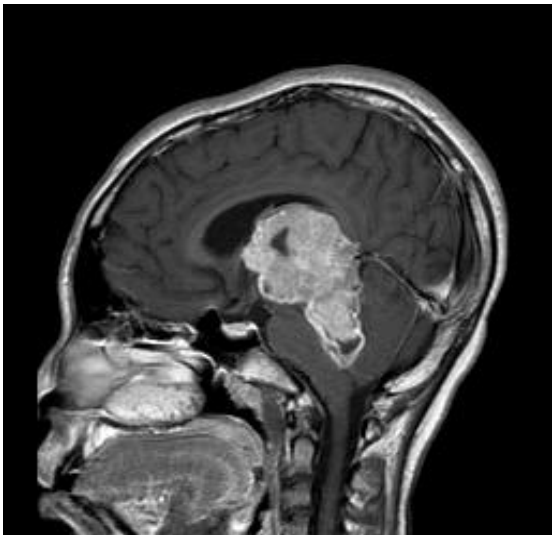

**Fig 1) E**

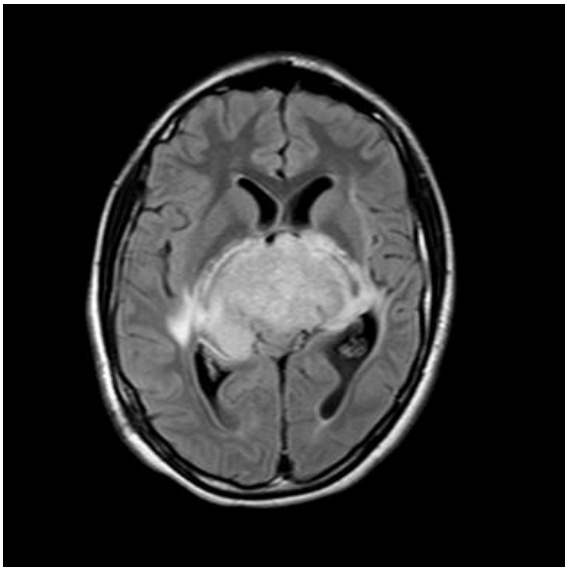

**Fig1) C**

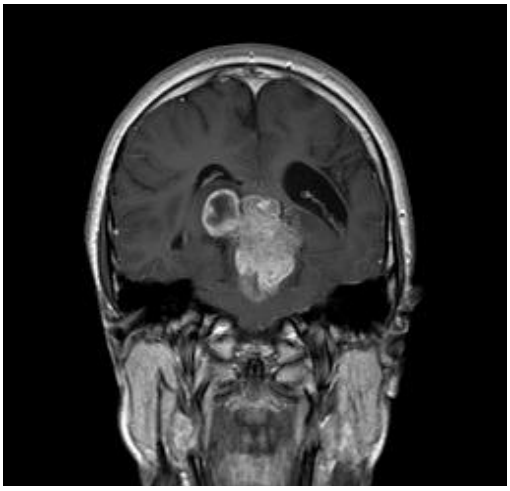

**Fig1) F**

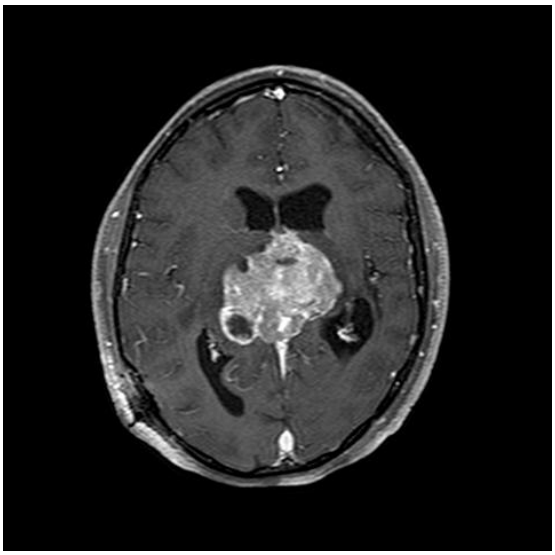

**Fig1 ) D**

**Fig (A-F):** MRI brain images in axial, sagittal and coronal planes with contrast show a large heterogeneously enhancing intraventricular tumor which is isointense on T1W, mildly hyperintense on T2W and FLAIR sequences. The mass extends into fourth ventricle. Perilesional edema and cystic areas noted.

## DISCUSSION

Intraventricular tumors usually present with clinical features of raised intracranial tension or with focal neurological deficits or ataxia. The common intraventricular tumors include ependymoma, subependymoma, central neurocytoma, subependymal gaint cell tumor (SGCT),choroid plexus neoplasms, meningioma, choroid glioma, rosette-forming glioneuronal tumor(RGNT), and metastases etc.[4] Ependymoma: common in young patients, involves fourth ventricle frequently.

These are large intraventricular tumors with areas of cysts and calcification. On MR these lesions are hypointense on T1W and hyperintense on T2Weighted images and show heterogenous enhancement with contrast. Blooming and diffusion restriction may be seen in some cases. Subependymoma: Arise mostly from fourth ventricle and lateral ventricle. They are usually less than 2 cms and majority are seen in patients above 15 years.

These lesions are well defined hypoattenuating on CT with occasional cystic degeneration, calcification and haemorrhage may be seen. On MR they are hypointense on T1 Weighted and hyperintense on T2 Weighted images with minimal or no enhancement with contrast adminstration.

**Cystic neurocytoma:** Occur in lateral ventricles with extension into third ventricle.

They are usually seen in third decade. These are well circumscribed, lobulated masses with cystic areas. Calcification and haemorrhage may be seen in some cases. On CT they are hyperattenuating and on MR they are isointense to gray matter on T1weighted and hyperintense on T2weighted images with strong enhancement with contrast. MR spectroscopy shows the presence of glycine (3.55ppm) which differentiates it from other intraventricular tumors. Subependymal gaint cell tumor: seen in patients with tuberous sclerosis. They are typically well defined masses seen at foramen of Monro. These tumors may show calcification and haemorrhage. On CT they are hypoattenuating and on MR they are hypointense on T1weighted and hyperintense on T2 weighted images with profuse enhancement with contrast.

**Meningioma:** Arise most commonly in the atrium of lateral ventricle, and rarely in third and fourth ventricle. 17% of pediatric meningiomas are intraventricular. These lesions are well defined hyperattenuating globular masses on CT and hypointense on T1weighted and hyperintense on T2 weighted images with show profuse contrast enhancement. Calcifications and cystic areas are seen in some cases.

Choroid glioma: is a slow growing anterior third ventricular mass with clinical features of hypothalamic dysfunction. They are well defined CT hyper attenuating masses which are isointense to gray matter on T1weighted and hyperintense on T2weighted images with profuse enhancement with contrast. Perilesional edema and cystic changes may be seen in some.

**Rosette –forming glioneuronal tumor**: slow growing tumors in young adults, seen in fourth ventricle. They are well defined masses with heterogenous areas of solid and cystic components. Other rare causes of intarventricualr tumors include lymphoma, metastases and primitive neuroectodermal tumors etc.

The most common site of involvement of choroid plexus neoplasms is the atrium of lateral ventricle followed by fourth ventricle. Histologically they are graded into three types: choroid plexus papilloma CPP (WHOgrade1), atypical choroid plexus papilloma CPP (WHO grade 2) and choroid plexus carcinoma CPC (WHO grade3). Of the three grades CPC occurs exclusively in young children. Histological diagnosis of CPC is confirmed only if any four of the following findings are seen. They include a) increased cellular density b) nuclear pleomorphism c) more than 5 mitoses per high power field d) blurring of the papillary pattern and e) necrosis.[5]. Since imaging of brain alone cannot differentiate choroid plexuspapilloma from choroid plexus carcinoma, imaging of entire neuroaxis has to be done[6,7]. Adjacent parenchymal invasion and peritumoral edema are well noted in all three grades of choroid plexus tumors. MR spectroscopy shows elevated choline with normal NAA and creatine peaks. Choroid plexus carcinoma may show elevation of lactate levels[8]. Sushila Jaiswal et al.[9] and others in their review of choroid plexus tumors found only two cases of carcinomas out of 23 cases operated. Chi-Man Yip[10] and others reported a case of CPC in a 21 year old patient which is extremely rare since more than 80 % of CPC occur in children.

Since local recurrence and metastases are more common with CPC gross total resection (GTR) with chemotherapy and radiotherapy is very important in preventing recurrence and prolonging survival[11].

## CONCLUSION

Choroid plexus carcinoma and meningioma constitute the most important intraventricular tumors in pediatric age group. Complete excision has to be contemplated to prevent recurrence and metastases.

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