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Spectrum Of Choroid Plexus Tumors - 5 Year Study In A Tertiary Care Centre

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Abstract

Introduction - Choroid plexus tumors (CPTs) are papillary neoplasms of neuroectodermal origin arising from choroid plexus epithelium of cerebral ventricles[1].
This study includes the spectrum of choroid plexus tumors occurring in foetus, children and adults with reference to their location and clinicopathological features in last 5 years.

Materials And Methods - This is a tertiary care hospital based study of ten cases of Choroid plexus tumors. All specimens were fixed in 10% formalin and entire biopsy tissue was processed as per standard guidelines. IHC markers were done after H&E microscopy.

Results - A total of 10 cases of choroid plexus tumor were diagnosed in 5 years (2018-2022) in our tertiary hospital. Two out of ten Choroid plexus tumors were atypical Choroid plexus papillomas and two were choroid plexus carcinomas and rest 6 cases were Choroid plexus papillomas.

Conclusion – Choroid plexus neoplasms are papillary tumors which can mimic other papillary tumors posing a difficulty in diagnosis especially in small biopsies. Immunomarker studies are helpful in diagnosis and confirmation of histogenesis in these tumors. MIB 1 labelling index is used for grading the choroid plexus tumors according to the WHO classification.

Keywords: Atypical choroid plexus papilloma, Choroid plexus papilloma, Choroid plexus carcinoma, Immunomarkers, MIB 1 labelling index

Introduction

Choroid plexus tumors are rare intraventricular papillary neoplasms derived from choroid plexus epithelium[4]. Choroid plexus tumors constitute 0.3 - 0.8 % of all the brain tumors.[5]

Aproximately 80% of CPCs arise in children, constituting 20 to 40% of all choroid plexus tumors in this age group[4]. Choroid plexus papillomas account for 58.2 % of choroid plexus tumors.[5]

Approximately 10% of all brain tumors in infants and 5% of perinatal brain tumors are of choroid plexus etiology.[1,3]

Their incidence is 10 - 20 % in first year of life, 2- 4 % in children aged < 15 years and 0.5 % in adults.[5]

Amongst the choroid plexus tumors, choroid plexus papillomas represent the lowest grade (WHO grade I). Higher grade tumors in this category include atypical choroid plexus tumor (WHO grade II) and choroid plexus carcinoma (WHO grade III)[3].

Choroid plexus papillomas are more common than choroid plexus carcinomas in a ratio of 5:1.[4]

Material And Methods

Study Design

This is a retrospective tertiary hospital based study of ten cases of Choroid plexus tumors in a period of 5 years from 2018 to 2022. All specimens were fixed in 10 percent formalin and entire biopsy tissue was processed as per standard guidelines. IHC markers were done after H&E microscopy.

Sample Size

10 histopathologically diagnosed Choroid plexus tumor cases.

Study Participants

Inclusion Criteria

Histopathological / IHC confirmed cases of Choroid plexus tumors.

Exclusion Criteria

Cases not confirmed as choroid plexus tumor on histopathology.

Results

In these 10 cases, one was foetus with gestational age of 18 weeks, one was infant of 3 months, & others were cases from 8 years to 60 years of age. Tumors were located in lateral ventricles in 5 cases (2 cases are in the temporal horn), Fourth ventricle in 2 cases, Posterior fossa in 1 case and Fronto temporoparietal SOL - Lateral ventricle in 2 cases. Majority were diagnosed as choroid plexus papillomas, two cases were of atypical choroid plexus tumor and two of choroid plexus carcinoma.

In our study, all tumors were positive for S100 and EMA , 90 % of the tumors were positive for PAN-CK, CK 7 was positive in 60 % while CK20 was positive only in 10 % cases. p53 was positive only in 20 % cases. INI1 was retained in all. MIB1 LI was used for grading the tumors according to WHO 2016 classification.

Features	Choroid Plexus Papilloma (WHO grade I)	Atypical Choroid Plexus Papilloma (WHO grade II)	Choroid Plexus Carcinoma (WHO grade III)
Histopathology	Fibrovascular stalk lined by a single layer of cuboidal epithelium arranged in papillary pattern.	Increased cellularity,nuclear pleomorphism,blurring of papillae, increased mitotic activity.	At least 4 of the following: increased cellularity, blurring of papillary architecture with poorly formed sheets,nuclear pleomorphism,necrosis, brisk mitosis.
Mitotic activity	< 2 per 10 HPFs	≥ 2 per 10 HPFs	> 5 per 10 HPFs
MIB1 Labelling Index	< 5% (Mostly<1 %)	9.1 % (Median)	20.3 % (Median)

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Table 1 : Correlation of age, sex & location of Choroid plexus tumors with Histopathological diagnosis

S.No	Age	Sex	Location	Histopathological Diagnosis	IHC Diagnosis
1	60 years	Female	Recurrent Posterior fossa SOL	Choroid Plexus papilloma (WHO grade I)	Atypical choroid plexus papilloma (WHO grade II)
2	16 years	Male	Left temporal horn papilloma	Choroid Plexus papilloma (WHO grade I)	Choroid plexus papilloma (WHO grade I)
3	18 weeks Foetus	Male	Foetal brain/ Right lateral ventricle	Choroid Plexus papilloma (WHO grade I)	Choroid plexus papilloma (WHO grade I)
4	21 years	Female	Fourth ventricle SOL	Atypical Choroid Plexus papilloma (WHO grade II)	Choroid plexus carcinoma (WHO grade III)
5	3 month	Male	Right temporal horn	Choroid Plexus papilloma (WHO grade I)	Choroid plexus papilloma (WHO grade I)
6	31 years	Male	Right lateral ventricular SOL	Choroid Plexus papilloma (WHO grade I)	Choroid plexus papilloma (WHO grade I)
7	55 years	Male	Left ventricular SOL	Choroid Plexus papilloma (WHO grade I)	Choroid plexus papilloma (WHO grade I)
8	8 years	Male	Recurrent Left Fronto temporoparietal SOL - lateral ventricle	Atypical Choroid Plexus papilloma (WHO grade II)	Atypical choroid plexus papilloma (WHO grade II)
9	8 years	Male	Recurrent Left Fronto temporoparietal SOL - lateral ventricle	Atypical Choroid Plexus papilloma (WHO grade II)	Choroid plexus carcinoma (WHO grade III)
10	35 years	Female	Fourth ventricular SOL	Choroid Plexus papilloma (WHO grade I)	Choroid plexus papilloma (WHO grade I)

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Table 2 : Immunohistochemical evaluation of choroid plexus tumors and their grading

Age	Sex	Immunohistochemistry							
		PanCK	CK 7	CK 20	EMA	S100	INI	P53	MIB 1-LI
60 years	Female	Positive	Negative	Negative	Positive	Positive	Retained	Negative	8-10 %
16 years	Male	Diffuse strong positive	Positive	Negative	Diffuse positive	Diffuse strong positive	Retained	Negative	1-2 %
18 week Foetus	Male	Positive	Negative	Negative	Positive	Positive	Retained	Negative	1-2 %
21 years	Female	Diffuse strong positive	Focal strong positive	Negative	Variable positive	Diffuse strong positive	Retained	Negative	15-20%
3 month	Male	Diffuse focal positive	Strong positive	Negative	Variable positive	Diffuse strong positive	Retained	Negative	Absent/ low
31 years	Male	Positive	Positive	Negative	Positive	Positive	Retained	Negative	1-2 %
55 years	Male	Diffuse strong positive	Strong positive	Negative	Diffuse strong positive	Diffuse strong positive	Retained	Negative	0-1%
8 years	Male	Diffuse strong positive	Focal Positive	Focal Positive	Focal Positive	Diffuse strong positive	Retained	Occasional Positive	8-12%
8 years	Male	Diffuse strong positive	Negative	Negative	Variable positive	Diffuse strong positive	Retained	Variable positive	16-22%
35 years	Female	Positive	Negative	Negative	Positive	Positive	Retained	Negative	0-1%

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FIGURES

Figure 1: Specimen of foetus of GA 18 wks , 6 days, which was kept in 40% formalin and it was also injected 1.5 ml in anterior and posterior fontanelle for better fixation of fetal brain

Figure 2 : Ultrasonography image of foetus - Routine Anomaly scan at GA of 18 weeks revealed a solid cystic mass of 2x0.95 cm in the right lateral ventricle with hyperechoic brain parenchyma obscuring details of underlying cerebral cortex

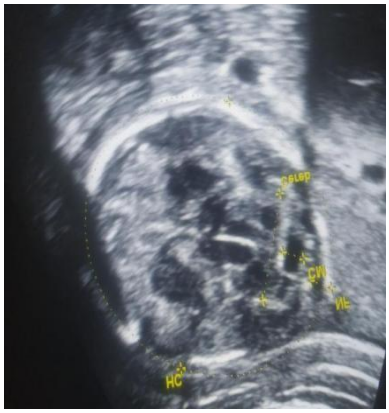
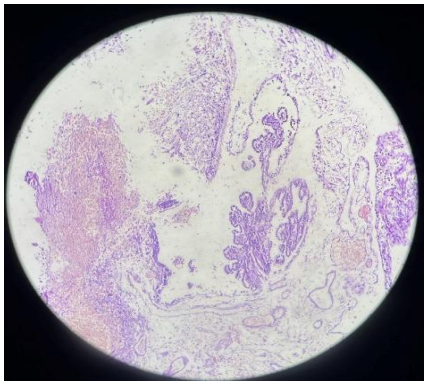


Figure 3: H and E image of foetus (Choroid plexus papilloma – WHO grade I)



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Choroid plexus papilloma (WHO grade I)

Figure 4 : H & E

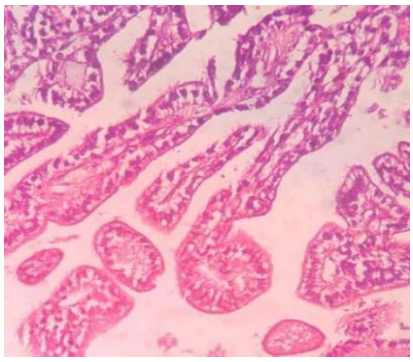
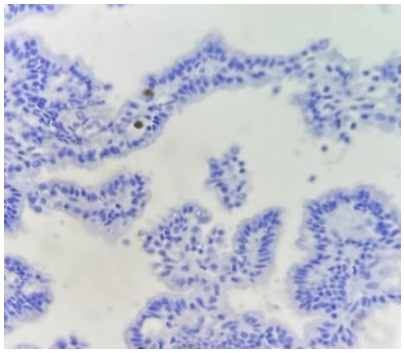


Figure 5 : MIB 1- LI = 1-2 %



Atypical Choroid plexus papilloma (WHO grade II)

Figure 6 : H & E

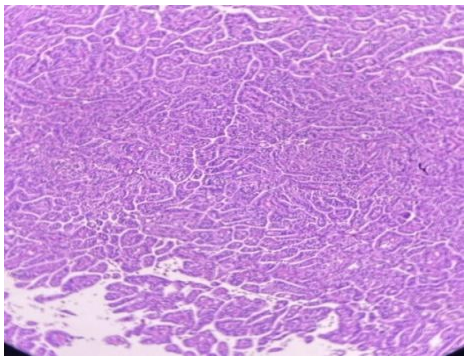
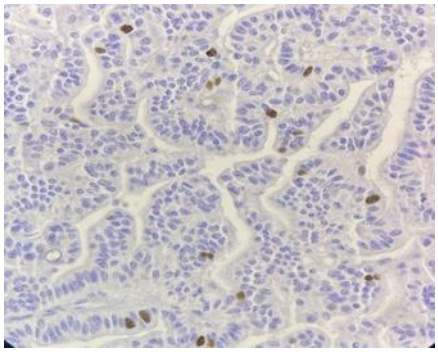


Figure 7: MIB 1- LI = 8-10 %



Choroid plexus carcinoma (WHO grade III)

Figure 8 : H & E - Necrosis seen

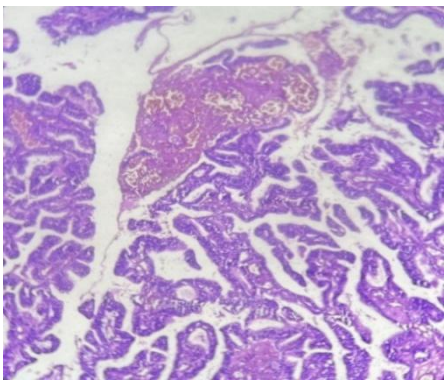


Figure 9 : MIB 1- LI = 16-22 %

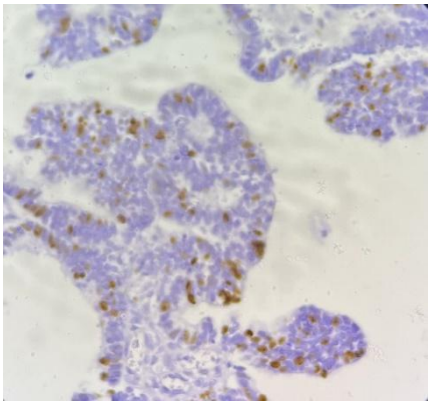


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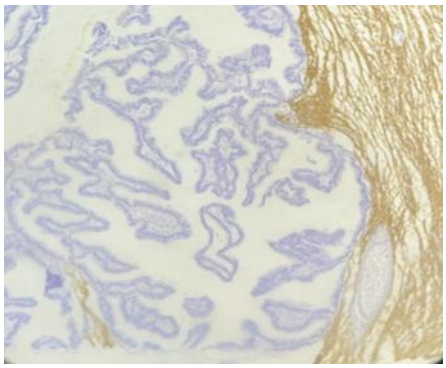
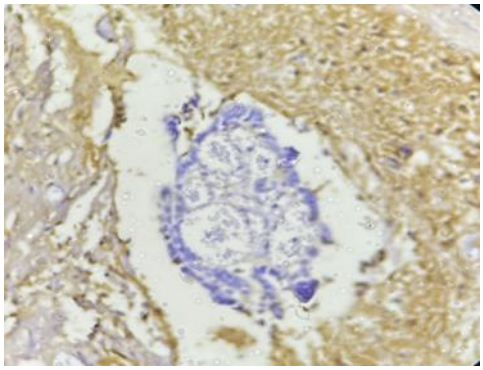
Figure 10 & 11 : GFAP – Invasion seen.





Discussion

During a period of five years, 10 cases of choroid plexus tumors were studied in our institute. They were 6 males (one foetus), 4 females. Male to female ratio is 2.3:1. Common presenting feature was raised intracranial pressure.

In these cases, one was foetus with gestational age of 18 weeks, one was infant of 3 months, & others were cases from 8 years to 60 years of age, covering all the age groups.

Tumors were located in lateral ventricles in 5 cases (2 cases are in the temporal horn), Fourth ventricle in 2 cases, Posterior fossa in 1 case and Fronto temporoparietal SOL - Lateral ventricle in 2 cases. Out of these, 3 cases of choroid plexus tumor came with recurrence of tumor. Two cases were identified as atypical choroid plexus tumor and two as choroid plexus carcinoma.

CPCs most commonly occur in the lateral ventricle followed by fourth ventricle & third ventricle. In our study one case was located in lateral ventricle & the other was in fourth ventricle.

One case was foetal brain, Baby of a 21 years old gravida 1, para 0, LB 0, female with no significant history of systemic illness during pregnancy. No history of teratogenic drugs or radiation exposure. Weight was increasing as expected. No anemia and edema present and vitals were stable during the whole pregnancy. NT scan was normal at 12 week of gestational age. Routine Anomaly scan was done in which a solid cystic mass of 2x0.95 cm was found in the right lateral ventricle with hyperechoic brain parenchyma obscuring details of underlying cerebral cortex. It was diagnosed as choroid plexus papilloma (WHO grade I).

Two cases were diagnosed as atypical choroid plexus tumors. Both of them presented as recurrent papillomas . The first case,60 yrs/F, presented with a recurrent SOL in the posterior fossa & was diagnosed as atypical choroid plexus papilloma (WHO grade II) with MIB 1 LI of 8 to 10 %.

Other case was a 8 yrs/M, presented with recurrent SOL in left fronto temporoparietal region & was diagnosed as atypical choroid plexus papilloma (WHO grade II), with MIB 1 LI of 8 to 12 % .

The second case of atypical choroid plexus papilloma again presented after about 7 months, in this biopsy it was diagnosed as choroid plexus carcinoma (WHO grade III),with MIB1 LI of 16 to 22 %. Brain invasion was also seen in this case. Patient was advised furthur evaluation and work up for Li-Fraumeni syndrome.

The other case was 21 yrs/ F,presented as fourth ventricle SOL and was diagnosed as choroid plexus carcinoma (WHO grade III) with MIB LI of 15 to 20 %.

In our study, all tumors were positive for S100 and EMA , 90 % of the tumors were positive for PAN-CK, CK 7 was positive in 60 % while CK20 was positive only in 10 % cases.

p53 was positive only in 20 % cases. INI1 was retained in all. MIB1 LI was used for grading the tumors according to WHO 2016 classification.

Distinguishing choroid plexus tumors from other CNS papillary tumors and metastatic papillary carcinomas can pose a diagnostic challenge . Cytokeratin and other immunomarker studies can help in the diagnosis and differentiating them from papillary variant of ependymoma, papillary

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meningioma, metastatic papillary carcinoma of thyroid, breast carcinoma and lung carcinoma.

Atypical teratoid/rhabdoid tumors (ATRT) are differentiated from choroid plexus tumors by INI1 which is retained in choroid plexus tumors and lost in ATRT.

In our study, INI 1 was retained in all (100%) the tumors.

P53 has been found to be positive mostly in choroid plexus carcinomas, similar to our study in which both cases (20%) of CPCs were positive for P53.

Conclusion

Choroid plexus papillomas are uncommon, benign neuroepithelial intraventricular tumors that can occur in both pediatric (more common) and adult population.

Choroid plexus neoplasms are papillary tumors which can mimic other papillary tumors posing a difficulty in diagnosis. These tumors include a papillary variant of ependymoma, papillary meningioma, metastatic papillary carcinoma of thyroid, breast carcinoma and lung carcinoma. Atypical teratoid /Rhabdoid tumor (ATRT) is a significant differential diagnosis.

Immunomarker studies are helpful in diagnosis and confirmation of histogenesis in these tumors. Immunomarker studies by MIB 1 labelling index is used for grading the choroid plexus tumors according to the WHO classification.

The majority of CPT's are diagnosed during the first year of life and are often sporadically detected. In some cases, CPTs are found in association with cancer predisposition syndromes including Aicardi and Li-Fraumeni syndromes. The Li-Fraumeni syndrome is associated with TP53 chromosomal anomaly. Aicardi syndrome is an X linked disorder whose manifestations are generally observed in utero and at birth[1]

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