

INTRAPARENCHYMAL CHOROID PLEXUS PAPILLOMA- A CASE REPORT AND LITERATURE REVIEW

Jini Lonappan Valooran¹, Anand T. P², Noushad A³, Joy Augustine⁴

¹Assistant Professor, Department of Pathology, MCH, Thrissur.

²Assistant Professor, Department of Forensic Medicine, MCH, Thrissur.

³Senior Resident, Department of Pathology, MCH, Thrissur.

⁴Professor, Department of Pathology, MCH, Thrissur.

ABSTRACT

BACKGROUND

Choroid plexus papilloma (CPP) is a rare, slow growing benign tumour of childhood with neuroectodermal origin. Their usual location is ventricle and associated with signs and symptoms of increased intracranial pressure and obstructive hydrocephalus. Rarely CPP can be intraparenchymal. Sudden deaths have been reported, but are very unusual. Here, we present a case of a 24-year-old male brought for medicolegal autopsy examination on grounds of sudden unexplained death. An autopsy examination showed oedema of brain and an irregular, firm, nodular mass deep inside the white matter of left occipital lobe. Histopathology and Immunohistochemistry (IHC) confirmed the diagnosis of a choroid plexus papilloma.

KEYWORDS

Choroid Plexus Papilloma (CPP), Intraparenchymal, Adult, Sudden Death.

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BACKGROUND

Choroid plexus papillomas are rare, accounting for less than 1% of all intracranial tumours in adults. They are relatively more common in childhood and constitute 1.5 to 4% of intracranial tumours.^[1] They are neuroectodermal in origin, usually located in the ventricle and is composed of multiple papillary fronds mounted on a well-vascularised connective tissue stroma very similar in structure to the normal choroid plexus. Grossly, they may appear as reddish cauliflower like growths, which often become heavily calcified.^[2] CPPs are not malignant; however, malignant evolution may occur, with an incidence of 10-30%. In a recent series by McEvoy et al, the five-year survival rate was 100%.^[3] Exceptional cases of unusual locations like intraparenchymal, suprasellar region and spinal epidural are on record. To the best of our knowledge, only three cases of intraparenchymal CPP have been reported. Here, we describe a rare case of intraparenchymal CPP. Our case has an unusual location and unique histopathological features.

Case Report

A 24-year-old moderately built and nourished male was brought for medico-legal autopsy examination on ground of sudden death. He was known to complain of headache quite often. Autopsy examination showed brain oedema with an irregular firm nodular swelling measuring 8 x 6 x 3 cm deep inside the white matter of left occipital lobe near the gangliocapsular region with surrounding area filled with clear fluid (Figure 1). Ventricles were free with smooth inner wall. Other organs were within normal limits.

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Corresponding Author:

Dr. Jini Lonappan Valooran,

#VII/2A, 228B,

Poovani, Kolazhy,

Thrissur-680010.

E-mail: jinivalooran77@gmail.com

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Gross Findings

Irregular, firm, nodular grey white growth measuring 8 x 6 x 3 cm with a grey white granular cut section. (Figure 2).

Microscopy

Section showed a neoplasm composed of cells arranged in papillary pattern and in sheets (Figure 3 and 4). Individual cells had round to oval nucleus and moderate eosinophilic cytoplasm. Mitotic figures were less than one per high power field. Focal areas of necrosis and calcification were noted. Focal blurring of papillary pattern, with sheet like arrangement and areas of necrosis suggested a tumour is high grade. The differential diagnosis included metastatic papillary neoplasm, papillary meningioma, atypical choroid plexus papilloma and papillary ependymoma. Immunohistochemistry (IHC) was done. Markers used were cytokeratin, Vimentin, S100, GFAP, Thyroglobulin, EMA and Ki67. Cytokeratin, Vimentin and S100 showed strong positivity (Figure 5 to 9). Thyroglobulin EMA and GFAP were negative. Ki67 showed very low index. Immunohistochemistry confirmed our diagnosis of Choroid plexus papilloma.

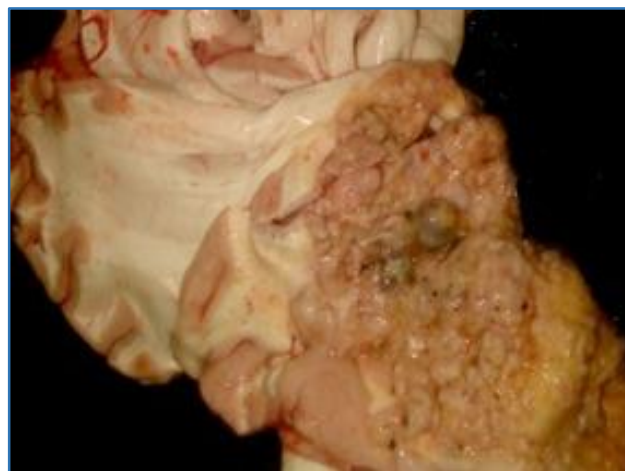


Figure 1. Gross Appearance of the Choroid Plexus Papilloma

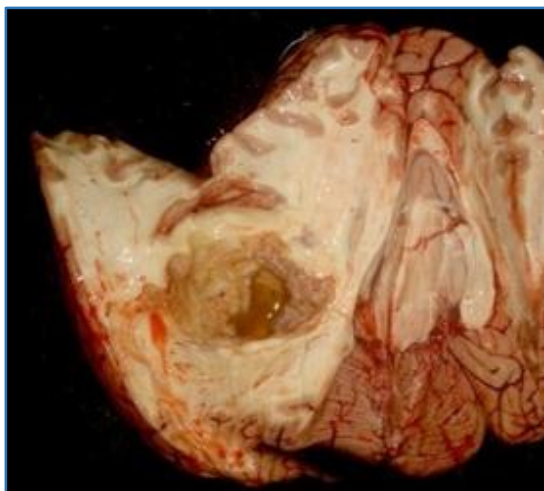


Figure 2. Gross Appearance of the Choroid Plexus Papilloma

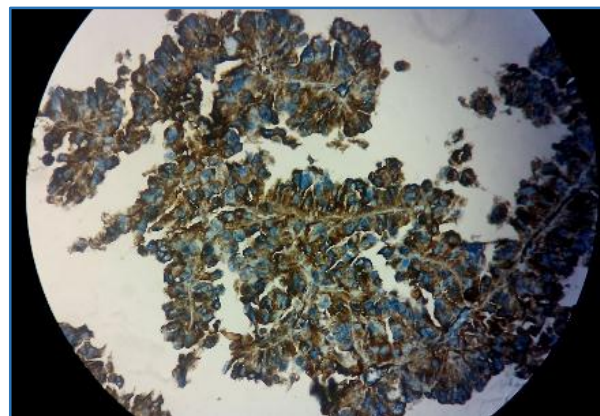


Figure 6. CPP with Expression of Vimentin

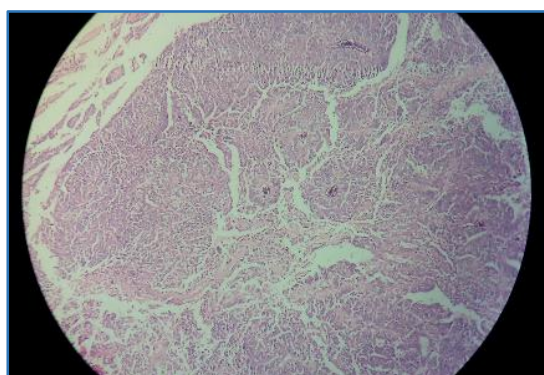


Figure 3. Scanner view of the Choroid Plexus Papilloma showing Papillary Areas (H & E Low power view)

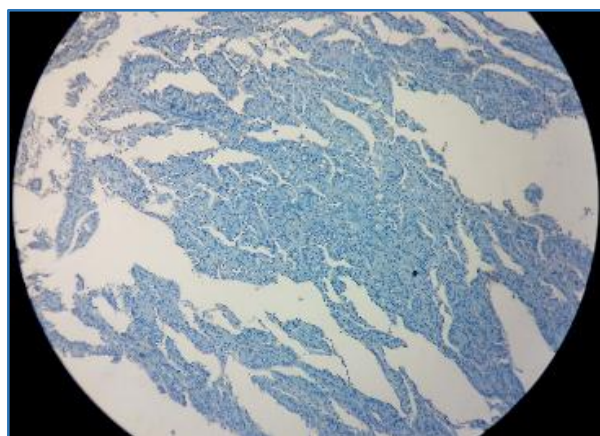


Figure 7. EMA done which is Negative

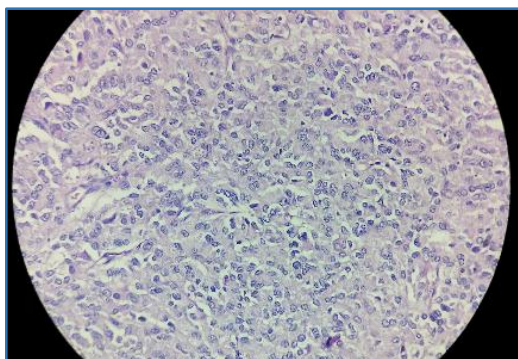


Figure 4. Microscopic Appearance of the Choroid Plexus Papilloma-(H&E High power view)

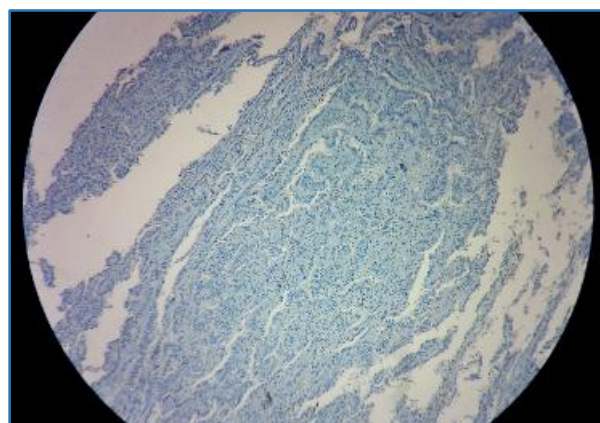


Figure 8. GFAP done which is Negative

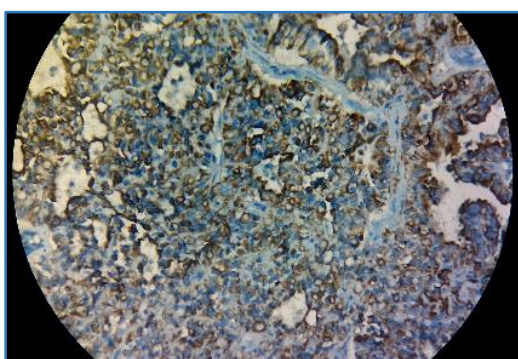


Figure 5. CPP with Expression of Cytokeratin

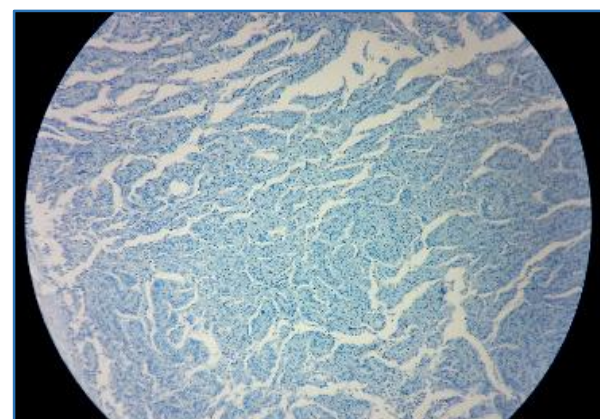


Figure 9. Thyroglobulin done- Negative

DISCUSSION

Choroid plexus tumours are rare. Among these, papillomas are far more common than carcinomas. They usually arise from the ventricles. In adults the origin is far more common in lateral ventricle. About 20- 40% of all choroid plexus tumours in children are carcinomas.^[4] Carcinomas are rare in adults. CP angle tumours in the fourth ventricle has a tendency to grow through the Foramen of Luschka into the CP angle.^[5] The usual duration of symptoms is reported to be about 1 month.^[3] There has been case reports of sudden death where tumour involved the third ventricle and caused acute ventricular obstruction.^[6] Grossly choroid plexus tumours are generally well-circumscribed brownish red cauliflower like masses. Malignant ones are invasive appearing haemorrhagic or necrotic. Histologically, there is a delicate connective tissue frond covered by a single layer of cuboid epithelium. The papilloma resembles normal choroid plexus but the cells are more crowded and elongated. The carcinoma is more cellular and shows features of anaplasia.

Rarely these tumours can undergo mucinous degeneration, melanisation, tubular glandular architecture or osseous and cartilaginous metaplasia.^[7] In adults, most choroid plexus papillomas are heterogenous, secondary to cystic degeneration and or calcification. Treatment depends on a number of factors including the size, location, type and grade of the tumour as well as the age and health of the patient. Surgery may be the only treatment required if the tumour can be completely removed.

Our case is unique due to intraparenchymal location, and its variation in histology including blurring of papillary pattern with sheet like arrangement of cells, pleomorphism of cells, high cellularity and areas of necrosis which made diagnosis difficult. To the best of our knowledge, only 3 cases of intraparenchymal choroid plexus papillomas have been reported earlier. Among the reported cases one was in temporal lobe with hydrocephalus,^(8,9) another one was in right parietal mass resulting in intractable seizure reported by Masaaki et al in 2011,^[10] and 3rd case was in medial aspect of the left temporal lobe.^[11] Our case is probably the first CPP to be reported in the occipital lobe.

CONCLUSION

Intraparenchymal CPPs are very rare. Differing from the usual bland pattern in CPP, blurring of papillary pattern with

sheet like arrangement, high cellularity, nuclear pleomorphism and areas of necrosis though rare do occur.

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