Case Report

The Smallest Choroid Plexus Papilloma at the Youngest Age of Presentation Ever Reported

Femela M*, Jane BI**, Vijayashree R***

*Professor, **Post Graduate student, ***Professor, Department of Pathology, Chettinad Hospital and Research Institute, Chettinad Academy of Research and Education.

Dr. Femela Muniraj is working as Professor in the Department of Pathology and Medical Director in the Department of Transfusion Medicine, CHRI. She did her graduation in Madurai Medical College and Post-Graduation in JIPMER. She has received the 'Best Postgraduate Award' from JIPMER University. She has published nearly 15 articles in international and national indexed journals so far. She has presented papers and posters in national conferences. She is a member of Tamil Nadu & Pondicherry chapter of Indian Association of Pathologists and Microbiologists, AABB and CAP. She is a reviewer in five of Pathology journals. Her areas of interest are Surgical Pathology, Hematopathology, Transfusion Medicine, Research and Quizzing.

Corresponding author - Dr. Jane Betsy Isaac (janebetsy@gmail.com)

Chettinad Health City Medical Journal 2020; 9(3): 205 - 207

DOI: https://doi.org/10.36503/chcmj9(3)-11

Abstract

Choroid plexus papilloma are tumours of the central nervous system occurring in children and usually presents in the first few years of life. Rarely, it has been diagnosed at birth or prenatally. Here, we present a case of choroid plexus papilloma in a foetus of gestational age 20 weeks.

Keywords: Childhood tumour, Choroid plexus papilloma, Intracranial Tumour, Youngest age of presentation

Introduction

Choroid plexus tumours represent about 2-4% of tumours of the central nervous system occurring in children.[1] Rare cases have been diagnosed at birth and prenatally.[1-4] Choroid plexus tumours comprise Choroid Plexus Papilloma (CPP) (WHO grade I), Atypical Choroid plexus papilloma (WHO grade II) and Choroid plexus carcinoma (WHO grade III).[2,3,5,6] CPP has better prognosis than other choroid plexus tumours.[5]

Case Report

A foetus was expelled from a 26-year-oldmother, due to intrauterine death, at 20 weeks of gestation and autopsy was done. Ultrasonogram done in second trimester did not detect any anomalies, except that the baby was small for gestational age. Her previous baby was delivered at 34 weeks with cardiac anomaly and died 2 days after birth.

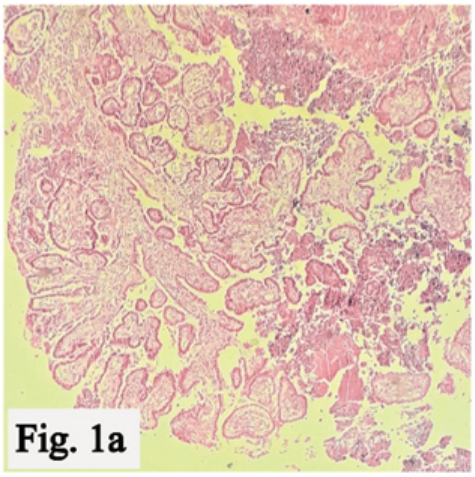
On gross examination, the scalp and neck were edematous, otherwise unremarkable. Microscopic examination of all the organs, umbilical cord, placenta, except brain, was unremarkable. The development of all the organs were appropriate for the age.

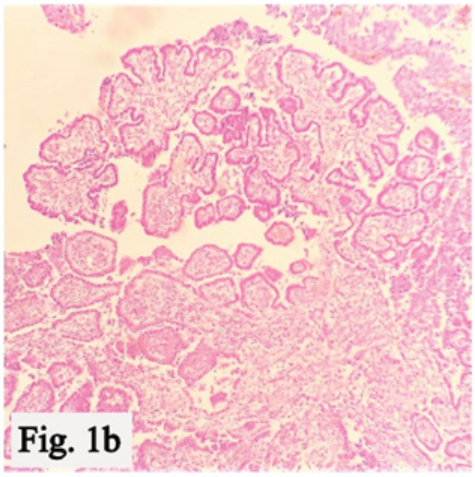
A section from the fourth ventricle had a tumour showing papillary architecture, a fibrovascular core lined by single layered cuboidal to columnar epithelium (Figure 1a, 1b, 2) and adjacent normal brain parenchyma (Figure 3). The tumour measured 1.47 mm. No necrosis or atypia of epithelium or mitotic figures were noted. The tumour was not present in the subsequent sections and hence immunohistochemistry with CK7, EMA, GFAP couldnot be done.

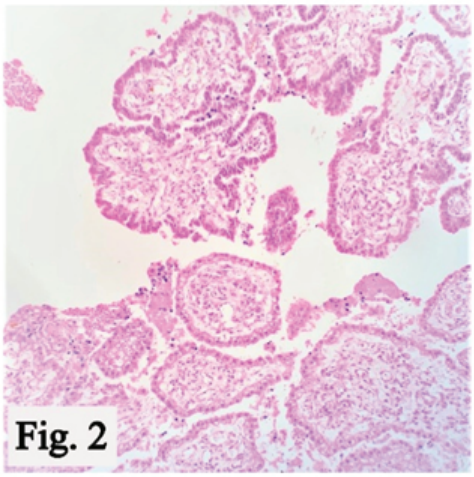
Discussion

Choroid Plexus Papilloma (CPP) are rare and constitute from 0.4 to 1.0% of intracranialtumours in adults and 1.5 to 6.0% in children.[2,6] About 70% of the cases have been reported in children of age less than 2 years.[6] No predilection has been observed in either sex.[3] In children they are usually located supratentorial and in adults they are usually infratentorial.[6] In children, they are found more commonly in lateral ventricles (43-80%), less commonly in fourth(10-40%)and third(5-10%) ventricles, cerebellopontine angle (9%) and, rarely in suprasellar region and cerebral convexity.[1-5] CPP has been reported to be associated with Aicardi and Li-Fraumeni syndromes, giant melanocytic nevus.[5]

Clinically, CPP presents with hydrocephalus, macrocephaly, 'sunset' sign in the eyes, signs of meningeal irritation, delay in developmental milestones, focal neurological symptoms such as headache, abnormal gait, epilepsy, vomiting.[2,5-7] They are tumours derived from the neuroepithelial cellsthat line the







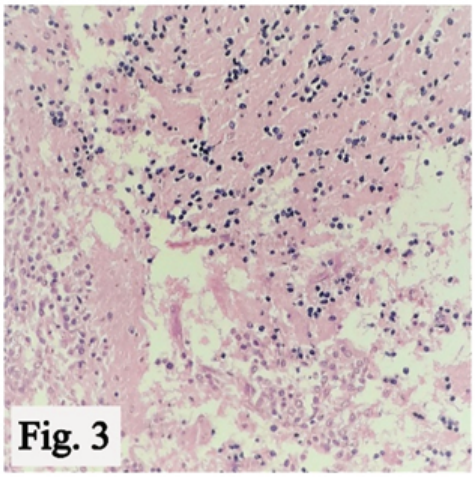


Figure 1a, 1b: Tumor with papillae (H & E, x100)
Figure 2: Papillae lined by cuboidal to columnar epithelium (H & E, x200)
Figure 3: Adjacent cerebellar tissue with normal histology (H & E, x400)

choroid plexuses in the ventricles.[2,5] There is a male preponderance and usually present by 3 years of age, 20-25% occurring in infants, 86% less than the age of 5 years.[2,4] Statistically, they constitute 3% of paediatric intracranial primary tumours and 0.6% of brain tumours in adults.[3-5] Most of the choroid plexus tumours are detected by ultrasonography.[5] In foetus, CPP is most often detected at the end of the third trimester of pregnancy, by ultrasono-graphy.[3-5]

Radiologically, the tumour appears lobulated and the contour is irregular, thus resembling "cauliflower".[2] Computed tomography shows homogenous, iso- or high-density tumour, sometimes associated with calcification, cystic change or hemorrhage.[2] Foetal MRI following an ultrasonographic finding of foetal hydrocephalus has been performed rarely.[4] In the MRI, CPP appears as contrast enhanced lesion; in the T1 weighted sequence, it appears asisodense or hypointense signal; and in the T2 weighted sequence, it appears as isodense or hyperintense signal.[2,6]

In the literature, the age of presentation of CPP ranges from 21 weeks of gestation to 18 months after birth.[4,5] The differential diagnosis of the intraventricular tumours in children include Ependymomas, Subependymal giant cell astrocytoma, Medulloblastoma, low grade astrocytoma, Meningiomas, Villous hypertrophy of choroid plexus.[5,6] The survival rate at 1 year, 5 and 10 years is 90%, 81% and 77% respectively.[2] Surgical resection is the mainstay of treatment; but it may be challenging because of the high vascularity of the tumour and the possibility of intraoperative haemorrhage.[2,5,6] Post-surgical complications may occur and include psychomotor retardation, epilepsy, hyperreflexia, developmental delay.[1]

Grossly, CPP is well demarcated cauliflower like masses that may adhere to the ventricular wall. The size of Choroid plexus papilloma in the cases reported ranges from 1cm to 6.5cm.[5] Microscopically, it is composed of single layer of orderly cuboidal to columnar epithelial cells resting on finger like papillary formations with distinct fibrovascular core. There is no atypia or mitotic figures. Ultrastructurally, these tumour cells are similar to normal choroid plexus epithelial cells.[1-3,5,6] Immunohistochemically, CPP shows positivity with cytokeratin 7, Vimentin,

GFAP, while transthyretin, EMA and S-100 protein are variably positive.[1,3,5,8,9] Proliferation index (ki-67) is low. Specific cytogenetic abnormalities are yet to be identified. DNA sequence of simian virus 40 has been identified in 50% of the cases.[1]

Two variants of choroid plexus papilloma have been described in the literature – Villous hypertrophy and cystic choroid plexus papilloma.[5,7,8] Villous hypertrophy is the diffuse enlargement of the choroid plexuses in bilateral lateral ventricles.[3,5,7] Cystic CPP manifests as a cystic tumour with or without a mural nodule. It contains cysts within the tumour or in the adjacent cortex and is found in 7.5% of patients with choroid plexus papilloma. The cysts are lined by cuboidal to columnar choroid plexus-like cells and contain pale yellow fluid.[3,5,8]

CPP is distinguished from atypical CPP by morphology, low proliferation index. Atypical choroid plexus papilloma, may resemble choroid plexus papilloma grossly, being nodular, with cauliflower-like appearance, but shows flattened grey areas scattered on the surface. Microscopically, it shows shortened and broadened villi, focal necrosis, hypercellular epithelium with crowding, overlapping cells, nuclear pleomorphism, hyperchromasia and few mitoses.[5]

Choroid plexus carcinoma has the nodular, cauliflower-like appearance similar to Choroid plexus papilloma and atypical choroid plexus papilloma but shows areas of necrosis and haemorrhage. Microscopically, it shows infiltration into the adjacent brain, hypercellular epithelium, pleomorphic nuclei, vascular proliferation, high proliferation index. Genetically, it shows hypo- or hyperdiploid copy number, mutations/germline variants in p53.[5]

In Vitro Fertilization is implicated in the pathogenesis of Choroid plexus papilloma, though many studies have contradicted it.[2] This tumour is misdiagnosed sometimes, since it mimics conditions like haemorrhage, infection, hydrocephalus due to congenital defects.[1-8]

Conclusion

This is the smallest Choroid Plexus Papilloma of fourth ventricle in a foetus of 20 weeks gestation, which is the youngest age at which this entity has ever been reported.

References

1. McKeever P E. Brain, spinal cord and meninges. In:Mills S E.Sternberg's diagnostic surgical pathology. 6th ed. Philadelphia: Wolters Kluwer Health. 2015;1260-3.
2. Cao L.R, Chen J, Zhang R.P, Hu X.L, Fang Y.L, Cai C.Q. Choroid Plexus Papilloma of Bilateral Lateral Ventricle in an Infant Conceived by in vitro Fertilization. PediatrNeurosurg. 2018;53:401-406.
3. Verma SK, Satyarthee GD, Sharma BS. Giant choroid plexus papilloma of the lateral ventricle in fetus. J PediatrNeurosci. 2014;9(2):185–7.
4. Noguchi A, Shiokawa Y, Kobayashi K, Saito I, Tsuchiya K, McMenomey SO, et al. Choroid plexus papilloma of the third ventricle in the fetus. Case illustration. J Neurosurg. 2004;100:224
5. Crawford J R, Isaacs H J. Perinatal (fetal and neonatal) choroid plexus tumors: a review. Child's Nervous System. 2019;6(35):937-44.
6. Muñoz Montoya JE, Maldonado Moran MA, Santamaria Rodriguez P, Toro Lopez S, Perez Cataño CS, Luque Suarez JC. Choroid Plexus Papilloma of the Fourth Ventricle: A Pediatric Patient. Asian J Neurosurg. 2019;14(2):585–88.
7. Welch K, Strand R, Bresnan M. Congenital hydrocephalus due to villous hypertrophy of the telencephalic choroid plexuses. Case report. J Neurosurg. 1983; 59(1):172–5.
8. Murata M, Morokuma S, Tsukimori K, Hojo S, Morioka T, Hashiguchi K et al. Rapid growing cystic variant of choroid plexus papilloma in a fetal cerebral hemisphere. Ultrasound ObstetGynecol. 2009;33(1):116–8.