CASE REPORT

Choroid Plexus Carcinoma (WHO Grade III): A Rare Case of Central Nervous System Tumour in Two Years Old Girl

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Abstract

Choroid plexus tumours represent 0.3 to 0.6% of all central nervous system tumors. The pediatric age group has a higher prevalence of malignancies originating from the choroid plexus epithelium. We reported a case of 2yr old female child who presented with projectile vomiting for the past 2 weeks to Neurosurgery. On CT scan brain showed a large heterogeneous density mass with perilesional oedema. Craniotomy and debulking of the tumors through a transylvian fissure were done. Intraoperatively, the tumor was fragile, soft, and highly vascular. On HPE, tumor was Grade III choroid plexus

Keywords

Choroid Plexus Carcinoma, Central Nervous System Tumours, Choroid Plexus Papilloma, Atypical Choroid Plexus.

Introduction

Choroid plexus tumours, which account for between 2 and 4 per cent of cerebral tumours in children and 0.5 per cent in adults, are uncommon intraventricular papillary neoplasms produced from the choroid plexus epithelium. [1] These tumours are classified by World Health Organization as grade one as choroid plexus papilloma, grade two as atypical choroid plexus papilloma and grade three as choroid plexus carcinoma. Ten per cent of Choroid plexus tumours are seen in children younger than a year.[2] Its common location is in the lateral ventricle in children and the fourth ventricle in adults. Choroid plexus carcinoma has a poor prognosis, and individuals with CPCs have an average 40% 5-year survival rate. [3] The importance of the gross complete elimination of CPC as a component of the therapeutic plan was stressed by a meta-analytical investigation. Significantly better overall survival is correlated with the degree of tumour eradication. While the impact of adjuvant radiation or chemotherapy on OS is still debatable.

Case Report

A 2-year-old female was admitted to the neurosurgery department with complaints of headache, nausea, and vomiting for 2 months. Clinical examination and routine haematological investigations did not reveal any significant findings. On Radiological examination, CT scan of the brain showed a large heterogeneous density mass with lobulated mass with an isointense signal relative to white matter and significant periventricular edema in the right

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Manuscript Received: 09.02.2023; **Revision Accepted**: 23.04.2023;

Published Online First: 10 Oct 2023

Open Access at: https://journal.jkscience.org

Cite this article as: Chander SK,Vinothkumar B, Yamini PM, Sulochana S. Choroid Plexus Carcinoma (WHO Grade III): A Rare Case of Central Nervous System Tumour in Two Years Old Girl. JK Science 2023;25(4):258-60.

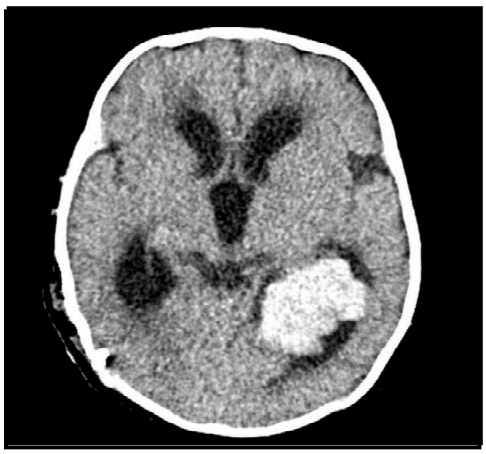

Fig 1. CT scan-large heterogeneous density mass with lobulated mass with an isointense signal relative to white matter and significant periventricular edema in the right lateral ventricle

lateral ventricle. (*Fig 1*). Craniotomy and debulking of the tumor through a transylvian were done. Intraoperatively, the tumor was fragile, soft and highly vascular, with features of high-grade tumor.

On gross examination, multiple fragile grey-brown soft tissue on aggregate measuring 5x 3.5x 2 cm. On microscopic examination, fragments of tissue show a neoplasm composed of cells which are arranged in sheets and in a papillary pattern with a fibrovascular core and congested blood vessels (*Fig 2A*). Areas of infiltration into the surrounding glial tissue and choroid plexus were seen. Areas of necrosis seen (*Fig 2B*). The tumor cells have scant to moderate basophilic cytoplasm and markedly hyperchromatic pleomorphic nuclei with prominent nucleoli and increased mitotic figures (*Fig 2C*). For further confirmation, immunohistochemical markers studies were performed and showed PAN CK-Focal moderate cytoplasm positivity in 30% of tumor cells (*Fig 2D*). Ki67-positive, percentage of cells with nuclear positivity 90% of tumor cells (*Fig 2E*) and Synaptophysin-negative (*Fig 2F*). Therefore, we came to the final diagnosis of Choroid plexus carcinoma (WHO grade 3).

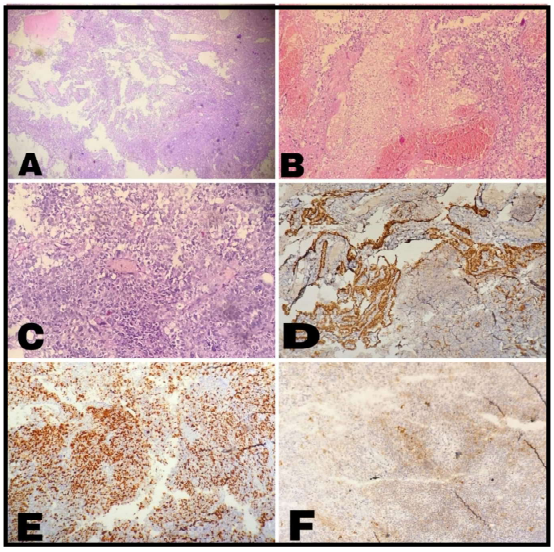

Fig 2. Microscopic examination showing neoplasm composed of cells which are arranged in sheets and in a papillary pattern with a fibrovascular core and congested blood vessels (Fig 2A). Areas of infiltration into the surrounding glial tissue and choroid plexus were seen. Areas of necrosis seen (Fig 2B). Tumor cells have scant to moderate basophilic cytoplasm and markedly hyperchromatic pleomorphic nuclei with prominent nucleoli and increased mitotic figures (Fig 2C). For further confirmation, immunohistochemical markers studies were performed and showed PAN CK-Focal moderate cytoplasm positivity in 30% of tumor cells (Fig 2D). Ki67-positive, percentage of cells with nuclear positivity 90% of tumor cells (Fig 2E) and Synaptophysin-negative (Fig 2F)

Discussion

Histopathological examination of the tumour in our case, diffuse blurring of papillary structures, increased cellularity, multiple areas of necrosis, significant pleomorphism and increased mitotic activity was leading to the diagnosis of Choroid plexus carcinoma. Study with immunohistochemical marks which are also in favour of Choroid plexus carcinoma. Sometimes, although not often enough to be categorised as CPC, choroid plexus papillomas exhibit one or more of the aforementioned malignant features. So, atypical choroid plexus papilloma is recognized as an intermediate entity regarding mitotic activity since 2007 in the WHO classification of central nervous system tumours.[4] Compared with papilloma, the diagnosis of Choroid plexus carcinoma is rendered in the setting of increased cell density, mitotic figures (usually greater than 5 per 10 high power fields), nuclear pleomorphism, and necrosis.[5] The parenchyma of the

adjacent brain is frequently diffusely infused. Although low-grade Choroid plexus papilloma is characterised by distinct papillary configuration, CPC has an ill-defined development pattern that can make identification difficult. The papillary features are blurred or may be lost in sheets of epithelial tumour cells.[6]

Surgical resection of tumours in this area is challenging and highly demands experience. Due to these tumours' high vascularity, significant blood loss after resection needs to be anticipated and successfully managed, especially in the juvenile population. So, neuroendoscopic biopsy followed by chemoradiotherapy is the best treatment option in cases where the tumour is not resectable. The choroid plexus papilloma, villous hypertrophy of the choroid plexus, meningioma, and metastatic papillary neoplasms are among the differential diagnoses. Associated clinical, cytologic, morphologic, and immunohistochemical characteristics are typically used to differentiate tissues.[7] The regimen for the treatment of choroid plexus cancer is not yet developed. The primary objective is the total removal of the tumour, which enhances the prognosis.[8]

Conclusion:

CPC is associated with poor prognosis, the 5-year survival rate of CPC patients is approximately 58% and 20% after complete or partial resection respectively. CPC cannot be differentiated from CPP based on radiological characteristics and this pathology should be in mind in approaching any lesion with suspected choroid plexus origin even in rare locations. Based on the meta-analysis of Wolff et al. adjuvant radiation can improve survival even after gross total resection however, this effect was only statistically significant in older age young patients but not in infants.

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