Case Report-I

Choroid Plexus Carcinoma in a Child : A Case Report

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ABSTRACT

Choroid plexus tumours arising from the epithelium of the ventricles are rare. Clinically these tumours produce hydrocephalus and symptoms of raised intracranial pressure. Histologically, most of these tumours are benign papillomas, while carcinomas are rare. We present a case of choroid plexus carcinoma in a child.

INTRODUCTION

Choroid plexus tumours, commonly located in the lateral ventricles, arise from its lining epithelium. These tumours produce hydrocephalus and raised intracranial pressure. Clinical signs may be subtle and includes behavioral changes and irritability. These tumours reveal a 'frond-like' appearance with intense enhancement after contrast administration on CT or MRI. While the majority of these tumours are benign papillomas, few of them may be malignant choroid plexus carcinomas. Surgery is the treatment of choice with total resection when possible, followed by adjuvant therapy.[1,2]

CASE: A 3 year old girl was brought to emergency service with altered sensorium following an episodes of right focal motor seizures which got secondarily generalized. She was born of full term normal pregnancy and had normal milestone development. Clinically she was irritable and had right hemi paresis. Optic fundi showed gross papilloedema. CT scan of the brain revealed an ovoid, hyper dense, well enhancing fronds like mass in the trigone, occipital and temporal horns of the left lateral ventricles with dilatation of all the four ventricles with periventricular luscencies (fig.1). Her routine biochemical and hematological parameters were normal. She underwent a left temporo- parietal craniotomy,

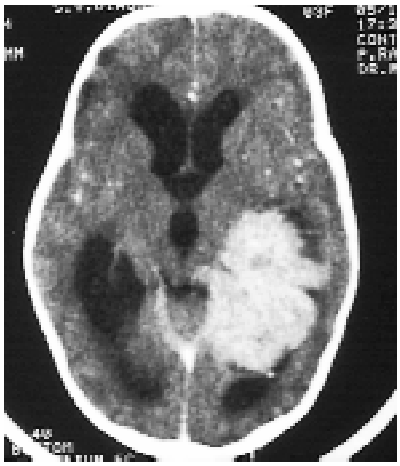

Figure 1: preoperative contrast CT scan

and near total decompression of the tumour, via trans sulcal approach. Tumour was very vascular, reddish and soft. Histopathological examination (figure-2) revealed intricate

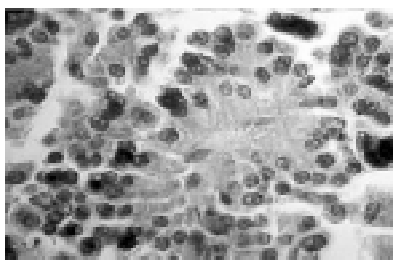

Figure 2: Microscopic appearance of the tumour (40x)

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papillary fronds with fibrovascular core which are lined by mildly oxyphilic to clear, pseudo stratified columnar epithelium, with severe nuclear pleomorphism, atypical mitotic activity and tumour giant cells. The solid areas of the tumour were congested, hemorrhagic with coagulation necrosis. The tumour cells on immuno-histochemistry staining were positive for cytokeratin, S100, Glial fibrillary acid protein and negative for Vimentin.

Parents of the child refused further treatment. The child died 5 months later due to progressive disease.

DISCUSSION

Choroid plexus papillomas (CPPs) are rare and comprises of 0.4-1% of all primary brain tumours. These commonly occur in children and constitute about 1.5-4% of childhood intracranial neoplasms.[3] CPPs are non-malignant and outnumber carcinomas by a ratio of 5: 1.[1] Malignant evolution may occur in 10 percent of CPP. Russel and Rubinstein defined choroid plexus carcinomas (CPCs) as WHO grade 3 tumours with characteristic features such as presence of invasion of adjacent neural tissue, loss of papillary architecture, mitosis, and cellular pleomorphism. These tumours frequently express CEA and CD44 as markers.[4] CT scan usually demonstrates a homogeneously hypodense to slightly hyperdense enhancing mass with cystic areas. There may be associated hydrocephalus. MRI is the investigation of choice for the diagnosis of choroid plexus tumours, and distinguishing between benign from more aggressive choroid plexus tumours.[5] The aetiology of CPPs is unclear. Germline mutations in P53 have been described.[6] Gross total resection of intra-ventricular CPCs should be the primary objective, as it affects the survival. The prognosis for CPCs is dismal, with a 5-year survival rate of 26%. Localized carcinomas often do well with adjuvant therapy in the form of chemotherapy and/or cranio-spinal irradiation. While pre-operative chemotherapy has been advocated to reduce vascularity and facilitate total surgical excision, radiotherapy has been associated with significantly better survival rates in CPCs. Relapse after primary treatment carries poor prognosis. Second surgery for relapsed disease can be beneficial.[7]

CONCLUSION

Choroid plexus carcinomas are rare intra cranial tumours and carry poor prognosis. Multimodality treatment consisting of aggressive surgery followed by Chemo – radiotherapy is the treatment of choice.

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