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Choroid Plexus Carcinoma

[1]Meera Chauhan,[1]Foram A. Patel,[1]Snehal M. Joshi,[2]Dr S P Srinivas Nayak

[1]Pharm D Intern,[2]Asst. Professor,

[1,2]Department of Pharmacy Practice, Parul Institute of Pharmacy & Research, Parul University, Vadodara, India-391760

Abstract:

Introduction: Choroid plexus carcinoma is an uncommon/rare cause of a hemispheric cerebral tumor arising from lateral ventricles.

Discussion: The exceedingly low prevalence of CPC in children has hampered the creation of standardized clinical trials, leaving therapy options to expert opinion and case studies. Surgery, chemotherapy, radiation, and autologous hematopoietic cell rescue are all options for treatment.

Conclusion: In rare conditions like these possible evidence-based treatment has low evidence and further establishment of more data-based studies are to be done to further out more options and adjuvant therapies for the future.

***KEY WORDS* – Choroid plexus carcinoma, Pediatrics, hemispheric**

I. INTRODUCTION

Choroid plexus carcinoma is an uncommon/rare cause of a hemispheric cerebral tumor arising from lateral ventricles in children with an annual incidence rate of 0.10 per 100,000 children)(2). It is associated with a dismal prognosis, especially if incompletely resected. Accurate histopathologic diagnosis is imperative, and this neoplasm should always be included in the differential diagnosis of a papillary intraventricular tumour. CPC lies under WHO grade III classification of tumors(1) These tumors present with hydrocephalus or macrocephaly. They are challenging surgically as their high vascularity often leads to significant blood loss in small children with a low circulating blood volume. Overall survival in most series of choroid plexus carcinomas has been poor. (3) Due to the lower survival rate,therapy for patients with choroid plexus carcinoma guidelines may not be possible and the treatment of chemotherapy or radiotherapy is a risk in pediatric population.

II. CASE SUMMARY

A four-month-old male child came with complaints of fever and increased head size since 2 weeks' acute onset generalized noticed at an abnormally rapid rate by the parents. The patient was taken to another private hospital wherein he was advised for a MRI, then was transferred to another hospital for further management wherein he was diagnosed as HYDROCEPHALUS WITH AFI (CPC). The patient was prescribed 0.45% DNS along with INJ. Febrinil for management of fever. Parents were explained regarding prognosis, nature, consequences of disease, need for surgery with high risk, mortality and morbidity risk and then was referred to higher centre for neurological intervention. There was no family history of any major medical illnesses, but it was a third degree consanguinity marriage. The Pateint was exclusively breast fed and was immunized upto Pentavac III and BCG scar was present. The patient was able to hold his neck, turn towards sounds, cooed and had a social smile which shows normal development. The patient's mother was on T. Labetalol during pregnancy.

At the time of admission, the patient was conscious and irritable with a head circumference of 48.5 cms (>97th centile). In the hematological examination, there was decrease in the haemoglobin and platelet count 7.00 g/dl and 65,000/cm^2 respectively. ESR was increased (68 mm) which shows signs of inflammation and CRP was raised (12 ug/ml). The rest of all lab investigations were within normal limits. The patient was given 1 Blood transfusion before surgery.

CT (computed tomography) brain was showing large irregular hyper dense lesion with calcification and peri-lesional edema in the left parietal region with infiltration of lateral ventricle with hydrocephalus. Therefore, for further investigation a Neonatal Cranial Sonography was done which showed a large extra-axial solid cystic mass lesion present in the left temporal-parietal region causing mass effect and displacement of left cerebral structure medially and causing midline shift of 2.2 cm. The solid component was present in the medial end of the lesion seen as lobulated well define homogenous hyperechoic lesion measuring (5.5cm x 5.5 cm x 4.5cm) in size. Multi-lobulated cyst components were seen lateral to the lesion reaching up to the meningeal surface, the overall dimension of the cyst component (7.5cm x 5.5cm x 5cm) (volume~100cc) the right ventricle was displaced medially and affected.

Magnetic resonance imaging (MRI) was showing a large size heterogeneous lesion in the left cerebral hemisphere involving the front-parietal and temporal convexities with cystic and avidly enhancing solid components with mass effect and middle shift possibility of choroid plexus carcinoma merit consideration. Mild dilatation of lateral ventricle with mild periventricular ooze. The cystic encephalomalacia changes in left front-parietal and temporal lobes with white matter volume loss and cortical thinning.(Figure 1 & 2)

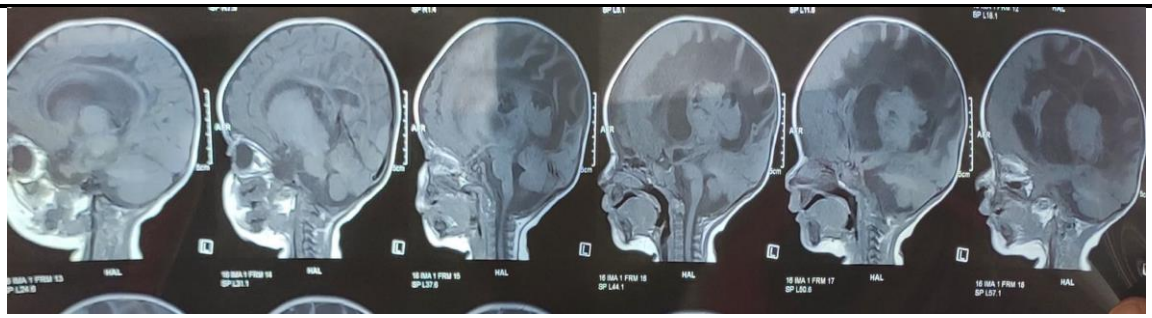

Figure 1MRI of the patient

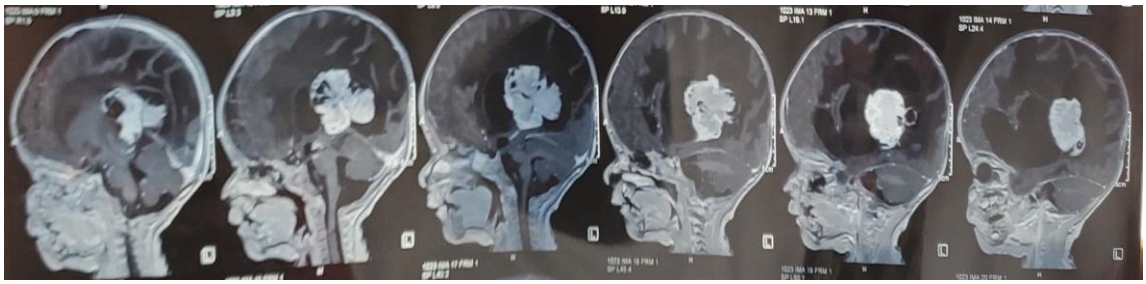

Figure 2 MRI of the patient

After the doctor discussion about the patient illness with the patient's parents for patient craniotomy with the consent, the patient was taken for emergency surgery. Risk of mortality was a concern in this patient during the therapy due to increase in tumor size. Perioperatively prophylactic antibiotics were started. A left parietal-occipital craniotomy was made and gross total excision of the tumor was done. While in the surgery, the patient was declared dead due to excess of blood loss.

Table 1 Pharmacotherapy

DRUG	**DOSE,ROUTE & FREQUENCY**	**DAY 1**	**DAY 2**	**DAY 3**	**DAY 4**	**DAY 5**
INJ. CEFOTAXIME	50mg/kg/day @ 350 mg IV 6 hourly	√	√	√	√	√
INJ. AMIKACIN	15mg/kg/day @ 35mg IV 6 hourly	√	√	√	√	√
TAB. DIAMOX (Acetazolamide)	50mg/kg/day @ 350mg/day PO ½ tab TDS	√	√	√	√	√
INJ. PARACETAMOL	10 mg/kg/day @70 mg IV TDS		√	√	√	√
INJ. ½ DNS + 5cc KCL	100ml/kg/day @ 30 ml/day		√	√		

Table 2 Daily Progress Chart of the Patient

DAY	**DAILY PROGRESS**
DAY 1	HR:126/min;$SP0_2$:98%;RBS:77mg/dl;MRT:98.4ºF;BP:98/64mmHg;1 x LGF
DAY 2	BP:102/66mmHg;RBS:118mg/dl;SPO_2:96%;HR:130 bpm;9 am-keep 2x PRC ready; 1 PCV;given very high risk content for Sx;Repeat CBC platelets after 48 hrs.7:30 pm-2 HGF spikes,taking BF well all reports noted CT Abs. acc. TPR 6 hourly.
DAY 3	8AM MRT: 98.9;HR:124 bpm; RR:34; SPO2:96%; BP:102/66mmHg; RBS:112mg/dl. 8PM MRT:101ºF /103ºF 2x HGF; Stool passed; feeding well;urine passed; Physician fitness taken for Sx-can be taken to OT with High Risk.
DAY 4	100ºF/104 ºF/101 ºF HR: 134 bpm; RR: 26; SPO2: 96% ;RBS: 112 mg/dl; Evening: wt: 7.15 kg;RR:34;Temp: 100ºF/ 101ºF
DAY 5	Wt: 7.20 kg;MRT:100ºF/102ºF;HR:124 bpm;RR:24;BP:98/66; RBS:102 mg/dl;SPO2:96%; Check for blood C/S; Neurosurgery Reference;Fundus exams ?optic atrophy; Opthal. Reference: Advised severe visual impairment secondary to hydrocephalus. Surgical consent for very high risk mortality taken and Patient shifted to Neurosurgery OT. Neurosurgery Note: Due to excessive blood loss patient was non-responsive and was declared dead.

III. DISCUSSION

Choroid plexus carcinoma (CPC) is an uncommon cause of hemispheric brain tumors in children that arise from the lateral ventricles. The possible differential diagnosis for such a hemisphere brain tumor includes choroid plexus papilloma (CPP), ependymoma, atypical teratoid rhomboid tumor, glioma, astrocytoma, and primitive neuroectodermal tumor (PNET) . In order to differentiate and establish a confirmed diagnosis, radiopathological correlation with tissue immunohistochemistry is critical.
The exceedingly low prevalence of CPC in children has hampered the creation of standardised clinical trials, leaving therapy options to expert opinion and case studies. Surgery, chemotherapy, radiation, and autologous hematopoietic cell rescue are all options for treatment. Although Surgery tends to be the fundamental option for the patient here . Radichemotherapy and Radiation therapy are the best regimen ,but a combination utilizing platinum and etoposide as backbone therapy is preferred. Early use of these therapies have affect in decreasing in the progression of disease and increase survival rates of patients. (2,3,5,8,10). A meta-analysis showed that the rate of progression free survival was around 37% in resection of tumor and overall survival was around 59%. Along with that survival rate for patient with whole-brain tumor removal was around 15% .(7)
We saw cauliflower-like structures in the MRI of this patient which confirms for tumor along with Hydrocephalus (4). It was noted in this article that the blood loss may be life-threatening due to the whole circulating volume of blood that may be lost during vascular resection which happened in this case. (2) A study on infant related CPP studies concluded that tumors were mainly in the lateral ventricle and had a male predominance but this can be biased but in this condition, it fits well.(9)

IV. CONCLUSION

In rare conditions like these possible evidence-based treatment has low evidence and further establishment of more data-based studies are to be done to further out more options and adjuvant therapies for the future.

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