Case Reports

Choroid Plexus Carcinoma

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Primary neoplasms of the choroid plexus are uncommon lesions, constituting 1% to 2% of all childhood brain tumors(l). Choroid plexus carcinoma (CPC) is a very rare tumor. Choroid plexus papilloma (CPP) is a common primary choroid tumor and its prevalence is four times higher than CPC. CPC is often extensive at diagnosis occurs in infants and has a poor prognosis. In view of the rarity of this condition and paucity of data from our country(2,3), this case is being reported.

Case Report

A 2-year-old developmentally normal boy presented with 15 days history of alteration in sensorium, vomiting, seizures, and weakness of right half of the body with associated history of low grade fever. There was no history of cranial nerve involvement, decerebration or enlargement of head. Birth and immediate postnatal history were normal. On examination the child weighed 8.6 kg and the head circumference was 45 cm. He was afebrile and normotensive. On neurological examination the sensorium by the Glasgow coma scale showed best verbal response of 3, best motor response of 4 and best eye response of 2. Fundus showed early bilateral optic atrophy. The child had right hemiparesis with generalized hypertonia, hyper-reflexia and extensor plantar response.

Investigations revealed a normal X-ray chest, normal blood counts, and a non-reactive Mantoux test. Examination of the cerebrospinal fluid was normal. CT scan of the head revealed a large hyperdense enhancing lesion in the left lateral ventricle with an area of necrosis and generalised dilation of the ventricular system. MRI head *(Fig. 1)* showed a large heterogenous mass arising from the left lateral ventricle.

The child was subjected to neurosurgical intervention. Tumor decompression was done through a left parietal craniectomy. The tumor was encountered at a depth of 0.5 cm from the surface. It was vascular, soft with an ill-defined plane of cleavage and grossly invasive. Histopathology of the tumor *(Fig. 2)* showed sheets of monomorphic cells with nuclei showing dispersed chromatin with small amount of amorphous cytoplasm. At places there was attempted gland formation and acinar pattern separated by thick fibrous septae with blood vessels; fair amount of mitotic activity was seen. The child was given radiotherapy post-operatively.

Discussion

CPP is the most common primary choroid tumor and in most series outnumbers CPC by a ratio of 4:1(1,4,5). Both CPC and CPP may arise in the lateral, 3rd or 4th ventricle. All CPC in children occur in the lateral ventricles (67% left, 33% right) while most tumors in adult are in the fourth ventricle(6).

There is clinical overlap between CPP

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and CPC. The tumors become symptomatic at any time during childhood, but most commonly in the first two yr of life(4,5,7,8).

Children with CPP have signs and symptoms of increased intracranial pressure due to hydrocephalus. Hydrocephalus occurs due to over secretion of the cerebrospinal fluid and also obstruction due to basal arachnoiditis following a bleed from the tumor. Children with CPC often have focal neurological deficits, without associated hydrocephalus. In one series(9) focal presentation was more common in children more than 3 yr of age. Subarachnoid seeding can occur with both CPC and CPP. Large invasive CPC can manifest as seizures.

Neuro-radiographic appearances are not characteristic in CPC. CPP are more homogenous, more commonly have hydrocephalus and rarely invade the parenchyma(9) whereas CPC are heterogeneous in appearance on CT and MRI scans. Malignant lesions that can mimic CPC include primitive neuroectodermal tumors, malignant gliomas, ependymomas or metastasis to the choroid plexus.

The criteria for the diagnosis (10, 11) of primary CPC are as follows: (i) Invasion

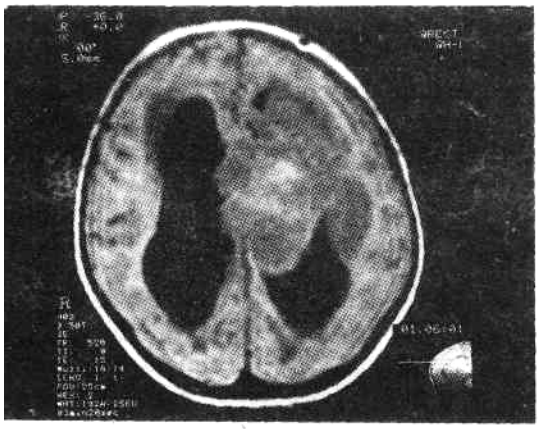

Fig. 1. MRI head showing a large heterogenous mass arising from the left lateral ventricle.

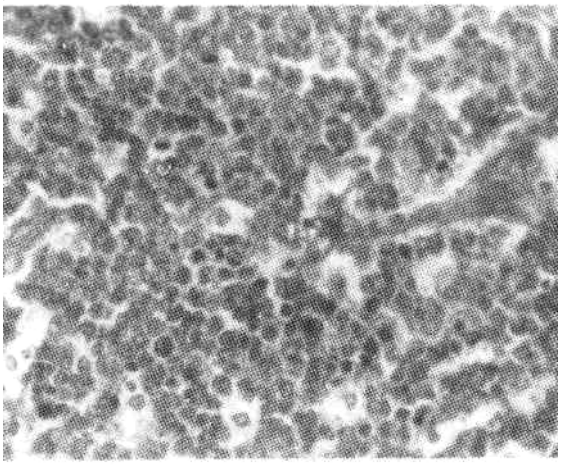

Fig. 2. Microscopic examination showing sheets of cells with prominent mitoses around a fibrovascular core, suggestive of choroid plexus carcinoma (H&E × 200)

of the adjacent neural tissue with infiltrating cells that assume a diffuse growth pattern; *(ii)* Loss of the regular papillary structure of the neoplasm where invasion is occurring and obvious malignant alteration in the cells; *(in)* transition of normal choroid plexus architecture to an undifferentiated pattern. Grossly both CPC and CPP are friable, vascular tumors. Histologically CPP are usually composed of a single layer of epithelium supported by well vascularised connective tissue. In CPC the regular architecture is lost, nuclei vary in size and mitoses are prominent. Immune-histochemical staining is not useful in separating the two conditions. The therapy of CPC consists of surgical resection with adjuvant radiotherapy or chemotherapy. The prognosis for these tumors is poor.

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NOTES AND NEWS

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