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Congenital Primary Cerebral Neuroblastoma

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Although brain tumors constitute the second largest group of tumors in children, they are extremely uncommon in the neo-

natal period and account for about 1% of all childhood brain tumors(1). Moreover, a difference in the pattern of disease from that seen in older children is observed which includes a high incidence of teratomas and a predominance of tumors in supratentorial sites(2).

A case of supratentorial neoplasm in a neonate of the nature of a "Primary Cerebral Neuroblastoma" is described. The tumor in itself is rare and this is probably the first case in literature with presentation at birth and which was diagnosed antenatally.

Case Report

A 30-year-old primi gravida with seven months amenorrhea was referred for an ultrasound report suggestive of congenital hydrocephalus in the fetus. The patient was examined in the antenatal clinic and referred to the radiologist for a repeat ultrasonography (USG).

Repeat USG confirmed the diagnosis of congenital hydrocephalus and also sug-

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gested the presence of an intracranial space occupying lesion. Unfortunately, the patient did not follow up in the intervening period and presented directly at term in labor. An emergency LSCS was performed for obstructed labor and a 2.3 kg male infant with mild birth asphyxia was delivered.

On examination, there was an obvious hydrocephalus. The head circumference was 38 cm (expected upto 35 cm), sutures widely separated and dilated veins on the scalp. There was a left supranuclear facial palsy but no other focal neurological deficits. Cry and activity of the infant were weak. There was no skin rashes, hepatosplenomegaly or lump in abdomen.

Cranial USG done on the infant revealed a huge echogenic mass in the brain with irregular margins crossing the midline. The right lateral ventricle was compressed and there was an obstructive dilatation of the left lateral ventricle (*Fig. 1*).

With this clinical picture and USG report a diagnosis of congenital obstructive hydrocephalus due to an intracranial space occupying lesion was made. Neurosurgical reference was made and a ventricular tap performed. Twenty ml of fluid was drained. Routine analysis of this fluid revealed elevated protein of 200 mg/dl. No abnormal cells were seen. A CT scan of the brain could not be performed because of poor general condition. The infant succumbed within 36 hours of birth.

At post mortem examination, the brain weighed 550 g and showed well developed cerebral hemispheres. A large, well defined, sharply demarcated greyish pink lobulated tumor mass was present in the right frontoparietal region extending upto the occipital lobe posteriorly and the midbrain inferiorly. Cut surface of the tumor appeared granular and presented large areas of hemorrhage and necrosis. Surrounding brain parenchyma was markedly edema-

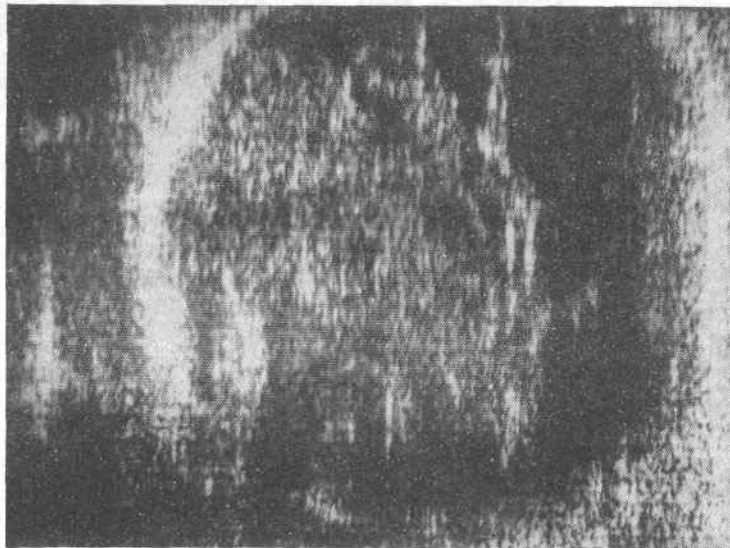


Fig. 1. Ultrasound showing tumor mass in right fronto-parietal region

tous. Liver, spleen and sympathetic tissue were found to be normal. There was no evidence of a tumor or metastasis at any other site.

Histology revealed a highly cellular tumor with moderate degree of desmoplasia. Clusters and cords of tumor cells were separated by fibrous septae. Small round to spindly tumor cells with darkly staining nuclei having moderate amount of eosinophilic cytoplasm was seen. Homer Wright rosettes were found. Mitotic figures and ganglion cell formation were inconspicuous. Immunohistochemical staining with neuron specific enolase was positive confirming the diagnosis of neuroblastoma (Fig. 2).

In view of the histopathologic appearance similar to peripheral neuroblastomas and absence of gross or microscopic involvement of the peripheral sympathetic

nervous system, presentation at birth a diagnosis of "Congenital Primary Cerebral Neuroblastoma" was made.

Discussion

Primary cerebral neuroblastoma is one of the rare types of primitive neuroectodermal tumors. The other more common variety being cerebellar medulloblastoma. The tumor is designated as neuroblastoma due to the histopathologic similarity to the more commonly seen neuroblastoma of the peripheral sympathetic nervous tissue(2).

Till 1976, about 12 sporadic isolated case reports were found in literature. In 1976 Horten and Rubinstein(3) published a comprehensive review comprising of 35 cases collected over a period of 12 years. Beyer and Edwards(4) have reviewed the long term follow up and suggested thera-

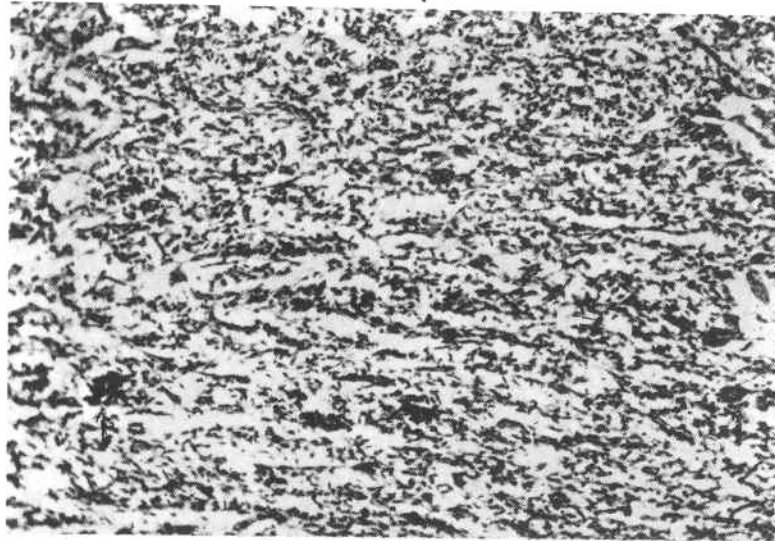


Fig. 2. Photomicrograph showing cellular tumor with clusters and cords of round to spindly shaped cells. (H & E \times 10)

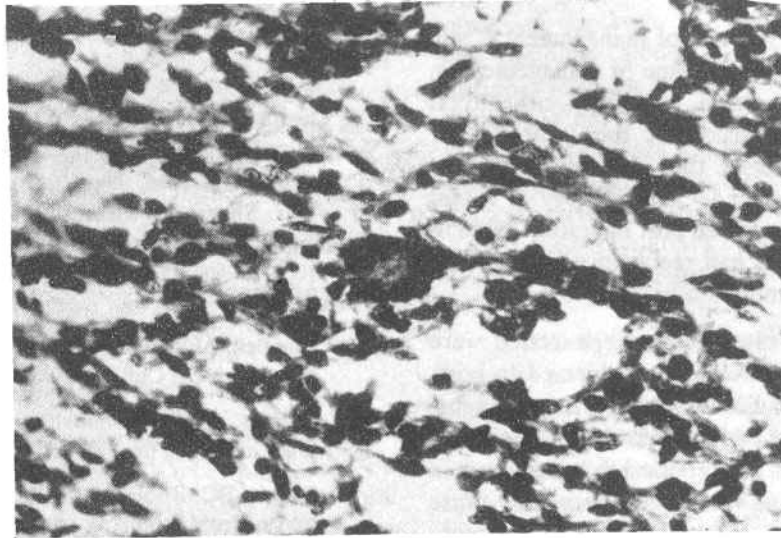


Fig. 3. (Inset) High power of the same with Homer-Wright rosette (arrow) suggestive of neuroblastoma (H & E x 40)

peutic guidelines for management of such patients.

Eighty five per cent of cases with this tumor have been encountered in the first decade of life of these 65% occurred below 5 years. Only 18-20% of these have occurred in infants less than 6 months of age. The age at diagnosis ranged from 2/2 months to 35 years. However, no case of tumor presenting at birth has yet been reported.

The duration of symptoms before diagnosis has ranged from 3 days to two years. Majority of the cases presented with raised intracranial tension and acute hydrocephalus. Focal deficits were reported in 4 out of 11 cases in Bergers series.

On investigation, raised CSF protein and presence of abnormal cells have been

reported in few cases. CSF examination in our patient was normal. Angiography may reveal a large avascular mass or rarely a tumor blush may be seen. Ultrasonography of the skull and CT scan may reveal 2 types of pictures: (i) Solid tumor mass and (ii) Cystic tumor which grows to a large size.

Elevated blood, urine or CSF catecholamines have not been demonstrated in cerebral neuroblastomas. However, no data is available on the catecholamine concentration in the tumor mass itself.

On histopathology, the feature which sharply distinguishes cerebral neuroblastoma from other neuroepithelial neoplasms is the unusual capacity of these tumor cells for stroma induction, based on which, these tumors have been classified into 3 subgroups: (i) Classical, (ii) Transitional and (iii) Desmoplastic. The histo-

pathology in our patient was characteristic of the classical variety.

The ideal mode of management is still uncertain. The first line of management in the cases reported, is surgery—either total or partial excision of the tumor mass. Twenty eight of the 35 cases in Rubinstein's series underwent surgery of whom 22 patients survived post operation. Nine of these patients had recurrences in a period ranging from 6 months to 7 years.

All 11 cases in Berger's series were operated and were also subjected to post-operative radiotherapy to prevent recurrences. Four patients showed a recurrence. Recurrences are reported to be common in those patients with solid tumors and those undergoing subtotal resection. Recurrences were managed by surgery or a combination of multiple chemotherapy with radiotherapy.

In summary, primary cerebral neuroblastoma is a rare type of neuroectodermal

tumor. Probably this is the first case reported, that has presented at birth.

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Post Mortem Radiography of Perinatal Deaths: An Aid to Genetic Counselling

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Post mortem radiography of perinatal deaths is a simple and informative investi-

gation which, in selected cases, can help in reaching a correct diagnosis and accurate genetic counselling thereafter(1).

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