Hepatoblastotna Associated with Congenital Hemihypertrophy

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Congenital hemihypertrophy is an uncommon condition with a reported incidence of 1 in 86,000 live births(l). There is a well documented association of hemihypertrophy with a number of malignant and non-malignant intra-abdominal masses including Wilms' tumor and adrenal cortical neoplasia(2). However, association of hemihypertrophy with hepatoblastoma is an uncommon condition and only two such cases have been reported in literature(3,4).

Case Report

A $1^{1/2}$-year-old girl was brought with a 3 months history of mass in the right side of abdomen. She had hypertrophy of right arm and right leg since the age of 3-4 months. There was no history of fever, loose motions, pain abdomen or loss of weight. On examination, she weighed 10 Kg, had right sided hemihypertrophy without any angiomatous malformation or local arteriovenous fistula and had normal blood pressure. On abdominal examination, there was a large intra-abdominal mass (10 cm x 15 cm) in the right hypochondrium which was firm, nodular, non-tender/moving with respiration and was neither bimanually palpable nor ballotable. An abdominal ultrasound showed multiple areas of decreased echogenecity throughout the liver with bilateral normal kidneys and adrenal glands. Liver functions, kidney functions, hemogram and platelet count, serum calcium, phosphorus, alkaline phosphate and X-ray chest were normal. Serum alpha-fetoprotein could not be done due to lack of facilities. On exploratory laparotomy there was a diffuse involvement of both the lobes of liver by a nodular, firm and vascular tumor. Hence, only a biopsy was taken from the tumor.

Histopathology showed fetal predominant hepatoblastoma (Fig. 1). Her postoperative stay was uneventful and she was discharged. However, she was lost to followup.

Discussion

The two most commonly reported tumors in association with hemi-hypertrophy are Wilms' tumor and adrenal tumor. Ringrose et al.(5) in a study of 129 cases of hemihypertrophy have reported a 5% association of Wilms' tumor and adrenal tumor. However, the association of hemihypertrophy with hepatic tumor is very uncommon. In one large series of 54 cases of hepatoblastoma, only one case had hemihypertrophy (3), and there is another report of a single

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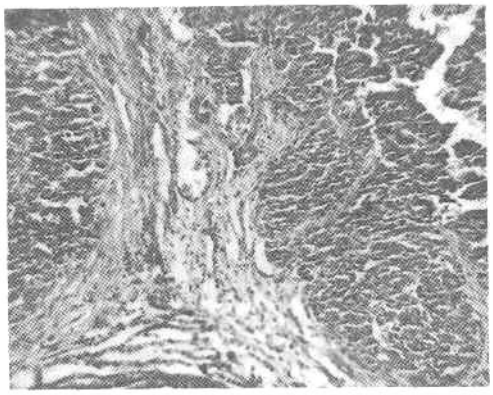

Fig. 1. Microphotograph showing tumor cells arranged in two cell thick laminae recapitulating those of fetal liver(H & E ×100).

case(4). In yet another case report, the child had hemihypertrophy in association with hepatic hemangioendothelioma(6).

Abdominal ultrasound is a, useful non-invasive modality for the detection of intra-abdominal masses in cases with hemihypertrophy. Raised alpha-fetoprotein is a non-specific marker yet it is helpful in diagnosing hepatoblastoma(3).

Hepatoblastoma is the most common primary hepatic tumor in children under 5 years of age(3,7)- Besides hemihypertrophy, hepatoblastoma can be associated with other congenital anomalies also; the exact incidence and cause of this association is unknown. In children, epithelial hepatoblastoma with predominantly fetal pattern is the most common type and has better prognosis(3). Complete surgical resection remains the key in achieving long term survival(8). Radiation or chemotherapy has little to offer as a primary treatment modality(3). Overall reported mortality is 76%(3).

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