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STUDY OF LIVER TUMORS IN INFANCY AND CHILDHOOD

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ABSTRACT: AIM: This study has been done with an aim to evaluate presentation, evaluation, treatment and outcome of liver tumors in childhood. **MATERIALS AND METHODS:** Over a period of 3 years, liver tumors, both benign and malignant, which presented to the Department of Paediatric Surgery, Niloufer Hospital were evaluated prospectively and retrospectively. **RESULTS:** 20 cases of liver tumors that presented to the Department of Paediatric Surgery, Niloufer Hospital during the study period were evaluated. 14 were hepatoblastomas and 6 were benign tumors. Only 4 out of the 14 hepatoblastomas were amenable for primary resection. Survival for malignant tumors was 64.2%. Prognosis of benign tumors was universally good. **CONCLUSION:** Liver tumours are relatively rare in paediatric population. They can be diagnosed preoperatively by biochemical and radiological investigations. Malignant tumours account for 70-75% of all liver tumours in children.

KEYWORDS: Liver tumors, Hepatoblastoma, Mesenchymal hamartoma.

INTRODUCTION: Tumors of liver although infrequent pose a considerable therapeutic and diagnostic challenge. Primary liver tumors constitute 0.5 to 2% of all pediatric tumors. 70 to 75% of liver tumors in children are malignant. Liver tumors commonly present as asymptomatic abdominal mass found on routine physical examination. They need to be differentiated from non-tumorous causes of hepatomegaly.

CLASSIFICATION OF TUMOR LIKE LESIONS OF LIVER

	BENIGN	MALIGNANT
EPITHELIAL	Focal nodular hypoplasia Hepatocellular adenoma Nodular regenerative hyperplasia Mixed hamartoma Developmental cysts	Hepatoblastoma Hepatocellular carcinoma Fibrolamellar carcinoma
MESENCHYMAL	Infantile hemangioendothelioma Mesenchymal hamartoma Cavernous hemangioma Infantile pseudotumor	Undifferentiated/ embryonal sarcoma Angiosarcoma Rhabdomyosarcoma Rhabdoid tumor
GERM CELL TUMORS TERATOMA		Endodermal sinus tumor

Benign liver tumors are rare and often present in infancy. Hemangioendothelioma which is the most common benign tumor is commonly treated with steroids, if symptomatic. Mesenchymal hamartomas are second most common tumor which is commonly amenable for surgical resection. Other benign tumors are commonly seen in teenage girls.

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Hepatoblastoma is the most common liver tumor seen in children with a male preponderance (1.7:1). Median age at diagnosis is 1 year. Hepatoblastoma is most often unifocal and right lobe of liver is commonly involved.

Hepatoblastoma can be classified into 6 histological subtypes based on predominant epithelial or mesenchymal components they are foetal, embryonal, trabecular, small cell variants and teratoid and non teratoid mixed variants. Liver tumours in children commonly present as abdominal mass.

DIAGNOSTIC WORKUP FOR CHILDREN WITH LIVER TUMORS

Younger than 5 years:

Complete blood picture.

Liver function tests.

Alpha fetoprotein.

B-HCG if precocious puberty.

Older than 5 years:

Complete blood picture.

Liver function tests.

Alpha fetoprotein.

Hepatitis B serology.

Vitamin B12 binding protein.

For all patients:

Plain X- ray abdomen.

Chest x- ray.

CT scan abdomen with IV contrast.

Children's cancer group (CCG) and south west oncology group (SWOG) had given a staging of hepatoblastoma based on the extent of tumor and surgical resection. International society of pediatric oncology (SIOP) has given a preoperative pretext staging by number of sectors of liver involved.

CCG and SWOG advocate initial laparotomy and resection of operable tumors and biopsy of those not resectable followed by chemotherapy for 2 to 3 cycles and a second look laparotomy. German cooperative study group (study- HB- 89) also confirms the importance of primary surgery. SIOP study group differs from the above and recommends primary chemotherapy. All children with hepatoblastoma require chemotherapy before or after liver resection. Most of the chemotherapeutic regimens include cisplatin, doxorubicin, vincristine and 5-fluorouracil.

MATERIALS AND METHODS: Over a period of 3 years, 20 liver tumors in infancy and childhood presented to the Department of Paediatrics Surgery, Institute of Child Health, Niloufer Hospital, Hyderabad. These patients were evaluated prospectively and retrospectively, postoperative follow up ranged from 6 months to 3 years.

All cases of liver tumors were subjected to thorough clinical and biochemical evaluation which included serum AFP levels. Radiological investigations included CT scan abdomen with IV contrast.

Figure 1

Fig. 1: CT scan with IV contrast showing liver tumour with thick internal septae s/o mesenchymal hamartoma

Treatment for benign liver tumors included prednisolone (4-5mg/kg/day for 28 days) for symptomatic hemangioendotheliomas and surgical resection for mesenchymal hamartomas.

Diagnosis of malignant liver tumors was confirmed by elevated AFP levels and CT scan findings. All these cases were stabilized and taken for laparotomy. If primary resection was not possible a biopsy was performed followed by 2 -3 cycles of chemotherapy followed by a second look laparotomy. All patients received 6 cycles of chemotherapy consisting of cisplatin 20mg/sq.m from day 1 to day 5 and doxorubicin 25mg/sq.m from day 1 to day 3. Patients were followed up for a period of 6 months to 3 years.

Fig. 2: Tumour arising from left lobe of liver in a infant

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RESULTS: Over a period of 3 years, 20 cases of liver tumors, 14 malignant tumors and 6 benign tumors were evaluated. Mean age for benign tumors was 8 months and for malignant tumors was 16.21 months. All the patients had presented with mass abdomen. All the 14 patients with malignant hepatoblastoma had weight loss, loss of appetite and hard hepatomegaly. All of the 14 malignant tumors had elevated AFP levels with mean AFP value of 15,316 ng/ml. CT scan with IV contrast could diagnose 3 out of the 6 benign tumours as hemangioendotheliomas and the other 3 as mesenchymal hamartomas. CT with IV contrast with elevated AFP could diagnose all cases of hepatoblastomas. Only one case of hemangioendothelioma was symptomatic and responded to steroids. All cases of mesenchymal hamartomas were completely resected and had an uneventful postoperative follow-up.

Primary surgical resection was possible in 4 out of the 14 cases of malignant tumors. Pre resection chemotherapy resulted in reduction of tumor size in 8 out of the 10 cases which were given chemotherapy. Mortality occurred in 5 cases of malignant tumors (35.7%). Total survival rate for hepatoblastoma was 64.2%.

DISCUSSION AND CONCLUSION: Liver tumors are relatively uncommon in pediatric population accounting for 2% of all pediatric malignancies and 4% of all solid tumors in children.¹ 90% of liver tumors in children present before 3 years of life². 75% of primary liver lesions are malignant.³ Clinical examination and CT scan with IV contrast can easily pick up benign liver tumors.⁴ Clinical examination, CT with IV contrast and grossly elevated AFP are usually enough to diagnose hepatoblastoma.⁵ Symptomatic infants with hemangioendothelioma are successfully treated with steroids. Complete resection is the best treatment option for mesenchymal hamartoma.

Despite remarkable progress in modern chemotherapeutic protocols, it is widely accepted that complete resection of tumor is essential for cure of hepatoblastoma.⁶ In a trial sponsored by CCG and POG. Complete tumour resection was possible at primary laparotomy in 25% of cases. In our study primary resection was possible in 28.5 % of cases. Neo adjuvant chemotherapy resulted in significant reduction of tumor size in 78% of tumors according to Bryan J. Dicken et al,² In our study 80% of cases were responsive to neo adjuvant chemotherapy.

In conclusion, liver tumors are relatively uncommon in childhood and 70% of them are malignant. Most of these benign and malignant tumors can be preoperatively diagnosed. Chemotherapy and surgery are the management modalities in hepatoblastoma in children. Overall survival in our series for hepatoblastoma was 64.2%.

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