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RESEARCH ARTICLE

A RARE CASE REPORT OF HEPATOBLASTOMA IN A 1.5YR OLD CHILD.

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Abstract

Hepatoblastoma is the most common form of primary liver cancer in children, although it is a comparatively uncommon pediatric solid tumor. It is the third most common abdominal neoplasm in this age group after neuroblastoma and nephroblastoma. The primary treatment is surgical resection. The prognosis for patients with resectable tumors is fairly good in combination with chemotherapy.

Introduction:-

Hepatic tumors are rare in children. Primary tumors of the liver account for approximately 1% of malignancies in children younger than 15 years of age. Between 50% & 60% of hepatic tumours in children are malignant, with 65% of these malignancies being hepatoblastomas. Most of these tumors are seen before 5 years of age [2]. Amongst all primary malignant tumors in children, hepatoblastoma accounts for 1-4% of all cases [3]. Hepatoblastoma occurs predominantly in children younger than 3yrs of age and the median age of diagnosis is 1yr. The etiology is unknown. Because of its rarity and inherent malignant nature, diagnosis and treatment is problematic [4].

Case report:-

A 1.5yr old male child was admitted to our hospital with complaints of abdominal distension and intermittent fever for 1 month. Examination revealed some pallor, icterus, bilateral pitting pedal edema, bitot spots. On abdominal examination, liver was palpable 10cm below subcostal margin in mid-clavicular line. CBC showed microcytic hypochromic anemia (Hb-8 gm%) with thrombocytosis(TPC-7.2 lacs/cmm). LFT was deranged, AST-119IU/L, ALT-53IU/L,ALP-1177IU/L, S.bil(T)-9.1mg%, S.bil(D)-3mg%, s.protein-6.5gm%, PT INR-1.7.

USG abdomen suggested well defined mass measuring 94X86 mm in right lobe of liver with the vascularity on color Doppler and hypoechoic areas with necrosis suggestive of mass lesion with possibility of hepatoblastoma. Further investigation in the form of CT scan confirmed the diagnosis of hepatoblastoma affecting right lobe of liver. Serum alpha-fetoprotein (AFP) was high: >1000 ng/ml (normal value <20ng/ml). Hepatitis B, C and HIV tests were negative. Patient was referred to the cancer institute for further workup and treatment.

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Discussion:-

The most common primary malignant liver neoplasm in children is hepatoblastoma [4, 5]. Two thirds of cases occur before 2 years of age and 90% of the cases are found below 5 years of age. Males are affected twice as compared to females [4]. Hepatoblastoma in adolescents and adults are worse off than in children as they are diagnosed late.

There are several environmental risk factors associated with hepatoblastoma. Prematurity and very low birth weight has been found to be associated with the later appearance of hepatoblastoma [6-8]. A large number of congenital syndromes have been described in patients with hepatoblastoma, but only Edward's syndrome, familial adenomatous polyposis and Beckwith Wiedemann syndrome have been clearly shown to increase the risk of hepatoblastoma [6].

Thrombocytosis is commonly found in hepatoblastoma. Hepatic tumor in pediatric age group, if associated with thrombocytosis and raised AFP, strongly suggests the diagnosis of hepatoblastoma [10].

Serum AFP is the most useful laboratory test for hepatoblastoma. AFP is produced in the fetal liver and yolk sac, and levels decline to adult values during the first 6 months after birth. AFP can be used as a tumor marker for screening and confirmation of diagnosis but it is not specific. Serum AFP levels can also be elevated in hepatocellular carcinoma, hepatitis, cirrhosis, germ cell tumor, hemangioendothelioma, testicular tumor and gallbladder carcinoma. Very high levels of serum alpha-fetoprotein (AFP) are noted in over 90% of patients, the level often being higher than 1×10^5 μg/l. Low AFP levels (<100ng/ml) are aggressive and associated with poor prognosis. AFP is an excellent tumor marker that not only reflects the extent of the disease, but also is very useful for monitoring both the effect of ongoing therapy and tumor recurrence at an early stage.

Hepatoblastoma is mainly divided into two histologic types: epithelial, accounting for 56% of cases, and mixed epithelial/mesenchymal. The epithelial type can be further divided into fetal (31%), embryonal (19%), macrotrabecular (3%), and small-cell undifferentiated subtypes (3%). It has been observed that the subtypes have an effect on the prognosis, with the pure fetal type having the most favourable outcome and the small-cell undifferentiated type having the worst.

The right lobe of the liver is affected more commonly than the left, and in 35% of patients there is bilateral involvement. The left hepatic lobe derives oxygenated blood totally from the umbilical vein. The right lobe derives

blood from portal vein with lower oxygen concentration. The low oxygen favours the embryonic differentiation of the hepatoblastoma and its frequent localization in the right hepatic lobe [11].

Complete resection of the tumor remains the best hope for long-term survival; however, the advent of effective chemotherapy may permit cure in the presence of initially unresectable or metastatic disease.

Any child below 3yrs of age presenting with unexplained hepatomegaly and USG revealing a mass lesion, one has to keep in mind the possibility of hepatoblastoma.

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