Case Report :

Hepatoblastoma - A Rare Case Report In 14 Months Old Child

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Abstract:

Though hepatoblastomas are rare tumours, but when a child presents with abdominal lump, decreased appetite, weight loss and triad of hepatic mass by imaging studies, high levels of serum AFP and thrombocytosis, should always raise suspicion of hepatoblastoma. All other conditions should be ruled out by clinical examination, bio-chemical tests and histological studies. So early recognition of the tumour may lead to early and prompt intervention and speedy recovery. LIVER BIOPSY remains the GOLD STANDARD diagnostic tool for confirming HEPATOBLASTOMA. The role of paediatricians, pathologist and paediatric surgeons are very important to diagnose and manage the condition.

Keywords: *hepatoblastoma ,malignant liver tumours,alpha fetoprotein,abdominal lump*

Introduction :

Hepatoblastoma is the most common malignant tumour of the liver in children. About two-thirds of malignant liver tumours are due to hepatoblastoma [1]. Most of these tumours are seen below 5 years of age [2]. Amongst all primary malignant tumours in children, hepatoblastomas account for 1-4% of all cases [3]. Because of its rarity and inherent malignant nature, diagnosis and treatment is problematic [4].

The typical age, clinical history, presentation and clinical findings of hepatic tumours along with thrombocytosis and elevated alpha-fetoprotein are highly suspicious of hepatoblastoma.

Case Report :

A 14 months old male child, product of non-consanguinous marriage, hailed from GGP colony of Bhubaneswar brought by his mother with presenting complaints of abdominal swelling for 25 days and poor feeding for 10 days. The child was apparently alright 25 days back. On examination the child was afebrile, with weight 10 kg (50th-75th centile)and height was 76 cms (50th-75th centile). Bowel and bladder functions were normal. Child attained milestones as per the age. Immunized as per national immunization schedule. There is no food and drug allergies.Had one sibling of 8 years of age On abdominal examination, liver was palpable 8 cms below the right costal margin in mid clavicular line and spleen was not palpable. Other systems were normal.CBC showed thrombo-cytosis(8 lakhs/cumm), Hb% was 11 gm/dl, WBC was 10,000/cumm and RBC was 5.5 millions/ cumm of blood. Had normal liver function tests .USG abdomen showed echogenic solid mass in the right lobe of the liver , with well defined capsule .The dimension being 65×87 mms. Further investigations in the form of CT scan confirmed the extent of the mass.

Serum AFP level was >1000 ng/ml. Hepatitis B and C and HIV tests were negative. Liver biopsy was performed under ultrasound guidance. The report showed cells arranged in solid sheets and trabecular pattern. The cells had distinct cell membrane and uniform nuclei with minimal

pleomorphism and there was presence of fetal pattern and foci showing rosset formation. The mass from the right lobe of the liver was exercised. No chemotherapy or radiotherapy was given as the tumor is localized and well circumscribed and also resectable with no metastasis.

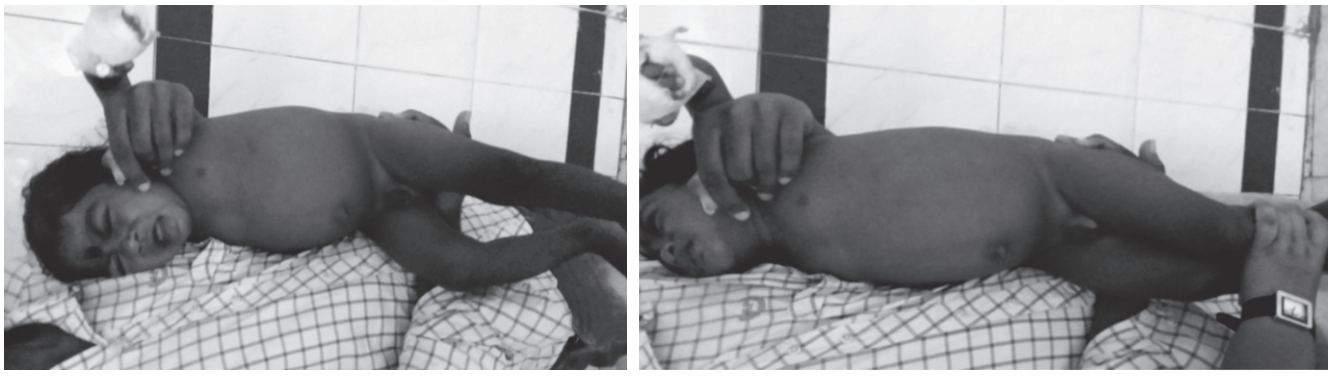

Figure 1: Child with abdominal lump and umbilicus was everted.

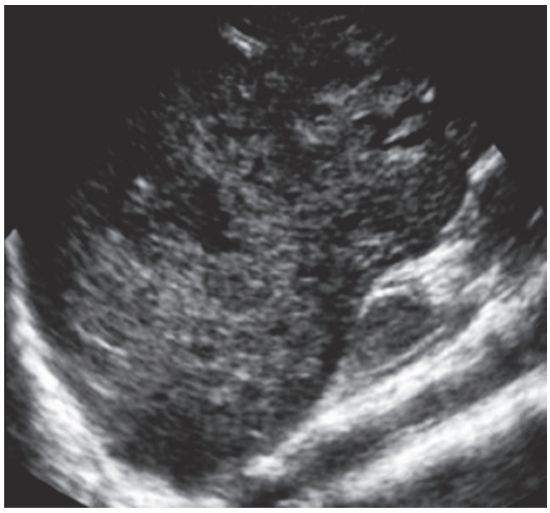

Figure 2: USG Abdomen showing echogenic soft tissue mass, well defined margin. Intralesional calcifications- are visible as areas of shadowing.

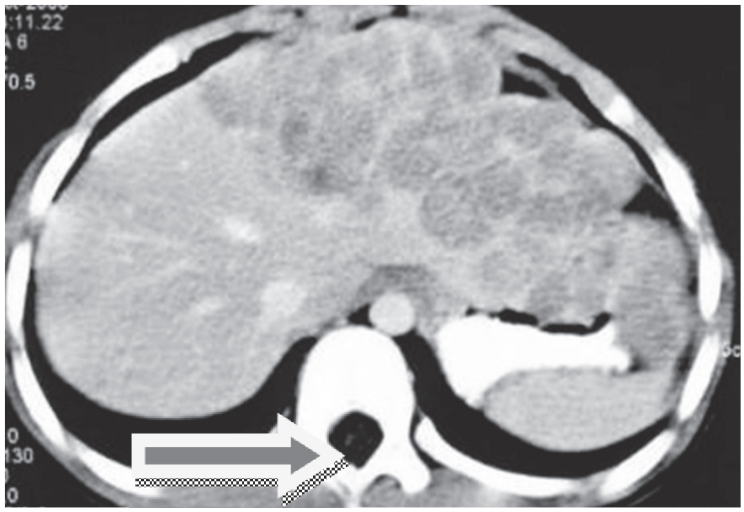

Figure 3: CT-Scan image showing heterogenousmass, and calcifications in the right lobe and left lobe of the liver. The mass is Hypoattenuated compared to surrounding liver. Necrotic and hemorrhagic areas were not seen.

Discussion:

Out of all childhood tumours, excluding leukemia and lymphoma, hepatic tumours form about 0.5-2% in frequency [5]. Most common age groups is between 2-5 years. Boys are 1.5-1.6 times more affected than girls. [7]. The incidence of hepatoblastoma is 1.2/million under 15 years of age.Hepatoblastoma in adolescents and adults carries worst prognosis, because they are diagnosed late [8,9].

The triad of a hepatic tumour, thrombocytosis and high level of serum AFP in a child between 6 months and 3 years of age is diagnostic of hepatoblastoma [10]. In this case all these pathognomic associations were present. In USG it appears as hyperechoic soft tissue intra hepatic mass [11]. Other investigations include CT, MRI, serum AFP and ? Hcg levels.

Histologically hepatoblastomas are classified into 2 types:

1. Epithelial (56%)
2. Mixed (44%)(epithelial + mesenchymal).

Epithelial is divided into 4 types:

1. Fetal (31%)
2. Embryonal (19%).
3. Macrotrabecular (3%).
4. Undifferentiated small cell. [12].

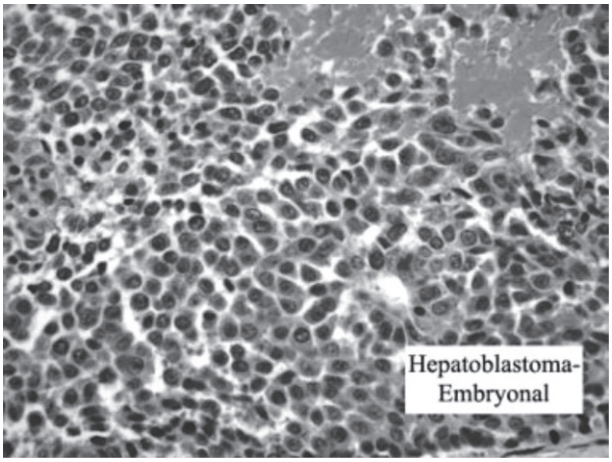

Figure 4:
Histopathology showing – tumor cells with large nuclei and scanty cytoplasm forming tubules and ribbons

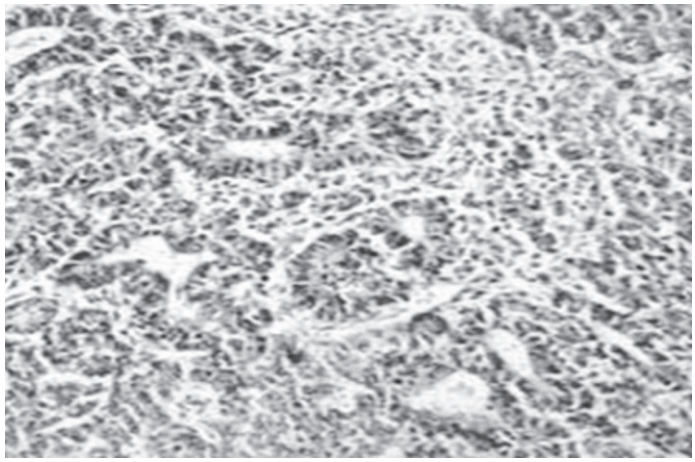

Figure 5:
Hepatoblastoma with a fetal pattern and a focus of osteoid (pink amorphous material).

High serum AFP levels in hepatoblastomas are not specific, because they are raised in other conditions also, such as, Hepatocellular carcinoma, yolk sac tumors, mesenchymal hamartomas, infantile hemangio endothelioma and focal nodular hyperplasia.

Certain variants of hepatoblastoma like small cell type are resistant to chemotherapy and they do not produce AFP, and carries bad prognosis. High levels of AFP, tumour extension and or metastasis signifies unfavourable prognosis.

Hepatoblastoma develops more frequently in the right lobe of liver [13]. This is due to low oxygen concentration, which favors the embryonic differentiation [13].

Cytogenetic analysis performed on hepatoblastomas reveal several alterations like chromosomal gains more frequent than losses. Trisomy 2, 8 and 20 were observed [14]. Deletion of 2p and 4q were associated with advanced disease and poor outcome [15].

Complete resection of the tumour remains the best hope for long-term survival. Chemotherapy is useful in case of unresectable and metastatic disease. Pre operative chemotherapy may decrease the tumour size and makes it more

resectable [16].

Conclusion:

Though hepatoblastomas are rare tumours, but when a child presents with abdominal lump, decreased appetite, weight loss and triad of hepatic mass by imaging studies, high levels of serum AFP and thrombocytosis, should always raise suspicion of hepatoblastoma. All other conditions should be ruled out by clinical examination, bio-chemical tests and histological studies. So early recognition of the tumour may lead to early and prompt intervention and speedy recovery. LIVER BIOPSY remains the GOLD STANDARD diagnostic tool for confirming HEPATOBLASTOMA. The role of paediatricians, pathologist and paediatric surgeons are very important to diagnose and manage the condition.

Contribution: GS – Final Manuscript, SB-Evaluation of case, review of literature, RM – Assisted pathology workup and management of case.

Conflict of Interest: None

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