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Case Study

Paediatric Hepatoblastoma in one year old female – A case report

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Keywords

Hepato-blastoma, Abdominal mass, Child

A B S T R A C T

In children, the most common malignancy in liver is the hepatoblastoma. A one year old female child presented with a lump in abdomen. Ultrasonography (USG) and CT scan revealed a solid hepatic tumor. Her complete blood count showed thrombocytosis. Serum alpha fetoprotein level was raised. A USG guided biopsy of the mass confirmed the diagnosis of hepatoblastoma. A hepatic tumor, in a child under the age of 3 years, associated with thrombocytosis and elevated serum alpha-fetoprotein is very likely to be hepatoblastoma. The mainstay of the treatment is complete surgical removal of the mass.

Introduction

In children, malignant tumors of liver are rare. About two-thirds of malignant liver tumors are due to hepatoblastoma. [1] 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] Because of its rarity and inherent malignant nature, diagnosis and treatment is problematic. [4]

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

Case report

A one year old female patient presented with abdominal mass. (Photo – 1) On general examination, patient was having good general status and was afebrile. The weight was 10.2 kg (75 percentile) and height was 76 cm (90 percentile). Patient had pallor but had good appetite and normal bowel functions. On abdominal examination, the liver was palpable, which is 7.5 cm below the costal margins and spleen was just palpable. Hematological investigation revealed moderate anemia (Hemoglobin - 8.4 g/dl; Hematocrit - 29.9%), thrombocytosis (platelets - 825 000/mm^3), normal liver function tests (ALAT 13 Iu/l, ASAT 38 Iu/l).

USG abdomen suggested well defined capsulated mass of measuring 94X86 mm noted in right lobe of liver with the vascularity on Color Doppler and hypoechoic areas 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(Photo - 2).

Serum alpha-fetoprotein (AFP) was high: >1000 ng/ml (normal values <20 ng/ml). Hepatitis B and C and HIV tests were negative. The patient's age, clinical-imagistic data, high value of alpha-fetoprotein and thrombocytosis raised the suspicion of a hepatoblastoma. However, the final diagnosis relied on the microscopic examination of the tumor biopsy therefore USG guided liver biopsy was taken.

The microscopic examination of the liver biopsy showed cells arranged in solid sheets and trabecular pattern. (Photo – 3) The cells had distinct cell membrane, uniform nuclei with minimal pleomorphism. There was presence of fetal pattern (Photo – 4) and foci showing rosset formation and tabeculae, having poorly defined cell borders and basophilic cytoplasm suggestive of embryonal pattern. (Photo – 5) Overall features were consistent with clinical diagnosis of hepatoblastoma.

Discussion

Out of all childhood tumors, excluding leukemia and lymphoma, hepatic tumors form about 0.5-2% in frequency. [5] The most common primary malignant liver neoplasm in children is hepatoblastoma. [1, 6] Two thirds of cases occur before 2 years of age and 90% of the cases are found below 5 years. Males are affected two times more compared to females. [7] Hepatoblastoma in adolescent and adults are worse off than in childhood because they are diagnosed late. [8, 9]

The triad of a hepatic tumor, thrombocytosis and high level of serum AFP in a child between 6 months and 3 years age is diagnostic of hepatoblastoma. [10] Our patient presented all these pathognomic associations. By Ultrasonography hepatoblastoma is seen as a hyperechoic, solid, intra hepatic mass. [11] Other investigations include CT, MRI, serum AFP and β Hcg levels.

High values of serum AFP can be detected in hepatoblastoma, [12] yolk sac tumor, hepatocarcinoma. Certain benign tumors like mesenchymal hamartoma, focal nodular hyperplasia and infantile hemangio-endothelioma may also elevate serum AFP levels. Certain histological variants of hepatoblastoma like small cell type do not produce AFP, grow rapidly and are resistant to chemotherapy. High values of AFP in hepatoblastoma may suggest massive tumoral extension, and /or presence of metastasis signifying an unfavourable prognosis.

Hepatoblastoma develops more frequently in the right hepatic lobe [13] and our patient also had involvement of right hepatic lobe. 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 favors the embryonic differentiation of the hepatoblastoma and its frequent localization in the right hepatic lobe. [14] Grossly, the hepatobalstoma is a yellowish white, large, solitary tumor, multi-nodular, with well defined margins, with areas of fibrosis, cystic change and necrosis. [15]

Photo.1 Large abdominal mass

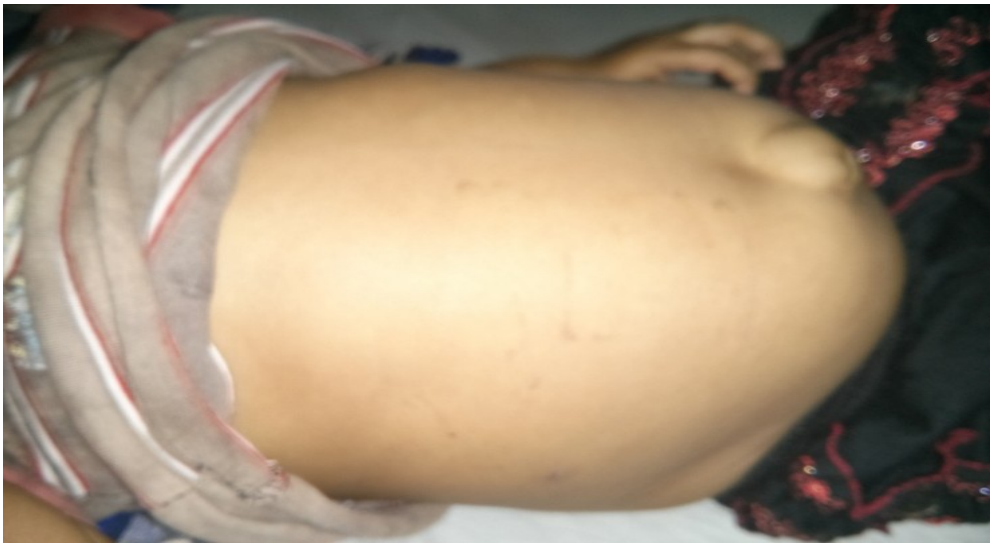


Photo.2 Abdominal computed tomography revealed a heterogeneous enhancing mass in right lobe of liver

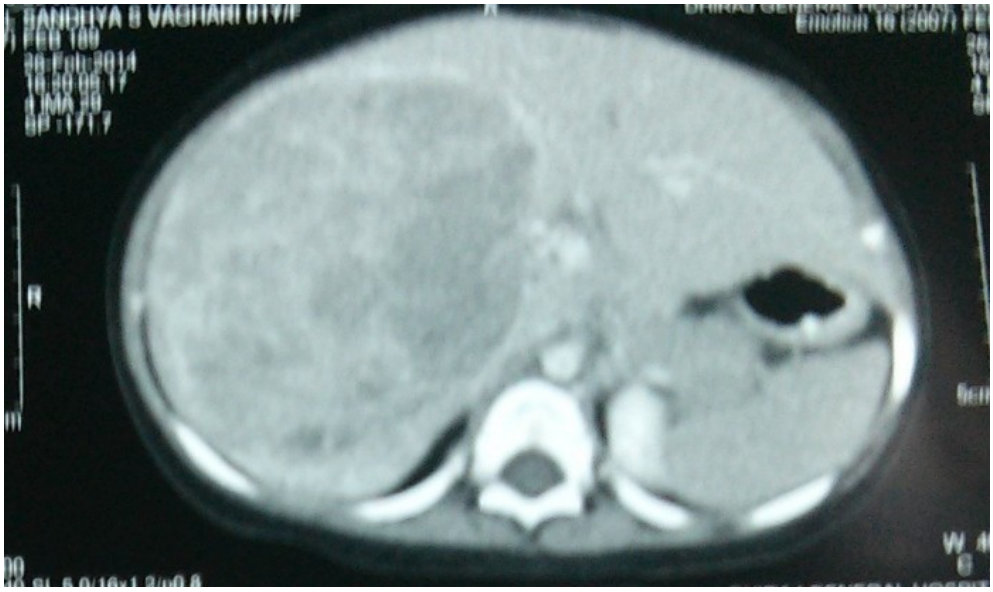


Photo.3 Scanner view of liver biopsy having light and dark areas with tumor cells arranged in solid sheets and trabecular pattern. [H & E, 10X]

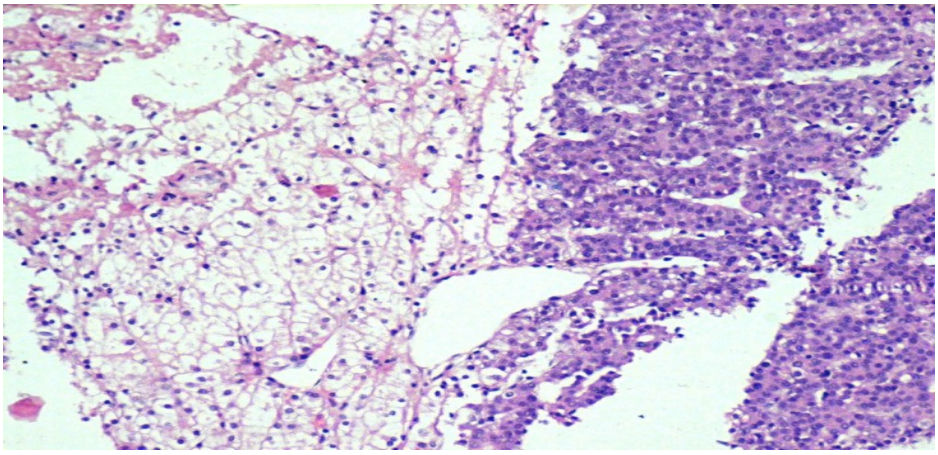


Photo.4 The tumor cells have distinct cell membrane, abundant clear vacuolated cytoplasm, uniform nuclei with minimal pleomorphism suggestive of fetal pattern. [H & E, 20X]

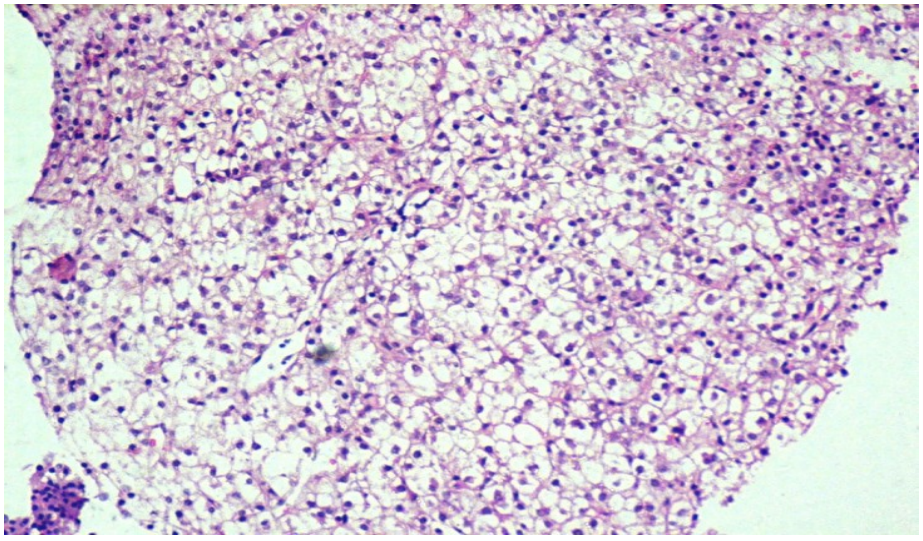
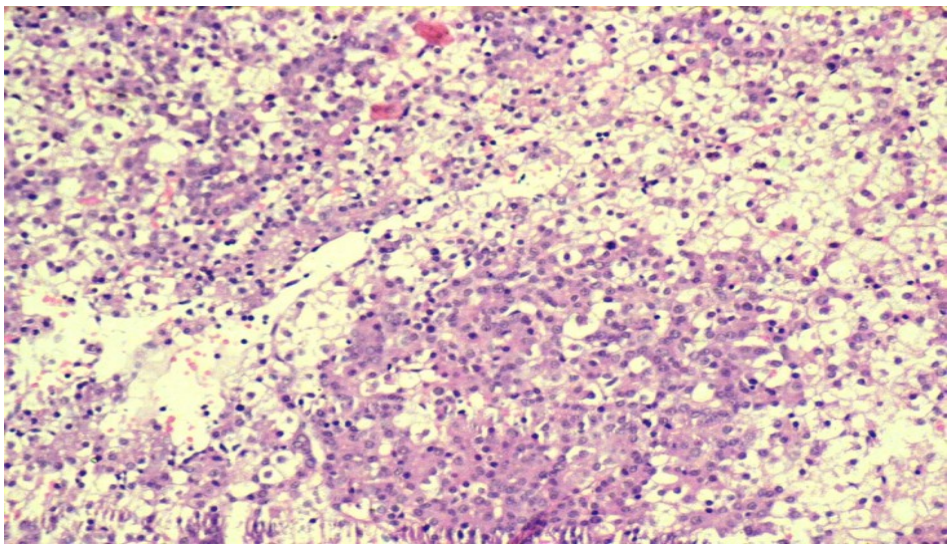


Photo.5 The foci of tumor cells shows rosset formation and tabeculae, having poorly defined

cell borders and basophilic cytoplasm suggestive of embryonal pattern. [H & E, 20X]



Microscopically, it presents with varying frequency and histology as under: [16]

Epithelial, Fetal, well differentiated (with minimal mitotic rate of ≤2 mitosis per 10 HPF, 40X) (7%) Epithelial, Fetal, mitotically active pattern (>2 mitoses per 10 HPF, 40X) (11%) Epithelial type, Fetal and Embryonal pattern only (39%)
Epithelial type, Macrotrabecular pattern (12%)
Small cell Undifferentiated pattern (5.6%)
Mixed Epithelial and Mesenchymal type without teratoid features (20%)
Mixed Epithelial and Mesenchymal type with teratoid features (4%)
Tumors which are purely Fetal, well differentiated lesions, mitotically inactive (of ≤2 mitosis per 10 HPF, 40X objective fields) are considered as favorable histopathologic feature, and when are in stage I, can be treated with surgery alone. Tumors with unfavourable histopathologic features belong to either small cell undifferentiated subtype or rhabdoid subtype or both. Thrombocytosis seen in hepatoblastoma is considered a paraneoplastic outcome [9, 17] due to excessive production of thrombopoietin by the neoplastic tissue.

Hepatoblastoma is sensitive to drugs like Doxorubicin, Cisplatin, Vincristine, 5-FU and Cyclophosphamide. [18] Chemotherapy helps in reducing the tumor size, makes them less prone to bleeding and delineates the tumor tissue and surrounding host parenchyma with vascular structures, so that surgical tumor resection is facilitated. Children with unresectable hepatoblastoma may undergo liver transplantation with significant success, with post transplant survival of 80%. [19] The present 5 years survival rate in children is 75%, which was 30% about 30 years ago. [6, 20] Monitoring AFP levels are a valuable means to determine pre-surgical chemotherapy, in the evaluation of post surgical excision results and for diagnosis of relapse or recurrence. Complete excision of the hepatoblastoma causes the serum AFP levels to become normal by 4-6 weeks.

Immunohistochemistry for hepatocyte antigen, α-fetoprotein, and, in some instances, ß-catenin may be useful in the

diagnosis of hepatoblastoma, mostly for tumors with favorable and unfavorable histologies. There is no immunostain to differentiate hepatocellular carcinoma from hepatoblastoma. Possible genetic markers (trisomies for chromosomes 2, 20 and 8, abnormalities of chromosome 1p) are being investigated and may help differentiate these two entities, but only in approximately 35% to 40% of hepatoblastomas that carry the abnormalities. [21]

The diagnosis of hepatoblastoma requires imaging studies in the form of Ultrasound (US), computed tomography (CT), serum AFP and β-HCG level, and the most useful diagnostic test microscopic examination of the tumor biopsy because histopathology is the gold standard for the final diagnosis.

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