Case Report

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Acute Presentation of Hepatoblastoma in A 9 Years Old Girl

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Abstract

Hepatoblastoma is the most common primary malignancy of the liver encountered in children. Primary tumors of the liver account for approximately 1% of malignancies in children. 50-60% of hepatic tumors in children are malignant, with >65% of these malignancies being hepatoblastomas. Hepatoblastoma occurs predominantly in children <3 yr of age. we present a 9 years old girl, who presented to our emergency ward with Jaundice and abdominal distension since 3 months. Examination showed pallor, icterus, hepatomegaly and ascites. Laboratory investigations revealed hyperbilirubinemia(TB:16.6, DB: 9) with thrombocytopenia (80000 cells / cumm). USG abdomen showed a mass lesion in right lobe of liver with irregular and nodular. CECT abdomen revealed a mass lesion of size 6.8x 5.5 x 5.9 cm in segment of right lobe with cirrhosis and metastatic nodules in the liver with moderate ascites and right pleural effusion. Spleen was moderately enlarged measuring 14 cm with infarcts. USG guided liver biopsy was done to confirm the diagnosis which showed histology pattern suggestive of Small Cell Undifferentiated Variant of Hepatoblastoma. Even a short period of history of jaundice and abdominal mass should rise the suspicion of hepatoblastoma, which help in early diagnosis and treatment.

Keywords: Hepatoblastoma; Jaundice; Thrombocytopenia.

Introduction

Hepatoblastoma is the most common primary malignancy of the liver encountered in children. [1] Primary tumors of the liver account for approximately 1% of malignancies in children. 50-60% of hepatic tumors in children are malignant, with >65% of these malignancies being hepatoblastomas. Rare hepatic malignancies include embryonal sarcoma, angiosarcoma, malignant germ cell tumor, rhabdomyosarcoma of the liver, and undifferentiated sarcoma. [2]

Hepatoblastoma occurs predominantly in children <3 yr of age. The etiology is unknown. Hepatoblastomas are associated with familial adenomatous polyposis. Etiology is thought to be alterations in the antigen-presenting cell (APC) / â-catenin pathway.[2,3,4] Low birthweight is associated with increased incidence of hepatoblastoma, with the risk increasing as birthweight decreases. Because of its rarity and inherent malignant nature, early diagnosis and treatment is difficult.

Case Report

9 years old girl, born to a 3rd degree consanguineous married couple, presented to our emergency ward with history of yellowish discoloration of eyes since 3 months, passing high colored urine since 3 months and progressive distension of abdomen since past 3 months. Child also presented with history of swelling of legs since 1 week with painful skin lesions, associated with itching. There was history of passing high colored urine. There was no history of passing clay colored stools, bleeding manifestations or convulsions. She had insignificant family history. For the above complaints child was treated by a quack with oral medications for 2 months.

On examination, vitals were stable and blood pressure was more than 95th centile. Anthropometry

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measurements revealed weight less than 10th centile and height between 25th to 50th centile. Pallor and icterus was present(Fig 1). Itchy, scaly skin lesions were seen in lower limbs up to knee (Fig 2). Pitting Pedal edema was present up to knee. In per abdomen examination, upper abdomen was distended with umbilicus pushed downwards and dilated veins were seen over abdomen(Fig 3). Shifting dullness was positive. Liver was palpable 3 cm below right costal margin and span was 12cm. liver was firm to hard in consistency There were no other signs of liver cell failure. Other systems were within normal limits.

Laboratory investigations are as follows: Hb: 10.6, TC: 9600, DC: N80 L19 E1, ESR: 30, platelets: 80000, total bilirubin: 16.6mg/dl, direct bilirubin : 9 mg/dl, SGOT- 330, SGPT: 171, ALP- 171, albumin- 2.4,RFT & Electrolytes: within normal limits, urine analysis: bile salt and bile pigments- present, HIV and Hepatitis B and C were negative. PT, APTT and INR were within normal limits. USG abdomen showed a mass lesion in right lobe of liver with irregular and nodular surface suggesting multiple heterogeneous nodules diffusely involving the liver. Further investigation with CECT abdomen revealed a mass lesion of size 6.8x 5.5 x 5.9 cm in segment of right lobe with cirrhosis and metastatic nodules in the liver with moderate ascites and right pleural effusion (Fig 4,5). Spleen was moderately enlarged measuring 14 cm with infarcts (Fig 6).

USG guided liver biopsy was done to confirm the diagnosis. The microscopic examination of the liver biopsy showed undifferentiated cells with minimal cytoplasm and round chromatic nuclei with inconspicuous nucleoli. Histology also revealed discrete small nests associated with minimal embryonal elements suggestive of Small Cell Undifferentiated Variant of Hepatoblastoma (Fig 7).

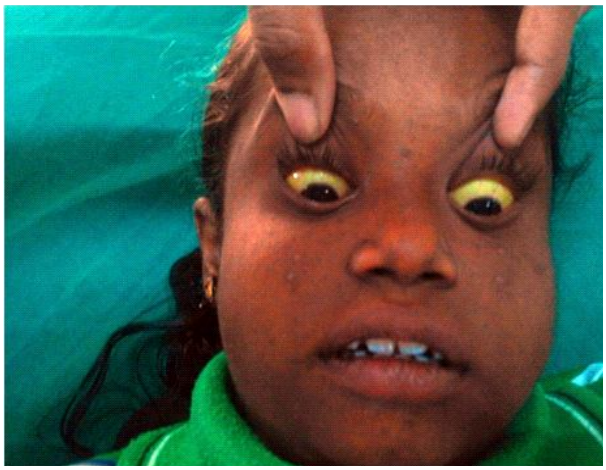

Fig. 1: Eyes showing deep icterus

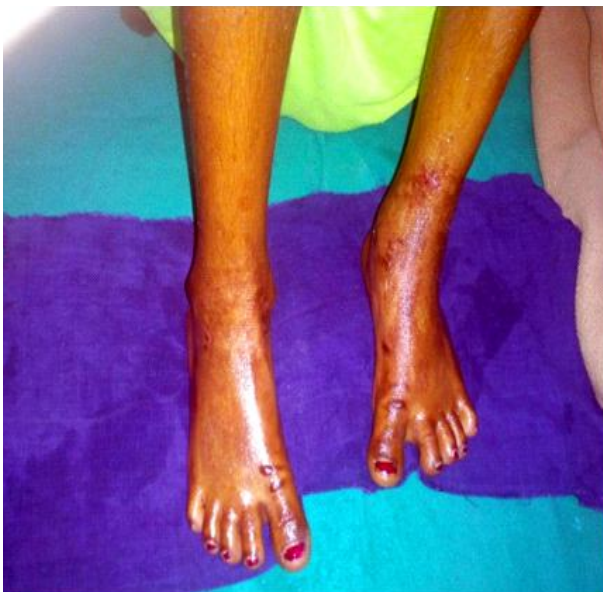

Fig. 2: Itchy scaly skin lesions with pedal edema

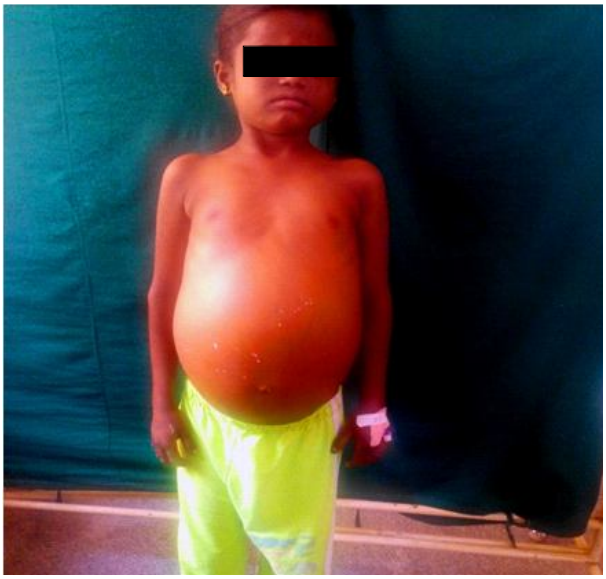

Fig. 3: Distended abdomen with umbilicus pushed down

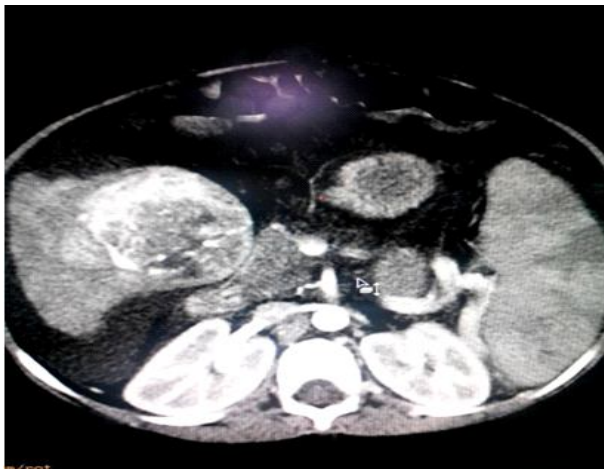

Fig. 4: CT scan showing mass lesion of size 6.8x 5.5 x 5.9 cm in segment of right lobe

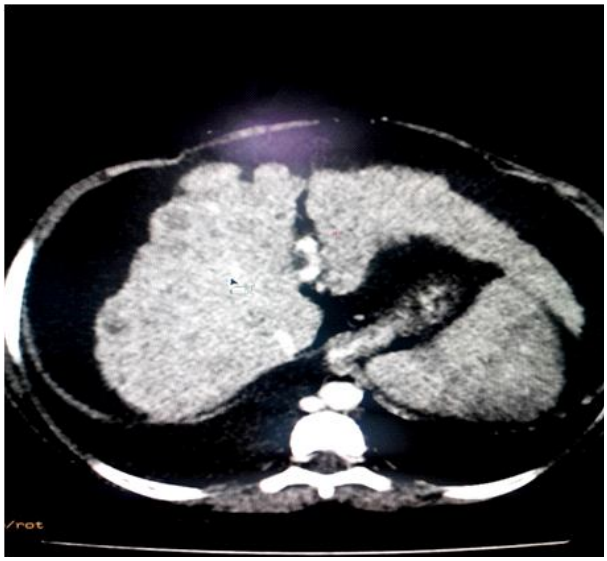

Fig. 5: CT scan showing metastatic nodules in liver

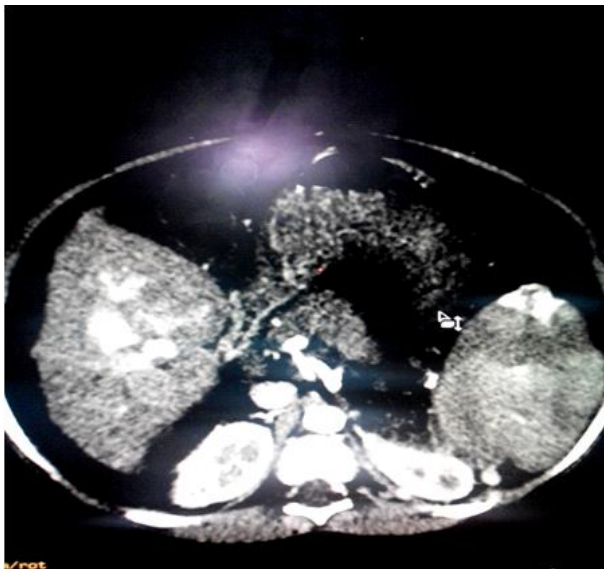

Fig. 6: CT scan showing splenomegaly with infarcts

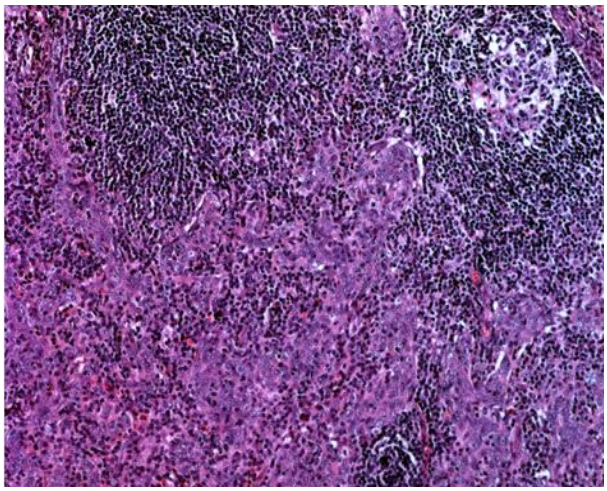

Fig. 7: Histology picture showing undifferentiated cells with minimal cytoplasm and round chromatic nuclei

Discussion

The most common primary malignant liver neoplasm in children is hepatoblastoma. Its incidence is approximately 1 to 2 in 1 million births. 2/3rds of cases occur before 2 years of age and 90% of the cases are found below 5 years [5]. Males are affected two times more compared to females. Hepatoblastoma in adolescent and adults are diagnosed late when compared to children and heralds poor prognosis. Our patient was also diagnosed as hepatoblastoma at 9 years of age which was over a short period and associated with metastasis.

There are several environmental risk factors associated with hepatoblasoma. Premature and very low birth weight have been found to be associated with the later appearance of hepatoblastoma [2].But our case did not have either of these risk factors.

A large number of congenital syndromes have been described in patients with hepatoblastoma, but only Edward's syndrome, familial adenomatous polyposis and Beckwith Wiede mann syndrome have been clearly shown to increase the risk of hepatoblastoma.

Most patients present with an enlarging abdominal mass. Our case presented with jaundice and distension of abdomen. The right lobe is involved three times more commonly than involvement seen in 20%-30%, and multicentric involvement in 15%. In our case also, lesion was seen in right lobe of the liver. Abdominal ultrasound is the first technique of choice as the initial diagnostic procedure for abdominal mass.

Magnetic resonance imaging (MRI) is the best modality for revealing morphologic details and to differentiate liver tumors. Computed tomography (CT) is better for detecting metastasis [6]. In our patient CECT was done which showed metastatic nodules in liver and infarcts in spleen.

Although thrombocytosis is commonly reported , low platelet counts can also occur in hepatoblastoma [6,7,8]. Our patient had thrombocytopenia with platelet count 80,000/cmm. Anemia is also described in previous studies [9].

Hepatoblastoma Mainly has 2 Variants

- The epithelial type, composed of small polygonal fetal cells or smaller embryonal cells forming acini, tubules, or papillary structures.
- The mixed epithelial and mesenchymal type, which contains foci of mesenchymal differentiation that may consist of primitive

mesenchyme, osteoid, cartilage, or striated muscle. The most common mesenchymal elements are osteoid and cartilage [5,10].

Epithelial hepatoblastoma is further broken down to pure fetal (31%), embryonal (19%), macrotrabecular (3%) and small-cell undifferentiated (3%). Our patient had small-cell undifferentiated type of hepatoblastoma which usually has bad prognosis. Our patient died within a week after the diagnosis was made. Death was due to hepatic encephalopathy. Our case is reported because of short duration history with acute presentation.

In general, the cure of malignant hepatic tumors in children depends on complete resection of the primary tumor. Even if 85% of the liver is resected hepatic regeneration is noted within 3-4 mo after surgery. Cisplatin in combination with vincristine and 5-fluorouracil or doxorubicin is effective treatment for hepatoblastoma and increases the chances of cure after complete surgical resection [2].

In low-stage tumors, survival rates >90% can be achieved with multimodal treatment, including surgery and adjuvant chemotherapy. With tumors unresectable at diagnosis, survival rates of approximately 60% can be obtained. Metastatic disease further reduces survival, but complete regression of disease often can be obtained with chemotherapy and surgical resection of the primary tumor and isolated pulmonary metastatic disease, resulting in survival rates of about 25%.

Liver transplant is a viable option for unresectable primary hepatic malignancies and results in good long-term survival [2,5].

Conclusion

Hepatoblastoma is the most common primary malignant tumour of liver seen in children and the main stay of treatment is complete surgical removal of the tumour mass which has a better prognosis when diagnosed early. Hence, even a short period of history of jaundice and abdominal mass should rise the suspicion of hepatoblastoma, which help in early diagnosis and treatment.

References

1. Raney B. Hepatoblastoma in children: A review. J Pediatr Hematol Oncol. 1997;19(5): 418-422.
2. Kliegman R, Stanton B, Geme St, Schor N, Behrman R. Nelson Text book of Pediatrics. 19th ed. United States of America. Saunders 2011
3. Herzog CE, Andrassy RJ, Eftekhari F. Childhood Cancers: Hepatoblastoma. The Oncologist 2000;5:445-53.
4. Atimati AO, Abiodun PO, Obaseki DE, Olubor OO. Hepatoblastoma in an adolescent girl: A case report. Niger J Paed 2014; 41 (4): 383 – 385.
5. Rathod G, Goswami S, Goyal R, Mehta S. Paediatric Hepatoblastoma in one year old female - A case report. Int J Curr Microbiol App Sci 2014;3(8):829-835.
6. Sharma N, Rathod G, Singla D. Hepatoblastoma of 12 years old female case report. Int Arch Integrated Med. 2015;2(6):223-26.
7. Nickerson HJ, Silberman TL, McDonald TP. Hepatoblastoma, thrombocytosis, and increased thrombopoietin C 1980; 45: 315-317.
8. Shafford EA, Pritchard J. Extreme thrombocytosis as a diagnostic clue to hepatoblastoma. Arch Dis Child, 1993;69: 171.
9. Kumar A, Poudel B. Hepatocellular Carcinoma presenting as a abdominal pain -A Case Report. J Biomed Sci 2013;2(1):1-3.
10. Kumar V, Abbas A, Fausto N, Aster J. Robbins and Cotran Pathologic Basis of Disease. 8th ed. United States of America. Saunders 2010.