

Case Report

Childhood Hepatoblastoma in two year old male- A case report

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Hepatoblastoma (HB) overall is a very rare tumor that affects the liver in infancy and childhood, it is the most common primary malignant tumor of the liver in that age . We reported a case of a 2-year old male child presented with pallor, abdominal distention; in physical examination there was a palpable large painless abdominal mass. CT scan revealed a solid hepatic tumor. His complete blood count shows thrombocytosis. Serum alpha feto- protein level was raised. A hepatic tumor in a child under the age of 3 years, associated with thrombocytosis and elevated serum alpha feto protein is very likely to be hepatoblastoma. The mainstay of treatment for this patient is surgical removal of the mass and then chemotherapy.

Keywords: Hepatoblastoma (HB); Abdominal mass; Thrombocytosis; Alpha feto protein(AFP); Child

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Introduction

During 2002–2008 the rate of HB in the United States Surveillance, Epidemiology and End Results (SEER) program data were 10.5 and 5.2 cases per million children <1 and 1–4 years, respectively, and was too infrequent thereafter to calculate incidence. An estimate of SEER data in 1999 suggested that about 100 cases of HB are diagnosed each year in the United States. However, as incidence of HB has risen roughly 4% per year between 1992 and 2004, faster than for any other childhood cancer, the current number of cases per year likely is higher(1) so Hepatoblastoma is a very rare cancerous tumor that starts in the liver yet it is the most common primary malignant hepatic tumor of infancy and childhood, occurring predominantly in the first three years of life(2).

This tumor accounting for just over 1% of pediatric cancers. The etiology is unknown; the pathogenetic mechanisms responsible for the development of HB remain unclear. HB, as an embryonal tumor, is derived from undifferentiated embryonal tissue. Rapidly growing HBs often are very sensitive to cytotoxic drugs and sometimes are very sensitive to radiotherapy. HB has been associated with Beckwith-Weidemann syndrome, familial adenomatous polyposis, and low birth weight(3). The primary treatment is surgical resection; neoadjuvant chemotherapy

particularly in unresectable tumors resulting in considerable preoperative tumor shrinkage and sometimes near total ablation of the tumor(4). Formerly a disease with a dreadful prognosis, children now have a 5-year survival rate of over 70%. This has been achieved by advances in surgical techniques and the use of chemotherapy(5).

Case report

A 2-year-old male patient presented with 1 month history of enlarging abdomen, noticed by the mother while bathing him. Initially painless, now complains of dull aching pain and occasional vomiting unrelated to food intake. There was no past medical history of jaundice, anorexia and the patient was up to date with his vaccination schedule. His temperature was 37.2 C (98.96 F), blood pressure was 90/50 mm Hg, pulse was 110/min and regular, and respirations were 30/min. His weight was 11kg(75 percentile) and the height was 83 cm(90 percentile). On general examination, patient was having good general status, pallor and anemia. On abdominal examination, the liver was palpable about 6cm below the costal margin (**figure 1**) and spleen was just palpable. Hematological investigation revealed moderate anemia (Hemoglobin-8g/dl; Hematocrit-28%), thrombocytosis (platelets-850,000/mm³), normal liver function tests (ALT

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13 Iu/I, AST 38Iu/I). USG abdomen suggested well defined mass noted in right lobe of the liver with the possibility of Hepatoblastoma. Further investigation in the form of CT scan (**figure 2**) confirmed the diagnosis of Hepatoblastoma affection the right lobe of the liver. Serum Alpha Feto Protein(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 data, high value of AFP 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. The cells had distinct cell membrane, uniform nuclei with minimal pleomorphism. There was presence of fetal pattern and foci showing roset formation and tabeculae, having poorly defined cell borders and basophilic cytoplasm suggestive of embryonal pattern. Overall features were consistent with clinical diagnosis of Hepatoblastoma. For this patient we did a Complete resection of the tumor (**figure 3**).



Figure 1: Abdominal examination of the patient demonstrate the large mass of the liver that can be palpated.

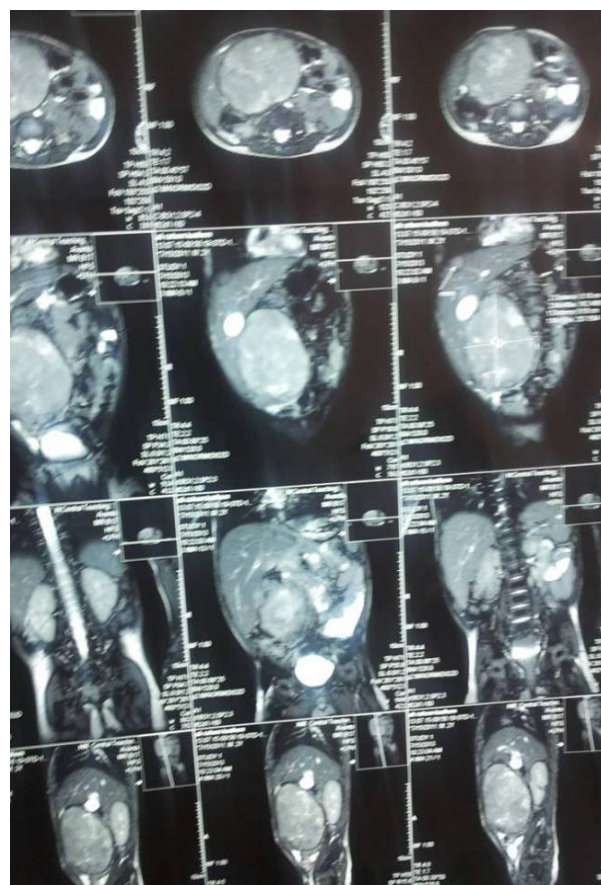


Figure 2: Abdominal computed tomography revealed the mass of the liver.

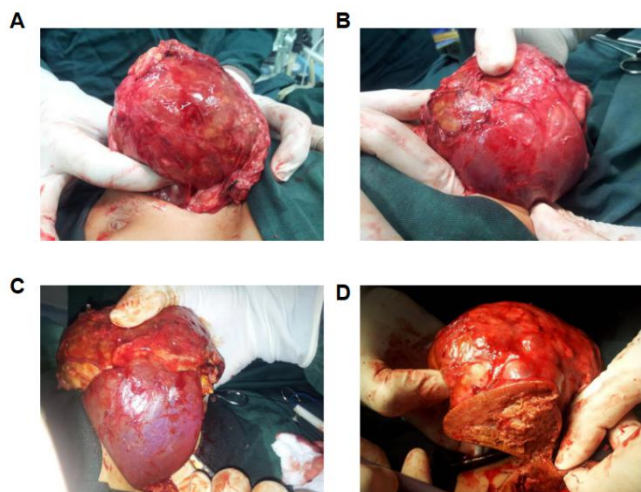


Figure 3 Intraoperative image represent the large liver tumor from our patient. **A.** represent the tumor (size 8.5cm x 10cm). **B.** represent the tumor (size 8.5cm x 10cm). **C.** represent the tumor attachment to the healthy liver(the tumor before the surgical resection). **D.** represent the beginning of the resection of the tumor after determining the margin of the tumor.

Discussions

The most common primary malignant liver neoplasm in children is hepatoblastoma(6). The median age of onset of hepatoblastoma is approximately 16 months with a slight male predominance. 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(7).

Serum level of Alpha Feto Protein(AFP) should be interpreted according to age, high values indicating hepatoblastoma . Moderately high values of blood serum fetoprotein are often detected in types of hepatoblastoma, tumor of yolk sac, hepatocarcinoma as well as in certain benign tumors (mesenchymal hamartoma, focal nodular hyperplasia and infantile hemangioendothelioma)(8).

The hepatoblastoma diagnosis is established according to clinical (young age), imaging, serum (very high AFP serum level) and histological criteria. Thrombocytosis, frequently seen in children with hepatoblastoma and other malignancies, is considered a paraneoplastic phenomenon. Thrombocytosis usually seen because of the excessive production of thrombopoietin at the level of the tumoral tissue. The human thrombopoietin, also called growth factor of the megakaryocytes, is secreted by hepatocytes, kidneys, spleen and stromal cells of the bone marrow. The level of the serum thrombopoietin stimulates the megakaryocytopoiesis and determines the increase of the thrombocytes number(9-11) . Immunohistochemistry for hepatocyte antigen, alpha fetoprotein, and, in some instances, β -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) may help differentiate these two entities, but only in approximately 35% to 40% of hepatoblastomas that carry the abnormalities(12).

The imaging work-up used at our institution begins with spiral CT for initial staging of the tumor and for assessing its resectability. It is also used to monitor tumor

response to preoperative chemotherapy and search for tumor recurrence.

only complete surgical excision (as in our patient) of the primary tumor was felt to correlate with cure. 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. If at initial surgical laparotomy the neoplasm seems resectable, safe attempt should be made to remove the tumor. If the tumor is deemed unresectable and the patient is at high risk for complications, a biopsy is taken and pre-resection chemotherapy is indicated(13-14).

Hepatoblastoma is taken into account to be non-surgical when: the tumour is extraordinarily giant, involving the danger of severe hemorrhage; both hepatic lobes are altered; the hepatic vein or the inferior vena cava is affected. The hepatic transplant is indicated either initially, for the non-excisable hepatoblastoma or after the relapse(15). Multicentric studies demonstrated a 10-year survival of 85 % in the case of children with a primary hepatic transplant and of 40 % in the case of those who needed a hepatic transplant after the relapse(16).

The AFP level monitoring represents a valuable marker of the response to the pre-surgical chemotherapy, in the evaluation of the excision result and for the precocious diagnosis of the hepatoblastoma relapse (17).

Conclusions

Hepatoblastoma(HB) is the most common malignant primary liver tumor in children. The primary treatment is surgical resection, and the use of pre-resection chemotherapy can increase the number of tumors that are resectable. The prognosis for patients with resectable tumor is fairly good in combination with chemotherapy. The outcome for those with non-resectable or recurrent disease remains poor.

Acknowledgement

None

Conflict of Interest

None

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