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Case Report

Hepatoblastoma of 12 years old female - A case report

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Abstract

Hepatoblastoma is the most common primary liver tumor of childhood and after neuroblastoma and nephroblastoma the third most common abdominal neoplasm in this age group. Hepatoblastoma in adolescent and adults are worse off than in childhood because they are diagnosed late. The primary treatment is surgical resection, and the use of preresection chemotherapy can increase the number of tumors that are resectable. The prognosis for patients with resectable tumors is fairly good in combination with chemotherapy.

Key words

Hepatoblastoma, Abdominal neoplasm, Surgical resection, Chemotherapy.

Introduction

Hepatoblastoma is the most common primary liver tumor of childhood and after neuroblastoma and nephroblastoma the third most common abdominal neoplasm in this age group. Most of these tumors are seen before 5 years of age [1]. Surgery remains the primary means of curative therapy, but the role of chemotherapy in both the adjuvant and neoadjuvant setting has become increasingly important over the past three decades. New insight has also been gained into the molecular biology of hepatoblastoma. Here we are presenting a case where we are able to find and document the typical features of hepatoblastoma in 12 year old girl patient with presence of metastasis.

Case report

A 12 year old female patient presented with abdominal mass. 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

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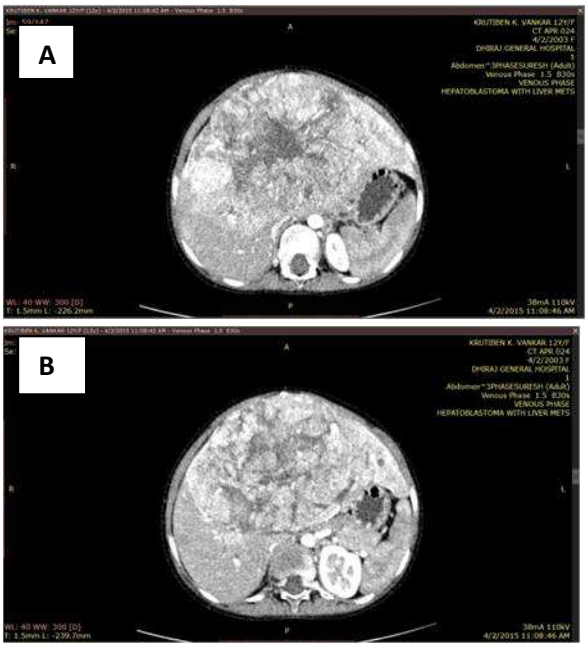
had pallor but had good appetite and normal bowel functions. On abdominal examination, the liver was palpable, which is 6 cm below the costal margins and spleen was just palpable. Hematological investigation revealed moderate anemia (Hemoglobin - 9.2 g/dl), thrombocytopenia (platelets – 70,000/mm^3), normal liver function tests (ALAT 14 Iu/l, ASAT 35 Iu/l) and positive sickling test. USG abdomen suggested well defined capsulated mass of measuring 12X11×11cm noted in right lobe of liver with central hypoechoic area and necrosis. There was another lesion of size 3.5× 3.1 cm with hypoechoioc ring noted in the right lobe of liver lateral to the main lesion possibility of secondary metastatic lesion. There was 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 with metastasis in right and left lobe of liver. **(Photo – 1A, 1B)** Serum alpha-fetoprotein (AFP) was low: >35 ng/ml (normal values <20 ng/ml). Hepatitis B and C and HIV tests were negative. Patient was referred to the cancer institute for further workup and treatment.

Discussion

The most common primary malignant liver neoplasm in children is hepatoblastoma [2, 3]. 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 [4]. Hepatoblastoma in adolescent and adults are worse off than in childhood because they are diagnosed late [5, 6]. Our patient was also diagnosed as hepatoblastoma at 12 year of age which was quite late and associated with multiple metastasis.

There are several environmental risk factors associated with hepatoblasoma. Premature birth and very low birth weight have been found to be associated with the later appearance of hepatoblastoma [7-10].

Photo – 1A, 1B: CT scan with hepatoblastoma affecting right and left lobe of liver with metastasis foci.



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 [7].

Most patients present with an enlarging abdominal mass. The right lobe is involved three times more commonly than the left, with bilobar involvement seen in 20%-30%, and multicentric involvement in 15% [11, 3]. The theory behind the involvement of the right lobe is that left lobe derives oxygenated blood totally from the umbilical vein. The right lobe derives blood from portal vein with lower oxygen concentration. In context to this theory our patient also had involvement of right lobe of liver with metastatic foci in left lobe of liver.

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Abdominal ultrasound is the first technique of choice as the initial diagnostic procedure for abdominal mass. Magnetic resonance imaging (MRI), in many centers is the best modality for revealing morphologic details and to differentiate liver tumors. Computed tomography (CT) is essential for the evaluation of pulmonary metastasis [7].Hepatoblastoma usually appears as a focal or multifocal solid tumor. Stippled or chunky calcifications can be detected in 40%-50% of patients, which is significantly higher than in patients with benign lesions such as hemangiomas and hemangioendotheliomas [12, 13].

Anemia and platelet abnormalities have been reported [3]. Although low platelet counts can occur in hepatoblastoma, thrombocytosis is commonly reported [14, 15]. Our patient also had thrombocytopenia with platelet count 70,000/cmm.

AFP is the most important marker for hepatoblastoma which is increased in 90% of patients. Low AFP levels (<100 ng/mL) are aggressive and associated with a poor prognosis [7]. Our patient also having low level of AFP levels with presence of metastatic foci in the right lobe of liver. In neonates the interpretation of AFP measurements is more difficult because of the naturally high serum levels.

Hepatoblastoma is classified by histology as epithelial (56%) or mixed epithelial/mesenchymal (44%) [16]. Epithelial hepatoblastoma is further broken down to pure fetal (31%), embryonal (19%), macrotrabecular (3%) and small cell Undifferentiated.

Cytogenetic analysis performed on hepatoblastomas investigators have found several alterations with chromosomal gains occurring more frequently than losses. The most common alterations are trisomy 2, 8, 20 [17]. The implementation of whole-genome DNA chipbased technologies led to the detection of many altered genomic regions, of which gain of material on chromosome 2q 13-22, 2q 36-37 and deletions of 2p and 4q were associated with advanced tumors and poor prognosis [17, 18].

Complete surgical excision 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 [19].

Conclusion

Hepatoblastoma is the most common primary liver tumor in children. The primary treatment is surgical resection, and the use of preresection chemotherapy can increase the number of tumors that are resectable. The prognosis for patients with resectable tumors is fairly good in combination with chemotherapy. However, the outcome for those with nonresectable or recurrent disease remains poor and new therapies are needed.

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