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Role of Imaging in the Diagnosis of Hepatoblastoma: A Case Study

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Abstract
In children below 5 years, most common malignant tumour is hepatoblastoma. A male child of one-year-age presented with a mass in the abdomen in our case and there is significant abdominal distension. Underwent USG abdomen and CECT thorax and abdomen and CT abdomen angiography which revealed a solid mass lesion in the liver that is a hepatic tumour. There is increase in AFP and serum βHCG. Thus, the diagnosis of hepatoblastoma made. CECT chest revealed multiple metastasis to both lungs and CECT abdomen revealed the mass lesion situated at the junction of right and left lobe of liver with mild heterogeneous enhancement in the hypoattenuating mass. CT abdominal angiography does not reveal any spoke-wheel pattern but suggestive of tumour nodularity with lobulation with small hypoechoic & anechoic areas suggesting hemorrhage or necrosis.

Keywords: Child, USG, CECT Abdomen, Abdominal Angiography, CECT Chest, Hepatoblastoma

Introduction

Hepatoblastoma is the tumour which is seen before 5 years of age. About two-thirds of malignant liver tumours are due to hepatoblastoma [3]. Amongest all primary malignant tumours in children, hepatoblastoma accounts for 1–4% of all cases. Patients most commonly present with abdominal distension and elevated AFP. Hepatoblastoma affects young children & mostly diagnosed in the first 3 years of life [1]. It is seen commonly in children born prematurely with very low birth weight [2]. It is\associated with familial adenomatous polyposis, trisomy 18 & other growth syndromes.

Because of its rarity & inherent malignant nature, diagnosis & treatment is problematic. The typical age, presentation, history, clinical findings of hepatic tumours along with imaging findings of USG, CECT Abdomen & elevated AFP are indicative of hepatoblastoma. Small cystic spaces & echogenic calcification can be present in hepatoblastoma.

Case Report

Our case is a one-year-old male child admitted with abdominal distension for 20 days more localized over right hypochondrium region and epigastric region associated with visible veins in the abdomen noted in less 4-5 days after admission. Patient was having fever for fifteen days(101°F). Child was a febrile for 3 days prior to USG which was relieved with medication. Child was born term had normal vaginal delivery and cried immediately after birth with birth weight 2.5 kg with no history of seizure, hypoglycemia, cyanosis, no history of change in bowl habits, no use of any medication. Immunization done as per age.

On examination patient was having pallor(HB 6.5gm/dl), hyperpigmentation on lips. No icterus, cyanosis, clubbing or lymphadenopathy. But in addition to pallor Patient had thrombocytosis (platelets 7.1 L), TLC 7800 cells, CRP was high, rest of the hematocrit were normal.

On abdominal examination Liver found massively enlarged 10 cm below costal margin, Left lobe of liver is also found enlarged 4-5cm in epigastric region and spleen was normal. Liver function tests showed increased ALAT, ASAT and alkaline phosphatase, KFt test was normal.

Ultrasound abdomen done soon after admission of the child which showed liver massively enlarged with a multilobulated heterogeneous solid lesion involving both lobe of liver with no increase in vascularity (Fig 1-4).

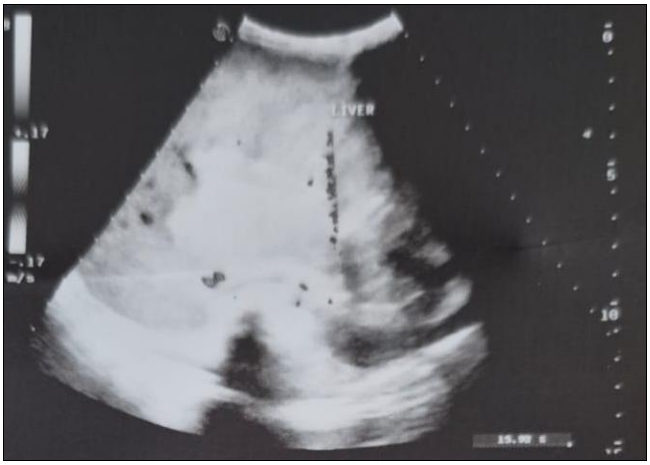


Fig 1: USG showing Lobulated mass lesion

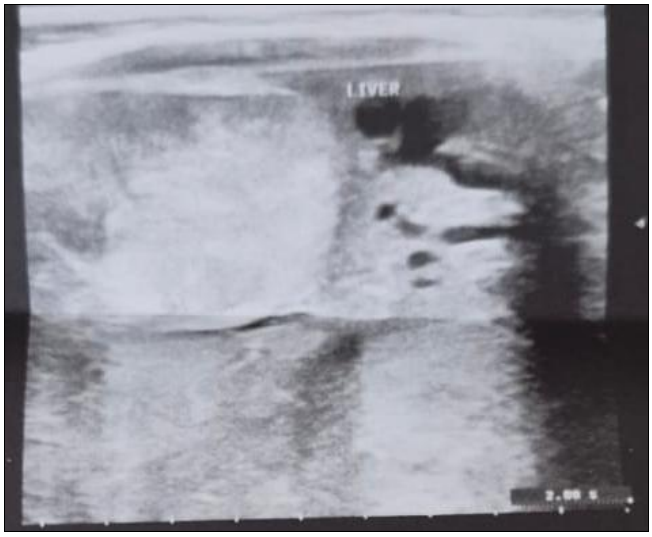


Fig 2: Lobulated Mass lesion at the junction of right and left lobe of liver

No calcification within the lesion found, USG diagnosis is hepatoblastoma mode. Liver biopsy was advised and also alpha fetoprotein estimation done which was (4.20000 ng/ml)done. Multiple rounded hyperdense lesions throughout both lung fields showed metastasis noted (Fig 5).

CECT Abdominal: Abdominal contrast enhanced CT showed massive liver enlargement with lobulated lesion. The tumour enhances unevenly i.e. heterogeneous enhancement. In arterial phase the lesion shows nodular enhancement which is a prominent feature. In portal phase, the lesion remains hypoattenuating compared to normal liver parenchyma.

CECT Abdominal Angiography: CT angiography sometimes describes a spoke-wheel pattern which is not found in our case. Tumour nodularity or lobulation was found in sonography, also noted in CECT abdomen. Small hypoechoic & anechoic areas noted within the tumour suggestive of hemorrhage or necrosis. Few necrosis areas seen within it Lymph node enlargement is not very significant. Serum Alpha fetoprotein was very high >420000ng/ml, Hepatitis test was negative. On the basis of multiple bilateral metastatic lesions to lungs by CT, stage IV disease is diagnosed and treatment accordingly started. Hepatoblastoma in our case is confirmed by imaging studies (USG abdomen, CECT chest & abdomen) and increased AFP & thrombocytosis.

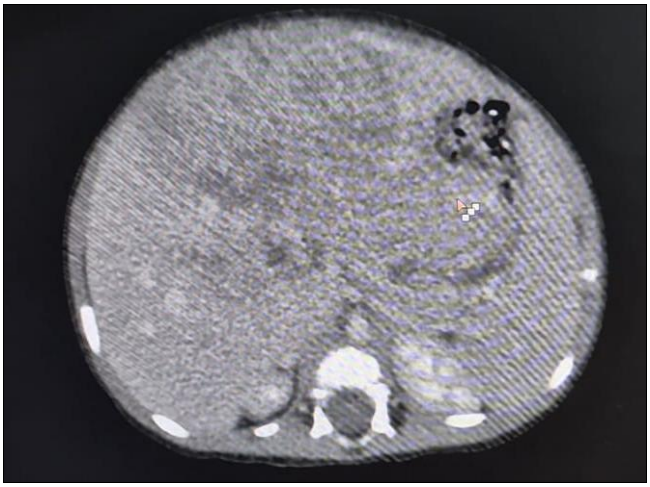


Fig 3: Nodularity and lobulation of the mass of liver

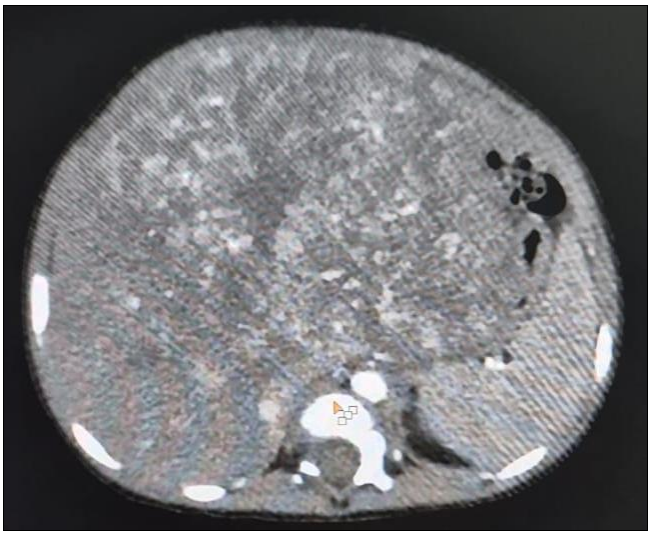


Fig 4: Heterogeneous enhancement of the liver mass

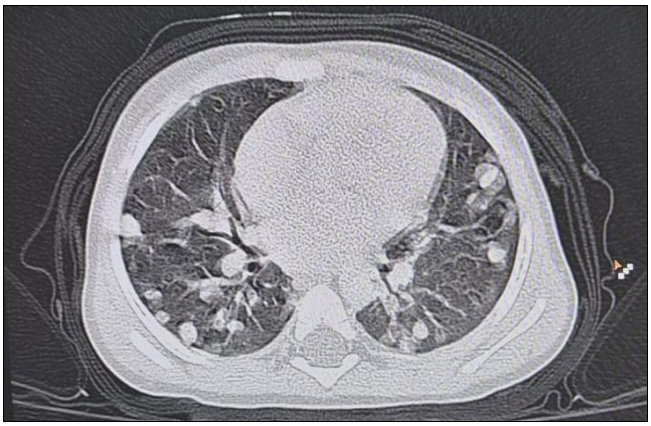


Fig 5: Multiple metastasis to lungs

Discussion

Hepatoblastoma affects children in the first 3 years of life [1]. It is more common in children born prematurely with very low birth weight [2]. The most common primary malignant neoplasm in children is hepatoblastoma [3, 4]. Males are affected two times more compared to females [5].

Hepatoblastoma is the most common primary malignant hepatic tumor of childhood, affecting 1 per 1.2 million children [6]. It is associated with familial adenomatous polyposis, Trisomy 18, and overgrowth syndromes [7].

Hepatoblastoma in adolescents and adults are worse than in childhood as they are diagnosed late [8, 9].

The triad of a hepatic tumor, thrombocytosis, and high levels of serum AFP in a child between 6 months & 3 years of age is diagnostic of hepatoblastoma [10].

Ultrasonography (USG) is seen as hyperechoic, solid intrahepatic mass [11]. Abdominal USG is the initial imaging modality performed in a child with an abdominal mass [6]. On USG, hepatoblastoma appears as a heterogeneous mass that is hypoechoic to the background liver. Small anechoic areas and echogenic calcifications can be present [6]. On USG hypoechoic and anechoic areas within the tumor represent hemorrhage and necrosis.

Color Doppler is used to evaluate hepatic vasculature, especially to identify hepatic veins, IVC, and PV, and determine their patency [12].

Other investigations being done are CT, CT Angiography MRI, serum AFP, and BHCG levels.

Hepatoblastoma develops more frequently in the right hepatic lobe [13]. MRI with hepatobiliary contrast agent is done as the next imaging modality after USG [14]. But because of easy availability of CT, contrast-enhanced CT (CECT) abdomen often done with CT abdominal angiography: CECT is performed in hepatic arterial and portal venous phases.

Non-contrast imaging and delayed phase imaging should not be performed [12]. On CT, the tumor is heterogeneous and typically enhances less than the background liver. Tumor calcification may be present [12]. Chest CT is recommended as lung metastasis are present in 20% at diagnosis [12].

CT abdominal angiography shows – ill defined, very very minimally heterogeneous nodular enhancement noted in the arterial phase, and in the portal phase, lesion remains hypoattenuating compared to normal liver parenchyma.

Thrombocytosis seen in hepatoblastoma is considered a paraneoplastic outcome [9, 15] due to excessive production of thrombopoietin by neoplastic tissue.

Surgical resection is recommended for hepatoblastoma and it is combined with neoadjuvant chemotherapy drugs such as doxorubicin, cisplatin, vincristine, and 5-fluorouracil [16].

Liver transplant is reserved for unresectable hepatoblastoma. These operations often have very successful outcomes [17].

Monitoring AFP levels is a valuable means to determine presurgical chemotherapy, in the evaluation of post-surgical excision results, and for diagnosis of relapse or recurrence. Complete excision of the hepatoblastoma causes serum AFP levels to become normal within 4–6 weeks.

Conclusion

Hepatoblastoma is the most common primary hepatic malignancy of childhood. The diagnosis of hepatoblastoma requires imaging studies such as USG, CT, MRI, serum AFP and βHCG levels.

Survival rates have been improved over the past several decades. The present 5-year survival rate in children is 75%, which was about 30% around 30 years ago.

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