Case Report

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A rare case of adult hepatoblastoma mimicking hepatocellular carcinoma- case report and review of literature

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ABSTRACT

Hepatoblastoma (HB) is a rare malignant primary tumour of the liver and occurs mostly in the pediatric group within the first 3 years of life. It is extremely unusual to find hepatoblastoma in adults and is a very rare cause of primary malignant liver tumour in adults and due to this patient may be diagnosed at late stages of the disease at leading to poor prognosis in this group. Reported here a case of a 20-year-old boy with a large liver mass with abdominal pain. Triphasic CT revealed the presence of a large heterogenous tumor in the right lobe of liver suggestive of hepatocellular carcinoma. Patient underwent a right hepatectomy. Final histopathology was reported as Epithelial type hepatoblastoma. The patient had an uneventful post-operative recovery. We present this case for its rarity and ability to masquerade other primary liver tumors in the adult age as seen in our patient.

Keywords: Adult hepatoblastoma, Chemotherapy, Surgical resection

INTRODUCTION

Hepatoblastoma (HB) is a rare malignant tumor of the liver, encountered in the pediatric population with most cases occurring before the age of 3. It accounts about 1-4% of all primary malignancy in children. Most of these tumors arise in the embryo, hence it is unusual for hepatoblastoma to occur in adult hence only rarely encountered in adult patients. Around 40 adult cases of adult HB have been reported in English literature and very few from India. 5-7

The diagnosis is often overlooked, and patients present at the late stages of the disease contributing to the poor prognosis in this age group.

CASE REPORT

A 20-year-old male presented to us with complaints of right upper quadrant pain of 3 months' duration, dull

aching and intermittent. There were no other associated symptoms. General physical examination was unremarkable without any evidence of generalized lymphadenopathy. Abdominal examination revealed presence of hepatomegaly. Other system examination was within normal limits.

Routine blood investigations did not reveal any abnormality. Transabdominal ultrasound picked up a large mass in the right lobe of liver involving segments 5,6,7,8 with areas of hemorrhage within it. A triphasic CECT abdomen was done to characterize the lesion which revealed a 16 x 13 cms well encapsulated mass in right lobe of liver with areas of hemorrhage within it, displacing the middle hepatic vein and enhancement in arterial phase and slow washout in portovenous phase suggestive of hepatocellular carcinoma. AFP values were within normal limits. In view of CECT findings and normal AFP values the patient was planned for a right hepatectomy.

Intraoperatively the lesion was involving segments 5,6,7,8 splaying the MHV laterally without any evidence of extrahepatic spread. Hence a right hepatectomy was performed using CUSA. Post-operative period was uneventful and the patient was discharged on POD 10. The final histopathology was reported as epithelial type Adult hepatoblastoma. He received adjuvant chemotherapy in the form of cisplatin, vincristine and 5-flurouracil and is asymptomatic at 1 year of follow up without any evidence of locoregional or systemic recurrence.

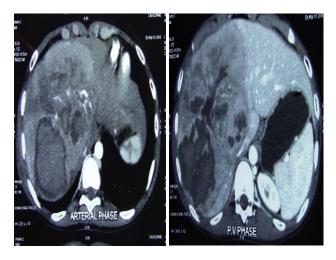


Figure 1: Triphasic CT showing a large mass in the right lobe of liver with enhancement in arterial phase and slow washout in portovenous phase.



Figure 2: Right hepatectomy specimen.

DISCUSSION

HB is the most common primary malignant liver tumor in the pediatric age group. 1,2 Around 90% of the cases occur in patients under 5 years of age and two thirds of the cases occur in the first 3 years of life. 3,4 Incidence is twice in males compared to females. HB in young adults is extremely rare and the prognosis is poorer than in children, as they are usually diagnosed late and hepatoblastoma is not commonly considered as a common differential diagnosis in adults. 6,7

The etiology of HB is not known clearly. Cytogenetic and molecular genetic abnormalities analysis have pointed towards involvement of chromosomal loci on 1q, 2q, 4q, 8q.8 Loss of heterozygosity imprinting at locus 11p 15.5 also has been implicated in the possible pathogenesis of HB.8 Nuclear p53 accumulation in tumor cells indicates that p53 mutation is also involved in its molecular pathogenesis.9

Individuals with familial adenomatous polyposis (FAP), frequently develop hepatoblastomas. ^{10,11} Also betacatenin mutations have been shown to be common in sporadic hepatoblastomas, occurring in as many as 67% of patients. ^{12,13}

Ishak and Glunz classified hepatoblastoma into two groups: an epithelial type, and a mixed epithelial and mesenchymal type. ¹⁴ The epithelial type consists of fetal and embryonic cells.

The most frequent symptoms of hepatoblastoma are abdominal pain, and rarely vomiting. Most frequent physical signs are hepatomegaly and weight loss.

Imaging

The initial diagnosis of HB based on imaging. Ultrasound is a non-invasive method particularly useful in infants. HB is seen as a hyperechoic, solid, intrahepatic mass on USG. 15 More specific imagings include triphasic contrast enhanced computed tomography (CT), magnetic resonance imaging (MRI), serum AFP. Kishimoto et al. mentioned that intratumoral calcification demonstrated by CECT was helpful in the diagnosis of adult HB. 16 Alpha-fetoprotein (AFP) levels are commonly elevated, but when AFP is very high (>100000) or low (<100), the prognosis is poor.

Treatment

Surgical resection is the mainstay of treatment for patients with HB and margin negative reaction is associated with better survival rates. Improvements in survival in recent years have been a function of standardized chemotherapy regimens that reduce tumor size and enable complete tumor excision, even allowing cure in the presence of initially unresectable or metastatic disease. The Chemotherapy has been proven effective in both an adjuvant and neoadjuvant treatment and can cause tumour shrinkage. It makes them less prone to bleed and delineates the tumor from the surrounding normal parenchyma and vascular structures so as to facilitate the resections. Chemotherapeutic drugs used in HB are doxorubicin, cisplatin, vincristine, 5-FU and cyclophosphamide. The control of the patients of the

Furthermore, liver transplantation has recently been associated with significant success in the treatment of children with unresectable hepatoblastomas. Post-transplant survival rates as high as 80% have been

reported for children with HB.¹⁹ The prognosis in adults is much poorer, the median survival being 6 months. Because of relative lack of experience of HB in adult patients, it is important to institute multimodality treatment in such cases to achieve a better outcome.

Because of its rarity and non-specific initial symptoms, HB in the adult presents a diagnostic challenge, requiring a high index of suspicion for correct diagnosis nd treatment. Because the prognosis could be improved with early detection, it is important for clinicians to list this disease in the differential diagnosis of adult hepatic masses.

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