CASE REPORT

A rare case report of extrahepatic hepatoblastoma in a child

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Abstract

Hepatic malignancies account for 1-4% of all childhood solid tumours and Hepatoblastoma is the most common malignant liver tumour in children. Its extrahepatic origin is rare. Here we present the case of a three-year-old boy who came with a large non-tender mass in the right upper abdomen for six months. Ultrasound abdomen revealed a huge heterogenous mass anterior to the right kidney and inferior to the liver with internal vascularity and calcifications, mimicking a neuroblastoma. Tru-cut needle biopsy showed foetal-type hepatoblastoma. After neoadjuvant chemotherapy, the tumour was explored. It was found to be adherent to the inferior surface of the liver with no capsular breech. Hence differentiating it from exophytic growth of hepatoblastoma. The tumour was completely resected. The postoperative course was uneventful and adjuvant chemotherapy was given. So far only a few cases of extrahepatic hepatoblastoma have been reported

Keywords: Extrahepatic, hepatoblastoma, suprarenal, tumor, excision..

DOI: 10.47391/JPMA.6895

Submission completion date: 10-05-2022
Acceptance date: 20-10-2022

Introduction

Hepatic malignancies account for 1-4% of all childhood solid tumours and Hepatoblastoma accounts for 80% of these malignant liver tumours[1]. It is the most common primary malignant tumour of the liver, affecting children younger than three years[2]. Nearly all hepatoblastomas arise from the liver. Extrahepatic origin of hepatoblastoma is very rare. We report the case of an extrahepatic hepatoblastoma in a three-year-old boy, which radiologically was labelled as neuroblastoma or less likely a teratoma, but biopsy revealed its definitive diagnosis.

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Case Report

A three-year-old boy presented in the paediatric surgery department, Mayo Hospital/ King Edward Medical University, Lahore in April 2021, with the complaints of right upper abdominal pain and fever on and off for the past six months. There were no other associated symptoms. He had normal vital signs and weighed 12 kilograms. Clinical examination revealed a large non-tender hard fixed mass of 12 x 10 cm palpable in the right hypochondrium. The rest of the systemic examination was unremarkable.

Blood biochemistry was within the normal range. Serum alpha-fetoprotein (AFP) level was more than 1210 ng/ml (normal levels <10ng/ml).

Computed tomography (CT) of the abdomen with intravenous contrast showed a huge mass with heterogeneous enhancement, internal vascularity, and specks of calcification measuring 11.4 x 11.4 x 9.4 cm and located in the right hemiabdomen anterior to the right kidney and inferior to the liver (Fig.1A). The liver was of normal size and appearance. The rest of the CT examination was unremarkable.

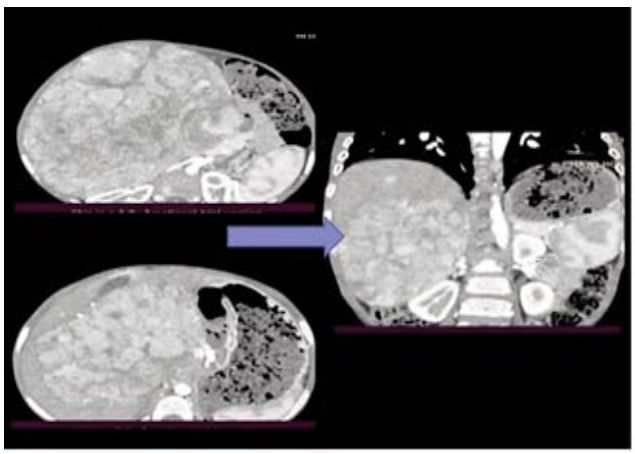

Figure-1A: Abdominal Computed Tomography (CT) showing a large heterogeneously enhancing mass in the right suprarenal region.

Ultrasound-guided tru-cut biopsy of the mass showed hepatocellular parenchyma with normal morphology and cells of pure foetal epithelial type differentiation. There was no evidence of distant metastasis on further workup. Neoadjuvant chemotherapy was planned, and he

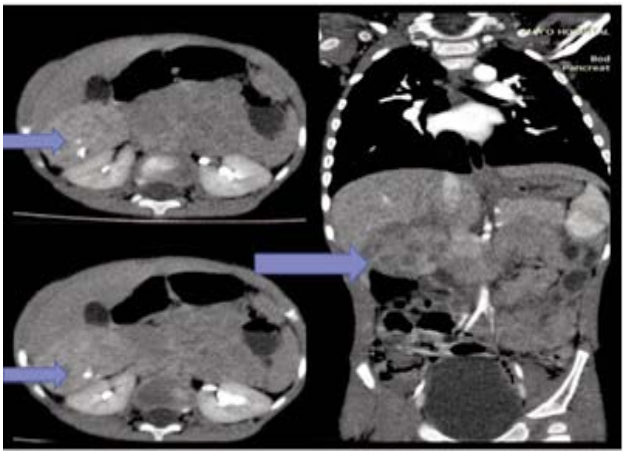

Figure-1B: Post chemotherapy abdominal Computed Tomography (CT) showing 70% reduction in tumor size, with the arrow showing infrahepatic location of the tumor

received six cycles of chemotherapy (cisplatin monotherapy). Post chemotherapy contrast-enhanced abdominal computed tomography (CT) showed a 70% reduction in tumour size (Fig.1B).

The plan of surgical resection was made under general anaesthesia after informed consent. Per-operatively, there was a 4 x 4cm mass in the right suprarenal area which was separate from the right kidney, with no involvement of major vessels and adherent to the undersurface of segment V and VI of the liver, with intact liver capsule (Fig.2A-a). Complete surgical resection of the mass was done (Fig. 2A-b).

Post-operative recovery was uneventful. Level of Serum alpha-fetoprotein (AFP) dropped to 6ng/ml. The patient was referred to an oncologist for adjuvant chemotherapy. Histopathological analysis of the tumour showed

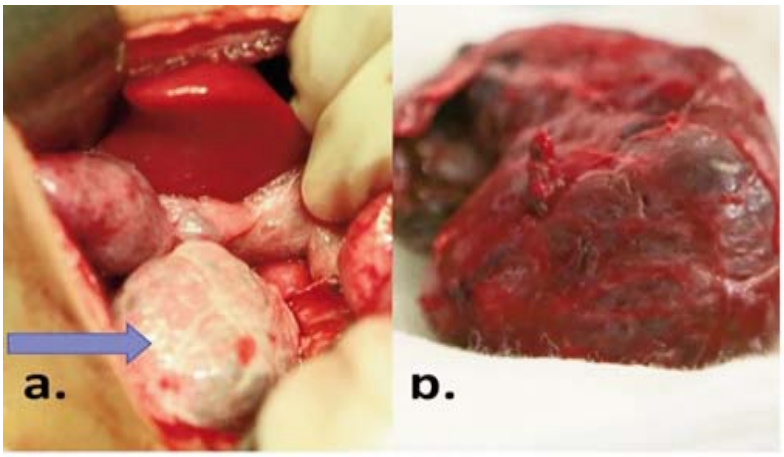

Figure-2A: Operative picture showing extrahepatic, suprarenal location of the tumor; cut specimen of the excised tumor

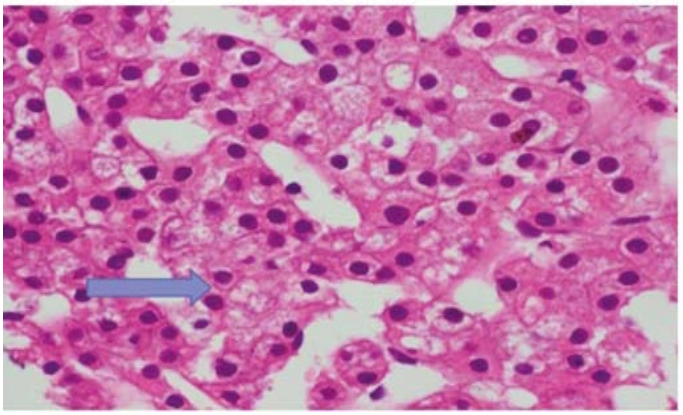

Figure-2B: Hepatocellular parenchyma with cells showing foetal epithelial type differentiation

hepatoblastoma of mixed type (foetal and mesenchymal) with 80-90% viable tumour. The tumour markers and cells were positive for Heparl and Glypican 3. The membranous and nuclear expression was positive for beta-catenin and negative for CK7 and INI-1 (Fig.2B). Consent for publication was taken from parents, head of department, and institution

Discussion

The extrahepatic location of hepatoblastoma has been rarely reported. In our case differentials included neuroblastoma, or teratoma. Based on imaging and biopsy, exophytic growth of hepatoblastoma was diagnosed.

Most cases of hepatoblastoma are sporadic and may arise from the liver as unifocal or bilobular lesion[3], but some have an association with genetic abnormalities like familial adenomatous polyposis and Beckwith-Wiedemann syndrome[4]. But for extrahepatic hepatoblastoma, this aspect has not been reported.

Our case presented with occasional abdominal pain and fever only. Affected children may present with abdominal distention or discomfort, fatigue, and anorexia and in case of tumour rupture with severe abdominal pain, vomiting, and anaemia[1]. The useful tumour marker is serum alpha-fetoprotein (AFP) which is used to detect the malignant change, treatment response, and relapse. In our case, serum alpha-fetoprotein (AFP) level was raised more than 1210ng/ml. Some variants of hepatoblastoma present with normal or low AFP levels [4,5]. However, the high level of

serum AFP is not specific, as it is also raised in other conditions, such as yolk sac tumours, focal nodular hyperplasia, hepatocellular carcinoma, mesenchymal hamartomas, and infantile haemangioendothelioma[6]. In our case, teratoma was also taken as a differential.

Abdominal ultrasonography and computed tomography (CT) are useful modalities for diagnosis. Radiologically due to heterogenous nature, presence of calcifications, and site being inferior to the liver and separable from right kidney, our case mimicked a neuroblastoma. This initial suspicion was partly ruled out by the presence of normal vanillylmandelic acid (VMA) levels and lack of hypertension. Histological diagnosis of the tumour lead to the definitive diagnosis and other differentials were excluded. The Japanese Study Group for Pediatric Liver Tumors (JPLT) suggests that treatment of paediatric liver tumours should be done after definitive diagnosis is made by histopathological analysis of the biopsy specimen, except in life-threatening urgent situations such as tumour rupture or invasion into cardiac chambers[7].

Complete resection of the tumour is the cornerstone of curative therapy. For advanced diseases and unrespectable tumours, chemotherapy is useful. Neo-adjuvant chemotherapy helps in decreasing tumour size and consequently, it is more likely to be resected completely[8]. According to the guidelines of the International Childhood Liver Tumors Strategy Group (SIOPEL) preoperatively[9], six cycles of cisplatin monotherapy were given to our patient, which resulted in a decrease in tumour size by 70%, after which complete resection of the tumour was done. Operatively, lack of capsular breech differentiated it from exophytic growth of hepatoblastoma. Adjuvant chemotherapy was done, and long-term follow-up is planned to prevent recurrence. Extrahepatic hepatoblastoma is rare. To our knowledge, only two cases of extrahepatic hepatoblastoma have been reported so far[3,10].

Conclusion

Histological diagnosis is the gold standard for differentiating extrahepatic hepatoblastoma from other tumours and improving survival by guiding medical practitioners about neoadjuvant chemotherapy. Complete surgical resection combined with adjuvant chemotherapy and a long term follow up is crucial, to avoid recurrence in these cases.

Disclaimer: None.

Conflict of Interest: None.

Source of Funding: None.

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