Emergency Hepatic Resection a Life Saving Procedure in Ruptured Hepatoblastoma: Case Report

Gururaj Deshpande • Mahesh D. Patel

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Introduction

Hepatoblastoma is the most common primary liver tumor in children, accounting for just over 1 % of pediatric cancers with most cases occurring prior to 5 years [1]. The incidence of this tumor has not changed since last decade with average male: female ratio of 2:1 [2]. The etiology is unknown, but it has been associated with Beckwith-Weidemann syndrome [3], familial adenomatosis polyposi [4], and low birth weight [5]. These tumors present with an asymptomatic abdominal mass palpated either by a parent or paediatrician. Abdominal pain, weight loss, anorexia, nausea, and vomiting may be present, particularly in advanced disease. Imaging is an important aspect of diagnosis of these patients, which includes an ultrasonography of abdomen and a contrast enhanced CT scan. The management is multimodal, involves medical oncologists, surgical oncologists, paediatricians and specialist nursing team. The primary treatment is surgical resection: however, chemotherapy plays an important role by increasing the number of tumors that are resectable. Serum Alpha Feto-Protein (AFP) is a sensitive biological marker of the disease. There is a correlation between AFP and extent of disease for all stages, and the rate of decline in AFP with treatment is prognostic. Low AFP levels are associated with anaplastic histology and poor outcome [6]. Tumor size, number of involved lobes, regional lymph node involvement, and distant metastases determine stage [7]. The prognosis for patients with resectable

tumors is fairly good; however, the outcome for those with nonresectable or recurrent disease is poor. Surgical resections done can be anatomical or non-anatomical resections. Resections as much as trisegmentectomies with Intraoperative trans-arterial embolisation have been done in patients with adequate hepatic reserve for ruptured hepatoblastomas [8]. In very few occasions hepatoblastomas have presented with spontaneous rupture with hemoperitoneum, we are presenting one such case.

Case Report

The patient was a 7-year-old male, who presented to us with 1 day history of sudden onset abdominal pain with gradual deterioration of general condition progressing to altered sensorium and shock. Abdominal sonography was done which suggested a mass lesion in the right lobe of liver with free fluid in the abdomen. Liver function tests, serum AFP and contrast enhanced Abdominal computed tomography (CECT) were also done. CT confirmed the mass lesion involving segments V, VI, VII and VIII of liver and also suggested it to derive its vascular supply from branches of right hepatic artery with intra-abdominal fluid accumulation with clots over the dome of right liver suggesting hemoperitoneum (Fig. 1). Hemogram was suggestive of active haemorrhage with hemoglobin of 5.0 g%. In view of raised serum AFP of 484.24 ng/ml and above imaging characteristics, a working diagnosis of ruptured hepatoblastoma was made. Laboratory studies showed the following values: AST 67 U/L; ALT 51 U/L, alkaline phosphatase 252 U/L ; platelet count 456 000/ml; Prothrombin time-12.7 s/13.0 s (INR-0.98). CT scans of the abdomen (Fig. 1) showed massive accumulation of intra-abdominal fluid and one large heterogeneous mass (c. 12.0 cm×9.0 cm×9.0 cm) with

G. Deshpande (✉) • M. D. Patel
Department of Surgical Oncology, Gujarat Cancer and Research
Institute, New Civil Hospital Campus, Asarwa,
Ahmedabad 380016, India
e-mail: drgururajdeshpande@gmail.com

cystic and solid components in the right lobe of the liver. Ideally patient should have undergone Trans-arterial embolisation of the right hepatic artery, but as patient was deteriorating and also patient had presented during emergency hours and we were not able to get embolisation done. After initial resuscitation with transfusions of packed cells patient was taken up for exploratory laparotomy. On laparotomy there was a ruptured tumor arising from right lobe of liver with a clot over it (Fig. 2). Right Hepatectomy with intra hepatic vascular ligation was planned. Intermittent Pringle's manoeuvre with 15 min occlusion and 5 min release was done. We performed right hepatectomy with five occlusions by Pringle's manoeuvre. Specimen was delivered after ligating and transfixing right and middle hepatic veins (Fig. 3). Final histopathology was reported as hepatoblastoma—mixed embryonal and fetal type. The stage of the disease was SIOP stage 2. In the post operative period patient had biliary leakage leading to jaundice, MRCP was done to evaluate the leak, and it was managed conservatively. Patient was referred for chemotherapy to the medical oncologists. Patient was started on chemotherapy, Cisplatin and Adriamycin. The dosage of the above chemotherapeutic agents are Adriamycin 25 mg/m² from day 1 to 3 and cisplatin 20 mg/m² from 1 to 5 days, both of them given as continuous infusion, repeated every 3 weeks for 6 cycles. Follow up CT Scans after 2 months revealed mild IHBR in the liver, nodules in the sub-diaphragmatic space, sub-hepatic space and bilateral basal pulmonary nodules suggestive of metastasis and AFP had increased to 206,210 ng/ml. Patient is responding well to chemotherapy and follow up imaging shows stable disease and 6 month follow up AFP is 5,000 ng/ml.

Fig. 1 CECT of the abdomen showing ruptured heterogeneous, multi-lobulated mass lesion in the right lobe of liver with hemoperitoneum

Fig. 2 Intraoperative image showing the ruptured tumor of the right lobe of liver covered with clots

Discussion

Hepatoblastomas are malignant tumor occurring in infants and most cases occur before the age of five years, and reports of adult cases are extremely rare. On imaging they are seen as large mass, frequently with calcifications, heterogeneous echogenic pattern on sonograms or heterogeneous enhancement with hyper dense internal septa on CT scans [9], and on MR images, hepatoblastoma appears as a heterogeneous mass, with areas of hypointensity seen on T1 -weighted images, corresponding to hemorrhagic necrosis [10]. Most of these tumours arise in the embryo during the development of the liver and contain embryonic features such as numerous mitoses. They sometimes show ribbon-like, rosette-like, or papillary formations. Hepatoblastoma is classified by histology as epithelial (56 %) or mixed epithelial/mesenchymal (44 %). Epithelial hepatoblastoma is further divided into pure

Fig. 3 Resected specimen of right hepatectomy

fetal (31 %), embryonal (19 %), macrotrabecular (3 %) and small cell undifferentiated (3 %) [8]. In completely resected tumors, pure fetal histology confers a better prognosis, while small-cell undifferentiated histology is associated with a poor prognosis and increased number of mitosis is associated with poor prognosis [11]. Studies suggest that alterations in the APC/b-catenin pathway contribute to the pathogenesis of hepatoblastoma [12]. Ishank and Glunz classified hepatoblastomas into three groups in 1967: epithelial type and mixed epithelial and mesenchymal type. The tumor type from this patient was distinguished from a hepatocellular carcinoma by the presence of angiodysplasia and also by the peculiar arrangement of bright and dark epithelial cells in sheets or ribbons. This patient, therefore, had a tumor of the mixed epithelial and mesenchymal type according to this classification. Most cases in the literature are of the mixed type. The precise mechanism to cause the tumor has been elusive; present investigations of the cytogenetic and molecular genetic aberrations in hepatoblastomas revealed involvement of chromosomal loci on 1q, 2 (or 2q), 4q, 8 (or 8q), and 20. Studies suggest that alterations in the APC/b-catenin pathway contribute to the pathogenesis of hepatoblastoma [13]. Current treatments for the tumor include surgery and chemotherapy. Combination chemotherapy with adriamycin and cisplatin has been considered effective for hepatoblastoma. A review search in the literature revealed only 6 paediatric cases have presented with spontaneous rupture which have been managed with emergency laparotomy. Of these only one survived the surgery. This case is the first case of ruptured hepatoblastoma who had undergone emergency hepatectomy that was reported in India. Our patient is on regular follow up.

Conclusion

Ruptured hepatoblastoma though rare entity needs prompt intervention at the earliest. Hepatic resections, trans-arterial embolisation as well as staged resection after control of initial haemorrhage have been done. Age of the patient is an important criterion for performing emergency hepatic resections, as hepatic reserves as well as haematological reserves are better in older patients compared to infants. In the above discussed case trans-arterial embolisation was not possible because of rapidly deteriorating general condition of the patient. Relation between the size of the tumor and the

propensity to rupture has not been studied as only few cases of ruptured hepatoblastomas have been reported but size of the tumor is more than 10 cm in three of the reported cases [14]. Rupture results in increased chances of intra peritoneal metastasis as we have noticed in this patient on follow up imaging. We conclude that surgical intervention is best life saving modality of management in such cases even when there is active haemorrhage.

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