CASE REPORT

Ruptured Hepatocellular Carcinoma in a Child with Budd-Chiari Syndrome

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Background: Hepatocellular carcinoma is an uncommon complication described in patients with Budd-Chiari syndrome. **Case characteristics**: A 12-year-old boy with Budd-Chiari syndrome, who was earlier treated with Transjugular intrahepatic porto-systemic shunt (TIPS), presented with acute onset hemoperitoneum and hypotension. **Outcome**: It was diagnosed to be a case of ruptured hepatocellular carcinoma. **Message**: Successful TIPS may not prevent the development of hepatocellular carcinoma, and children with Budd Chiari syndrome should be monitored for the same.

Keywords: *Complication, Hemoperitoneum, Liver tumor, TIPS*.

Budd-Chiari Syndrome (BCS) is a progressive disease leading to portal hypertension, liver dysfunction, hepatopulmonary syndrome, and cirrhosis. Hepatocellular carcinoma (HCC) is a rare but potential complication of BCS. It has been reported in few adult patients despite undergoing radiological intervention like Transjugular Intrahepatic Porto-systemic Shunt (TIPS) [1]. We present a 12-year-old boy, a known case of BCS treated successfully with TIPS, who presented with hemoperitoneum due to a ruptured HCC.

CASE REPORT

A 12-year-old boy, known case of BCS, presented with an acute onset abdominal pain with hypovolemic shock. He had tachycardia, tachypnea and hypotension, and was pale. His abdomen was distended along with tenderness and guarding.

His illness first manifested at the age of four years with abdominal distension (ascites) and hematemesis. At 6 years, he presented to our institute with failure to thrive, refractory ascites and firm hepatosplenomegaly. Complete blood count showed evidence of hypersplenism. Endoscopy revealed small esophageal varices. There was no coagulopathy or liver dysfunction. Diagnosis of BCS was confirmed on the basis of ultrasound (USG) Doppler and computed tomography (CT) angiography which revealed complete occlusion of the middle and left hepatic veins and short segmental occlusion of the right hepatic vein (RHV) ostium. A combined percutaneous- transjugular approach was used to stent the RHV ostium. He showed good clinical response, but after 6 months developed stent occlusion. He underwent TIPS procedure with 10 mm × 60 mm GORE VIATORR stent. The portosystemic gradient reduced from 28 mmHg to 3 mmHg. There were no immediate complications. On follow-up, ascites subsided and weight improved (from <5th centile to 25th centile). He had no further variceal bleeds. The patient was put on anticoagulation with warfarin. Thrombophillia workup (Protein C, Protein S, Anti thrombin III, Lupus anticoagulant, Anti cardiolipin antibodies) and JAK2 mutation studies were negative.

Three-monthly USG Doppler studies were done to evaluate the TIPS patency. Three years after insertion of TIPS, USG Doppler showed a shunt block (proximal end) following which hepatic venogram and balloon angioplasty was done (gradient reduced from 23 mmHg to 3 mmHg). Patient remained asymptomatic since then till the current episode.

Investigations during the current episode revealed hemoglobin: 2.8 g/dL, total leucocyte count: 3x10^9/L, platelets count 120×10^9/L, prothrombin time: 21.0 seconds, INR: 1.9, AST: 29 IU, ALT: 41 IU/L, TSB: 2.1mg/dl, GGT: 195U/L, Alkaline phosphatase: 301 IU/L. USG abdomen showed a mass in the right lobe of liver with breach of liver capsule, and hemorrhagic fluid in the peritoneum. CT abdomen showed a large (9.2cm × 4.5cm) ill defined, mixed density lesion along segment 8, 5, 7 extending into the perihepatic space suggestive of ruptured HCC (***Web Fig.* 1a**). Tumour with characteristics of a malignancy was seen in the lower lobe of the right lung along with right-sided pleural effusion suggesting

metastasis to lungs. Alphafeto protein (AFP) was 2380 µg/L. Hepatic arteriogram (Digital subtraction angiography) was done which revealed a hyper vascular mass in the segment 7, segment 8 of the right lobe of liver, showing neovascularization supplied by multiple branches of the right hepatic artery, with contrast extravasation in delayed phase. (***Web Fig*. 1b**). Hepatic artery embolization (super selective embolization with PROGREAT microcatherter and injecting 200-300 PVA particles) was done on emergency basis to control bleeding. Shock was corrected with intravenous fluids and packed cell transfusions.

A diagnosis of ruptured HCC in the setting of BCS was made. Due to advanced nature of the illness, he was managed conservatively. Palliative care in the form of recurrent blood transfusions was given. He succumbed 7 months later due to progressive abdominal distension, jaundice and anemia.

DISCUSSION

Hepatocellular carcinoma is an uncommon complication of BCS and has been only reported in adults. Our patient developed BCS at the age of 4 years and with radiological intervention his clinical course improved, but after 6 years he succumbed due to HCC.

Park, *et al*. [2] published their experience with 67 patients of BCS from Korea, where 17 developed HCC on long term follow up. Median time lag between diagnosis of BCS and HCC was 51 months [2].

BCS leads to chronic congestion of liver due to impaired drainage. Long standing fibrosis and dysplastic regenerative nodules may predispose to HCC [3]. Our case had underlying cirrhosis of liver; nodules seen on screening USG were thought to be regenerative and the possibility of development of malignancy was unforeseen. The size of regenerative nodule larger than 4 cm may be helpful in suspecting HCC but larger size with benign etiology has been reported [4,5]. Contrast enhanced USG with a cut-off size of 4 cm may be helpful in screening children with BCS [6].

The modality of treatment of spontaneous rupture of HCC is not well-defined in children as only one case has been reported [8]. As per adult studies, transarterial embolization/chemoembolization and resection, if possible, are two approved treatment options for ruptured HCC; though long term outcome remains poor [9]. Our patient underwent hepatic artery embolization to control bleeding as a palliative management in view of lung metastasis.

As more children with BCS are being diagnosed at an early age and managed with radiological intervention, they need to be screened for hepatocellular carcinoma on long term follow-up. Early diagnosis may provide a chance for curative treatment.

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