ORIGINAL RESEARCH PAPER

Hepatobiliary Surgery

ACUTE ABDOMEN IN BUDD CHIARI SYNDROME: A RARE CAUSE AND DIFFERENTIAL DIAGNOSIS

KEY WORDS: Ruptured HCC, Budd Chiari, acute abdomen.

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ABSTRACT

Budd Chiari syndrome (BCS) caused by Hepatic venous or inferior venacaval obstruction, can have an acute presentation. Hepatocellular Carcinoma in patients with BCS is a rare complication. An acute presentation with rupture is even more rare. We describe a 63 year old man diagnosed with BCS, 9 years back. He had been treated with a portocaval shunt a few years ago, but currently presented with sudden onset abdominal pain and raised lactate levels. Evaluation revealed a 10 cm large, ruptured HCC with hemoperitoneum. Diagnostic dilemmas and treatment options are discussed.

INTRODUCTION

Hepatocellular carcinoma (HCC) is the most common primary malignant tumor of the liver. However, the incidence of rupture in HCC is rare, with incidence ranging from 3-15% with poor short- and long-term prognosis[1-3].

Budd Chiari Syndrome (BCS) is a rare disorder caused by occlusion of hepatic vein or inferior vena cava. It has two forms according to the obstruction site: primary hepatic vein obstruction (classical BCS) and obstruction of the hepatic portion of the inferior vena cava (IVCO). The IVCO form is common in Asia and Africa but rarely reported in Western countries. It may clinically present itself in the acute (fulminant), subacute or chronic (decompensated) forms. Usual symptoms are, abdominal pain, hepatomegaly, ascites and pedal edema. HCC in BCS is rare, with an average lag time from diagnosis of several years to decades [4].

We report a 63 year old gentleman with BCS, who had had a portocaval shunt, and lost to follow up, who presented with a sudden onset abdominal pain which on further workup was diagnosed with a ruptured HCC.

CASE REPORT

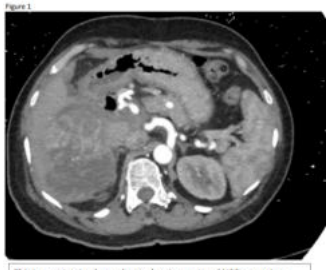
A 63-year-old gentleman, with Budd Chiari Syndrome diagnosed in 2012, following which he had a side to side portocaval shunt the same year, presented with history of 4 days of central and right sided abdominal pain which was sudden onset, persistent and associated with one episode of vomiting. There was no history of hematemesis or melena. He was a diabetic, a chronic smoker with history of alcohol use for 20 years. On examination, he was icteric, with tachycardia (112/minute) and tachypnea (22/minute) with normal blood pressure (110/70mmhg). Abdominal examination revealed mild abdominal distention with generalized tenderness and guarding. Initial clinical differential diagnosis included spontaneous bacterial peritonitis, hollow viscus perforation with peritonitis and acute mesenteric ischemia.

Investigation showed the following: Hemoglobin-10.2gm/dl; Total count-12800 cells/cumm Platelets- 2.07 lakhs/cumm; Total Bilirubin- 3.51mg/dl; Transaminases: normal; ALP- 180 U/L; S. Albumin 2.9 gm/dl; D- Dimer – 7.7mg/L; C Reactive Protein- 7.7 mg/dl; Creatinine- 1.8mg/dl; Serum AFP - 1200 ng/ml; Serum Lactate - 7.8 mmol/L. Cardiac echo showed features of severe pulmonary hypertension (estimated systolic Pulmonary arterial pressure- 87mmHg), with an ejection fraction of 53%.

On non-contrast computed tomography of the abdomen, there was hemoperitoneum. After hydration, a contrast enhanced CT scan was performed, which showed a heterogenous 10x7x8 cm, partly exophytic lesion, occupying segments V,VI,VII,VIII of right lobe of liver, with active contrast extravasation into the lesion via the branches of the right hepatic artery. This lesion had ruptured leading to the hemoperitoneum (Figure 1).

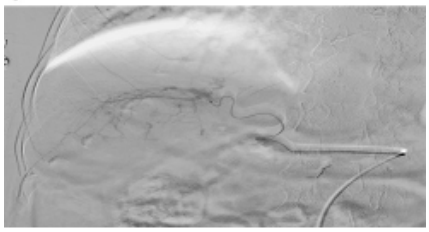
A ruptured HCC (stage IV) with hemoperitoneum, in a background of BCS (MELD 23 and CTP-B) was diagnosed. He was managed with blood transfusion and emergency angioembolization. Pre-embolization hepatic artery run is shown in Figure 2. The inferior segmental branch of the right hepatic artery was embolized and no tumor blush was observed post procedure (Figure 3). Patient was stable post procedure and was discharged on the 5th day, post procedure. In view of the rupture, liver transplant was not an option. Because of his performance status, severe pulmonary hypertension and MELD of 23, liver resection was also deferred.

Figure 1



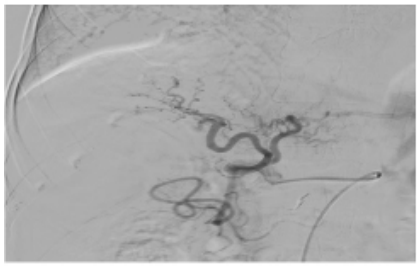
This is a contrast enhanced scan showing ruptured HCC occupying segment V,VI,VII,VIII with hemoperitoneum.

Figure 2



Pre embolization with hepatic artery run.

Figure 3



Post embolization of the inferior segmental branch of the right hepatic artery with absence of tumour blush.

DISCUSSION

BCS may present as a fulminant form (3-26%)[5] wherein, an acute thrombosis of the hepatic venous outflow causes an acute and rapidly progressive liver injury with progressive jaundice, high serum transaminases. This presents clinically with abdominal pain, distention, ascites and hepatomegaly. The other causes of an acute abdomen in BCS are spontaneous bacterial peritonitis, hollow visceral perforation and ischemic bowel from mesenteric venous or arterial thrombosis due to the underlying prothrombotic state. These were our initial clinical working diagnoses when our patient was assessed in the emergency room. However, imaging revealed another rare cause of an acute abdomen in this context, namely, hemoperitoneum from a ruptured HCC.

Rupture of HCC in cirrhosis is uncommon, but a well-known complication. Although this precludes liver transplant as an option, in these patients, an elective liver resection in selected patients provides survival benefit when compared to non-resected patients.[6]

The development of Hepatocellular carcinoma after Budd Chiari Syndrome is uncommon, with a lag period of several decades. HCC rupture occurring in the context of BCS is extremely rare and hence the diagnosis is often not considered in the initial working differential diagnosis, when these patients present to the ER as an acute abdomen. The initial management in such patients should include resuscitation and emergency angioembolization to arrest the bleeding. The definitive management options are limited and largely palliative in nature. This is because a ruptured HCC effectively is stage IV disease and contraindicates liver transplantation, which otherwise would be an ideal option. Moreover, a major liver resection in BCS is difficult due to a congested liver with outflow obstruction. Our patient in addition was a poor surgical risk due to his performance status and severe pulmonary hypertension. Stereotactic external beam radiotherapy or trans arterial radioembolization (TARE), in combination with systemic therapy are the palliative options that have been recommended for this patient.

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