Case Report

Acute Budd-Chiari syndrome during hepatic vein catheterization ☆

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ABSTRACT

This case report describes a 4-year-old male with a history of hepatoblastoma, treated with chemotherapy followed by a right extended hepatectomy. Secondary to this, the patient experienced recurrent hepatic vein stenosis. He was treated initially with conventional angioplasty, followed by paclitaxel-coated balloon dilatations in an attempt to prevent episodes of re-stenosis. During the catheterization of the hepatic vein in one of the treatments, hemodynamic instability due to an acute Budd-Chiari syndrome occurred. The hemodynamic compromise became unresponsive to intravascular resuscitation and inotropic support. The patient was then treated with an emergency conventional angioplasty of the hepatic vein, which resulted in a rapid response and eventually in a full recovery. The etiology of this complication remains unclear; however, it may have been secondary to endothelial damage leading to acute thrombosis and/or venous spasm. Conventional angioplasty was successful in managing this complication. Awareness of iatrogenic acute Budd-Chiari syndrome as a potentially fatal complication during hepatic catheterization/dilatation, especially in the posthepatectomy setting, successful management of this complication, and the importance of a multidisciplinary and rapid response, is emphasized.

Introduction

This report describes the case of a 4-year-old boy with a history of hepatoblastoma, right extended hepatectomy, hepatic vein stenosis treated with conventional angioplasty and then with paclitaxel-coated balloons alongside primary disease recurrence. During the 11th dilatation, acute hepatic vein obstruction secondary to catheterization occurred, resulting in acute hepatic vein thrombosis. These findings were compatible with an acute iatrogenic Budd-Chiari syndrome (BCS). This is an extremely uncommon event which can be life threatening and difficult to manage.
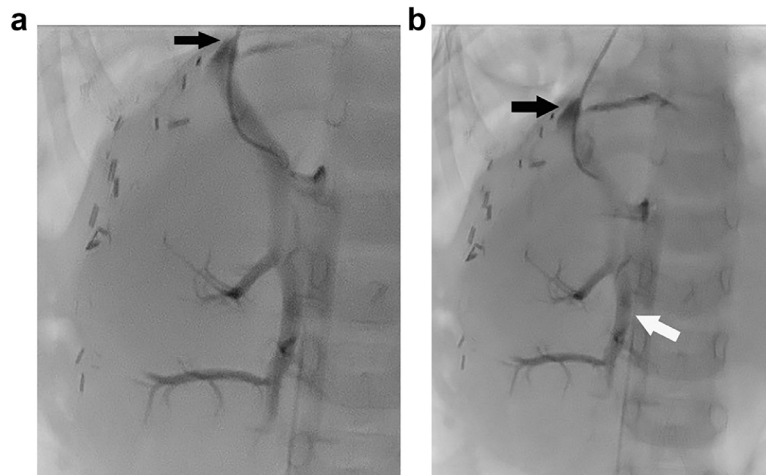
Fig. 1 – Hepatic vein venograms during (a) and after (b) contrast injection (a). A significant stenosis at the anastomosis is observed (black arrow) associated with dilatation of the hepatic vein and very poor clearance of contrast in the delayed image (white arrow).

Case report

Informed consent was obtained from the patient's parents for this publication. This report describes the case of a 4-year-old male who first presented at 15 months of age with abdominal distention and a palpable abdominal mass. Ultrasound and computed tomography (CT) of the abdomen showed a large heterogeneous liver mass consistent with a hepatoblastoma associated with IVC invasion. After chemotherapy, the tumor became resectable and a right extended hepatectomy was performed and a surgical venous conduit was constructed. Thrombosis of this conduit was diagnosed 1 month later and anticoagulation was started. After 4 weeks of treatment, the patient presented with signs of portal hypertension secondary to stenosis of the anastomosis of the single hepatic vein diagnosed by venography. This was treated initially with conventional angioplasty and later with paclitaxel coated balloons, in an attempt to better manage the stenosis. The patient had recurrence of the primary disease in the right lung, which resulted in a right pneumonectomy. Cycles of experimental chemotherapy were initiated.

The patient presented, now at age 4, for elective hepatic venogram and potential dilatation. A port removal was also scheduled during the same anesthetic. This was his 11th dilatation, with the most recent one being 31 weeks prior to this visit. The time between dilatations had been extended due to the patient's lack of symptoms, thought to be due to the use of the paclitaxel-coated balloon. The patient had a history of cough and influenza the week prior to this procedure. He was American Society of Anesthesiology physical status classification system III. The patient was on warfarin, but was switched to enoxaparin prior to the procedure. The right port-a-cath was removed successfully. The right internal jugular vein was used for access. A 9-French vascular sheath was placed. The hepatic vein was then catheterized with a 4-French Bernstein catheter and a 0.035″ hydrophilic guidewire. Immediately after the initial contrast injection, which demonstrated
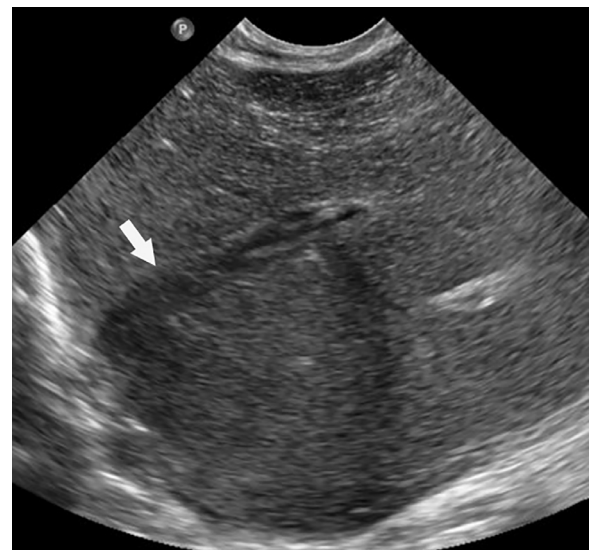

Fig. 2 – The sonographic examination of the hepatic vein showing the lumen filled with echogenic material (arrow) compatible with thrombosis.

significant stenosis at the venous anastomosis and very stagnant flow of contrast (Fig. 1), the patient experienced an acute clinical deterioration. The patient became tachycardic, with a heart rate rising from 120 to 160 beats per minute. He then became hypotensive, with his blood pressure dropping from 100/50 mm Hg to 70/30 mm Hg, with no changes in O_2 saturation, $ETCO_2$, and peak airway pressure. The anesthesiologist requested that the procedure be put on hold to stabilize the patient. A limited sonographic examination of the liver showed slow flow in the hepatic vein, an early sign of thrombosis (Fig. 2). A bolus of 1,300 units of heparin was administered. Phenylephrine and epinephrine boluses were also given. The blood pressure increased partially in response

Fig. 3 – Left pulmonary angiogram performed via the right neck sheath showing adequate opacification of the pulmonary artery and branches, with no evidence of pulmonary embolism. The patient had a history of right pneumonectomy due to metastatic disease.

to this; however, the patient's tachycardia reached 190 beats per minute. A pulmonary angiogram of the left lung was performed and a pulmonary embolism was excluded as the cause of the acute clinical deterioration (Fig. 3).

A temporary central venous line was placed. A right radial arterial line was also inserted. Two large bore peripheral IVs were inserted bilaterally into the saphenous veins. Pediatric intensive care unit, oncology, general surgery, and thrombosis staff physicians were called to the procedure room. The patient continued to deteriorate. The patient's blood pH dropped from 7.2 to 7.1, HCO$_3$ from 20 to 16.2 mEq/L, base excess −6.8 to 11.4 mEq/L, lactate 2.4 to 3.4 mmol/L. Intravenous boluses of normal saline and 5% albumin were administered, sodium bicarbonate was given and an epinephrine infusion was started. A total of 1.5 hours was spent attempting resuscitation. It was concluded that the patient was in shock and the prognosis was unclear. The clinical examination of the abdomen demonstrated an acutely edematous liver, which correlated on ultrasound with progressive hepatic vein thrombosis and new onset of free fluid. The multidisciplinary discussion concluded that the cause of the clinical presentation of shock was an acute obstruction of the hepatic vein resulting in decreased cardiac preload. As medical management was thus far unsuccessful, it was agreed upon that the optimal therapeutic option was to attempt to restore flow through the hepatic vein with angioplasty.

An emergency conventional angioplasty was then performed using a 6-mm balloon (Fig. 4). A significant improvement of the flow of contrast through the area of stenosis was observed, with residual clot noted in the hepatic vein. Following the angioplasty, which re-established flow, the patient immediately responded with an increase in blood pressure. A slow decrease in heart rate followed. An ultrasound was

performed showing increased flow in the hepatic vein and a reduction in the size of the thrombus (Fig. 5). A heparin infusion was started. The patient was transferred intubated to the pediatric intensive care unit, and remained stable overnight. He was discharged from the hospital after 3 days, clinically well, with no sequelae related to the procedure and on anticoagulation (switched to enoxaparin). The only complication related to the clinical episode was a hematoma of the port-a-cath subcutaneous pocket, which was managed conservatively with dressing changes on an outpatient basis.

Discussion

BCS is a disease complex that results from impairment of hepatic venous outflow, mostly at the level of the hepatic veins, IVC, or both [1,2]. BCS is rare, with an overall incidence of 0.1 to 10 cases per million people annually [1]. When untreated, BCS results in portal hypertension, cirrhosis, and liver failure that is oftentimes fatal. If left untreated, there is a 3-year mortality of 90% [1]. If treated, patient survival can range from 42% to 100% [1]. BCS can be classified based on different elements: etiology (primary or secondary), clinical course (acute or chronic), or morphology (truncal, radicular or veno-occlusive type) [1]. With respect to clinical course, most patients present with symptoms of chronic BCS, associated with gradual onset abdominal distention, tortuous abdominal veins, and portal hypertension [3]. The acute BCS is a rare presentation, and one that is associated with acute onset abdominal pain, ascites, hepatomegaly, and fulminant and progressive liver failure [3]. The differential diagnosis for causes of BCS include hypercoagulable states (such as with myeloproliferative diseases), tumoral invasion or extrinsic compression, idiopathic, as well as rarer causes such as parasitic infection or liver abscess. Diagnosis of BCS relies on the demonstration of venous outflow obstruction as well as structural changes to the liver or portal venous system, or a secondary pathology seen on ultrasound, CT or MRI [1,2]. Management of BCS is based on the underlying cause. Treatment often proceeds in a stepwise manner. Consensus of expert opinions indicates that all patients should receive anticoagulation [1,4]. Following medical treatment, endovascular treatment to restore vessel patency is typically performed, which may include local thrombolysis, angioplasty, and stenting [1,4]. The next step of the treatment algorithm is placement of a transjugular intrahepatic portosystemic shunt [1,4]. Surgical portosystemic shunting is also possible, but expert opinion favors radiologic intervention over this option [3]. Finally, in rare cases, treatment may need to escalate to orthotopic liver transplantation [1,4].

BCS is an uncommon disease in children, with very few descriptions of its clinical features and treatment options in the pediatric population [3]. One study of 46 pediatric BCS patients found that all had chronic BCS, diagnosed via Doppler ultrasound in 95% of cases. Additionally, almost 75% of the cases investigated were found to have a prothrombotic state [3]. Most commonly, it was the hepatic vein that was obstructed. They found radiological intervention (including angioplasty, stenting, and transjugular intrahepatic portosystemic shunt) to be successful in 100% of cases in association with
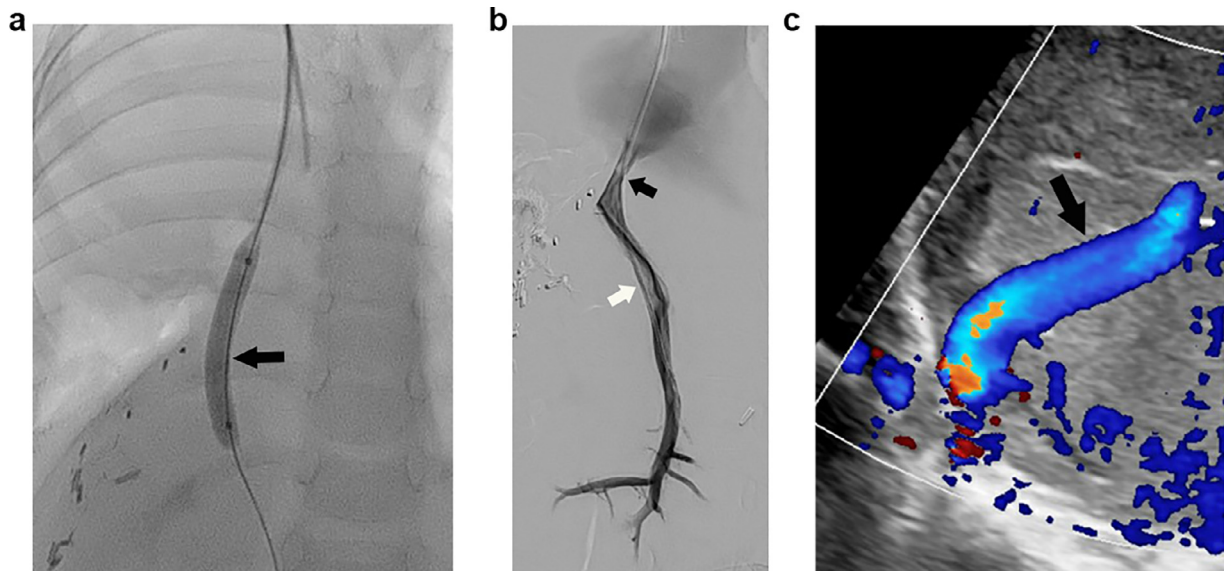
Fig. 4 – (a) Emergency conventional angioplasty of the area of the anastomosis with a 6-mm balloon (arrow). (b) Contrast flow was re-established in the area of the anastomosis (black arrow). Residual nonocclusive thrombus was observed in the hepatic vein (white arrow). (c) The color-Doppler study showed satisfactory flow in the hepatic vein (arrow).
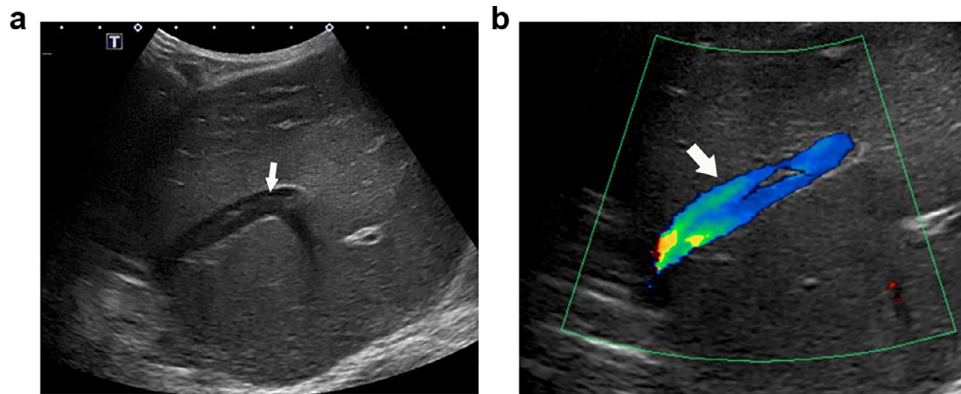

Fig. 5 – (a) Gray-scale sonographic examination of the hepatic vein 24 hours after the event showed progressive resolution of the nonocclusive residual thrombus (arrow). (b) The color-Doppler study demonstrated satisfactory hepatic vein flow.

medical therapy. Complications of neck hematoma and hemorrhagic ascites occurred in one patient each [3]. Another study investigated radiological interventions for pediatric patients with gastrointestinal diseases, including 14 with chronic BCS who were treated with stenting, and the mean stent patency at 2 years was approximately 79% [5]. Pediatric BCS has also been described in the literature as a serious complication of liver transplantation, and has been reported to be successfully treated by percutaneous radiologic interventions [6–8]. One case series examined 25 pediatric patients with acute BCS, of which 21 patients underwent balloon angioplasty, and this was found to be a safe and successful treatment strategy, with surgical management being an appropriate attempt in failed radiological management cases [9].

The current case represents one of acute BCS that was veno-occlusive. The differential diagnosis for the etiology includes venous endothelial trauma from catheterization, and venous spasm in the context of significant progressive venous stenosis that was left untreated for a long time interval due to the lack of clinical symptoms. The patient's recent respiratory infection and potential decrease on lung capacity appears unlikely to have played a role. There have been sporadic reports in the literature of iatrogenic BCS. Most of these have been in the postliver transplantation setting [4,10]. There have been reports of BCS from nonliver interventions, such as a case of BCS following laparoscopic cholecystectomy, but this was due to an underlying undiagnosed hematologic condition [11]. Another example is a case of a patient undergoing a pancreaticoduodenectomy who then experienced an acute BCS due to a compressive hematoma of the retro-hepatic IVC [12]. Additionally, 1 case series described 3 adult patients with BCS who experienced acute iatrogenic IVC thrombi, following a failure of re-establish patency of the occluded IVC, were successfully treated with agitation thrombolysis

[13]. Two cases of iatrogenic BCS following hepatectomy for hepatolithiasis had been described [14]. A few cases have been reported of acute BCS following hepatectomy due to torsion of the remnant liver causing compression of the IVC or kinking of the left hepatic vein [15,16]. Another case in the literature detailed recurrent acute BCS following right hepatectomy due to left hepatic vein kinking, which was ultimately treated definitively with IVC angiogram and left hepatic vein stenting [17]. One particular case has been described of acute BCS occurring intraoperatively after extended right hepatectomy [15]. In this case, similar to the one described here, the patient experienced sudden life-threatening hemodynamic collapse that was unresponsive to intravascular volume therapy and inotropic support [15].

This case of acute iatrogenic BCS posthepatectomy during a routine catheterization of a stenotic hepatic vein emphasizes the need to consider this as a risk when performing hepatic vein interventions, particularly in patients who have undergone hepatectomies and therefore may rely on a single hepatic vein. Additionally, it illustrates acute thrombosis secondary to intravascular instrumentation as a potential cause for BCS. The hemodynamical impact of acute BCS results from reduced preload and right atrial filling [1,2]. The acute clinical presentation is a sudden life-threatening hemodynamic collapse that is unresponsive to intravascular volume therapy and inotropic support. A rapid response was seen following correction of the hepatic venous outflow obstruction, which supports this as a fundamental step in the management. An interdisciplinary approach with effective communication and collaboration is crucial in responding to acute BCS.

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