

# Hydatid Cyst of Liver Complicated with Budd-Chiari Syndrome and Portal Vein Thrombosis

Dear Editor,

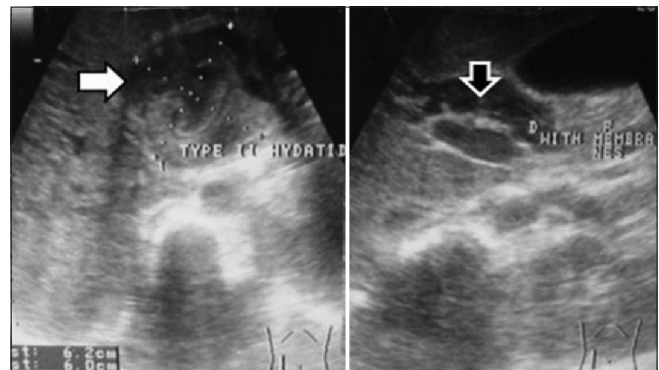
Prevalence of hydatid cyst is increasing in developed countries due to immigration. However, complicated hydatid disease of liver causing inferior venacaval and portal vein thrombosis is exceedingly rare. In literature, five similar cases have been reported worldwide; however, most of them have been treated with radical surgery. Our case differs in the fact that it was successfully treated with non-operative approach using sequential endoscopic retrograde cholangiopancreatography (ERCP) and catheter-directed thrombolytic therapy.

A 42-year-old Asian female presented with 6 days history of right upper quadrant pain, progressive jaundice, high grade fever, persistent vomiting, and progressive distention of abdomen. Examination revealed dry mucous membranes, scleral icterus, temperature of 103°F, pulse 120/min, blood pressure 80/60 mm Hg, and tender hepatomegaly. Table 1 demonstrates initial and subsequent laboratory parameters. Sonography of abdomen demonstrated a well defined cyst (6.2 × 6cm) in the left lobe of liver with floating membranes inside the cavity and ascites [Figure 1]. The patient was initially resuscitated with intravenous fluids, vasopressors, blood transfusions, and antibiotics along with albendazole were also started. Emergency ERCP was performed which revealed dilated common bile duct with multiple filling defects [Figure 2a]. A Naso Biliary Drain (NBD) was placed in the left hepatic ductal system, which drained large amount of pus mixed with membranes and bile. NBD cholangiogram revealed large cavity in left lobe of liver communicating with left hepatic duct [Figure 2b]. Culture of blood, urine and ascitic fluid was sterile but pus from NBD came positive for scolices and *Escherichia coli*. *Echinococcus* serology assessed by ELISA was also noted to be positive.

Contrast Enhanced Computerized Tomography of abdomen demonstrated well-defined cystic cavity in left lobe of liver, multiple small hepatic abscesses, ascites and well-defined filling defect within the lumen of inferior

**Table 1: Comparison of laboratory parameters at admission, hospital stay and discharge**

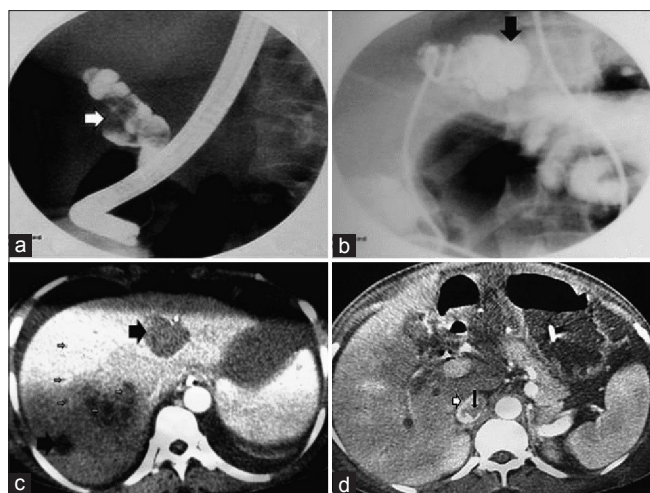
Parameters	At admission	Day 10 <sup>th</sup>	At discharge
Hemoglobin	6.7 gm/dl	8.8 gm/dl	9.5 gm/dl
WBC	16.0×10 <sup>3</sup> /ul	18.0×10 <sup>3</sup> /ul	7.32×10 <sup>3</sup> /ul
Differential	N80 L12	N82L15	N72L14
Platelets	49×10 <sup>3</sup> /ul	95×10 <sup>3</sup> /ul	1.49×10 <sup>3</sup> /ul
BUN	82 mg/dl	30 mg/dl	22 mg/dl
Creatinine	1.6 mg/dl	0.60 mg/dl	0.5 mg/dl
Total Bilirubin	22.3 mg/dl	11.5 mg/dl	6.7 mg/dl
Direct Bilirubin	14 mg/dl	7 mg/dl	4 mg/dl
Aspartate transaminase	48 unit/l	99 unit/L	45 unit/L
Alanine transaminase	42 unit/L	41 unit/L	20 unit/L
Alkaline phosphatase	1,124 unit/L	1,380 unit/L	1,117 unit/L
Albumin	2.5 gm/dl	2.7 gm/dl	3.1 gm/dl
Prothrombin time	12	22	28
INR	1	1.42	2.60



**Figure 1:** USG abdomen showing Hydatid Cyst (6.2 × 6.0cm) in left lobe of liver with floating membranes inside (white arrow). Also, dilated CBD with membranes inside (black arrow)

venacava suggestive of thrombus [Figures 2c and d]. Doppler study revealed thrombotic occlusion of inferior venacava, right portal vein, and main portal vein along with portal hypertension [Figure 3a]. Patient was started on heparin drip. On day 8<sup>th</sup>, patient's condition begins to deteriorate again with worsening jaundice, leukocytosis, and increasing alkaline phosphatase [Table 1]. An elective ERCP demonstrated bulging papilla of vater with protruding membranes. The common bile duct was dilated 20 mm with numerous leaf-like filling defects. A papilotomy was performed and copious amount of pus with hydatid material was evacuated [Figure 3b and c].

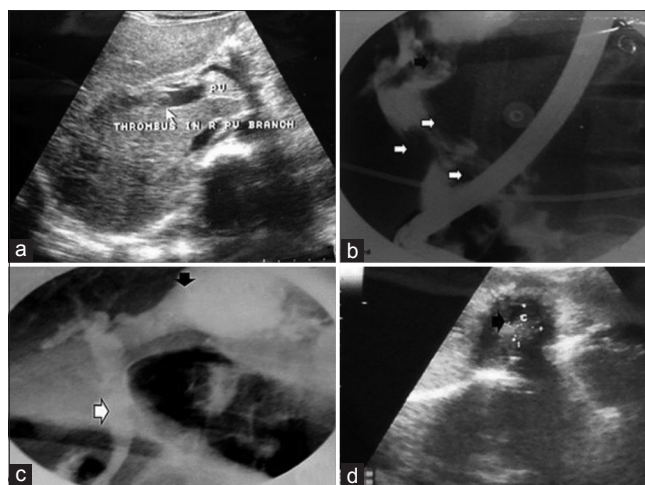
Although, fever, leukocytosis, and features of cholestasis improved after ERCP, but her abdominal pain and ascites



**Figure 2:** (a) Initial ERCP showing dilated common bile duct with multiple filling defects. (b) ENBD cholangiogram revealed large cavity in left lobe of liver communicating with left hepatic duct. (c) CT Abdomen with contrast showing well defined cystic lesion in left lobe of liver, multiple small hepatic abscesses. (d) CT Abdomen with contrast showing filling defect in inferior venacava (small black arrow). Compression on IVC by enlarged liver (small arrow)

progressively worsened. A peritoneal tap was performed and ascitic fluid analysis was negative for scolices and hooklets with serum ascitic albumin gradient (SAAG) of 1.3 g/dL suggestive of a transudate likely due to portal hypertension secondary to portal vein thrombosis. As the thrombosis was acute and irreversible even after decompression of liver, and patient was high risk for bleeding due to low platelets and anticoagulant use. It was, therefore, decided to attempt catheter-directed thrombolysis to prevent long-term complications of inferior venacaval and portal vein thrombosis and risk of bleeding. Using transfemoral catheter directed approach, 250,000 units of streptokinase was given followed by infusion of 100,000 units/hour for 36 hours. No complication was encountered during or after the procedure and patient was monitored in intensive care unit. Subsequent to thrombolysis, abdominal pain, ascites and portal hypertension significantly improved and patient was discharged on albendazole for 3 months and oral warfarin for 6 months. On follow up, repeat sonography revealed decrease in the size of hydatid cyst cavity suggestive of patient is improving with no disease recurrence [Figure 3d].

Despite the rise in prevalence of Echinococcosis, it still remains a very rare disease (<1 case per 1 million inhabitants) in United States. With intrabiliary rupture, the classic triad of jaundice, biliary colic, and fever occurs but ascites and lower extremity edema is rare; however, it can be the presentation of thrombus in either portal vein or inferior venacava. The Budd-Chiari syndrome (BCS) indicates an obstruction of hepatic venous outflow at any level from the small hepatic veins to the junction of the inferior vena cava (IVC) and the right atrium.



**Figure 3:** (a) Doppler ultrasound study of abdomen revealing thrombotic occlusion of portal vein. (b) ERCP showing CBD dilated 20 mm with numerous filling defects (white arrows) and filling defects in left hepatic duct (black arrow). (c) Cholangiogram showing dilated CBD (white arrow) and communication between cyst and left hepatic duct (black arrow). (d) Follow up USG abdomen showing decrease in size of cyst cavity with no membranes or daughter cysts (black arrow)

Acute form of BCS is a rare condition and up to 15% of cases are complicated with portal vein thrombosis (PVT). Prognosis in patients with Budd-Chiari syndrome with portal vein thrombosis is poor.<sup>[1]</sup> In Western countries, its spontaneous mortality is reported to approach 70% at 1 year and 90% at 3 years.<sup>[2]</sup> Therefore, early diagnosis and treatment is necessary.

The initial screening test of choice is ELISA (sensitivity 80%). Sensitivity of Ultrasound abdomen and ERCP for diagnosis of intrabiliary rupture of hydatid cyst is almost 70% and 90%, respectively. ERCP is gold standard for diagnosis.<sup>[3]</sup> To evaluate inferior venacaval and portal vein thrombosis, ultrasound and contrast enhanced triple phase computerized tomography (CT) are considered primary diagnostic modalities. Unlike venography, they can accurately demonstrate the cephalic extent of thrombus and assessment of other surrounding viscera.<sup>[4]</sup> USG is best and safest means to follow up such patients.

Endoscopic therapy appears to be an effective mode of treatment for biliary fistulas complicating liver abscesses and cysts.<sup>[5]</sup> Preoperative ERCP is very helpful in patients with cysto-biliary fistula and, in selected cases, it can solve the problem without further surgical intervention.<sup>[6]</sup> Although most of these cases have been managed with decompression by ERCP or Puncture Aspiration Irrigation and Reaspiration (PAIR) followed by surgical resection of hydatid cyst which also led to the resolution of inferior venacaval thrombosis as well.<sup>[4,6]</sup> Interestingly, in our case cyst decompression with ERCP alone did not lead to resolution of IVC thrombosis and portal hypertension. Thus, thrombolysis was performed.

A number of thrombolytic agents, such as urokinase, streptokinase, and tissue plasminogen activator have been administered. Thrombolysis is more likely to be successful in acute thrombosis and when the thrombolytic agent is locally infused into a hepatic vein and/or inferior venacava.<sup>[7]</sup> Recently a quick and easy thrombolytic approach; that is, agitation thrombolysis for the treatment of fresh thrombosis have been devised.<sup>[8]</sup>

There are a few case reports in the literature describing the benefits of systemic thrombolysis, whereas, few investigators have used combined local and systemic thrombolysis.<sup>[9]</sup> Although we used local thrombolysis for inferior venacava thrombosis, possibly its systemic effect led to resolution of portal vein thrombosis. Vascular access may be attempted via the transjugular or transfemoral route when inferior venacava interventions have to be performed. Mostly, the femoral route is preferred as negotiation is easier and risk of bleeding is less compared to transhepatic approach. Transhepatic route has proved to be a safe and effective method in the treatment of portal vein thrombosis. But it carries a risk of post-procedure uncontrollable bleeding through the transparenchymal tract after removal of the catheter and is increased in patients with ascites, coagulopathy or those on anticoagulant therapy.<sup>[10]</sup> This is the reason transfemoral approach was chosen in our patient.

In conclusion, as yet there is no consensus on treatment guidelines for patients with intrabiliary rupture of hepatic hydatid cyst complicated with inferior venacava and portal vein thrombosis. Keeping in view Echinococcosis is not a malignant disease; consideration should be given to all nonsurgical therapeutic approaches before contemplating operative procedures.

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