Intrabiliary rupture of hydatid cyst in a patient with cirrhosis

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Abstract
A case of 48-year-old male, hepatitis B cirrhosis, hepatic hydatid cyst, jaundice, fever, chills suffered from severe abdominal pain in the right upper quadrant. He was suffered from acute cholangitis and spontaneous bacterial peritonitis, and received intravenous antibiotics but his condition rapidly deteriorated to sepsis and severe hepatic failure. The presence of dilated Common Bile Duct (CBD) containing small cystic lesions suggesting daughter cysts on ultrasonography, which was further verified by Endoscopic Retrograde Cholangiopancreatography (ERCP), along with significant eosinophilia and positive serology test for hydatid cyst, made the diagnosis of intrabiliary rupture of hydatid cyst definite. We performed a delayed endoscopic sphincterotomy which resulted in complete resolution of the clinical picture. The patient was treated with Albendazol and Lamivudin and was referred for surgery.

Keywords
hydatid cyst, cirrhosis,
ture, the concurrence of the two mentioned dis-
eases has not been reported. The rapid deterio-
ration of cholangitis to severe sepsis accompa-
nied by significant hepatic decompensation
created various clinical challenges that made
the management of this case problematic. The
rapid and complete resolution of clinical pic-
ture along with the improvement in biochemi-
cal tests after performing endoscopic sphincter-
omy, reinforces the fact that endoscopic retrograde cholangiography with sphincteroto-
my has been successful as the sole and defini-
tive means of treatment of intrabiliary ruptured
hydatid cysts [1].

Case presentation
A 48-year-old male, known case of chronic
hepatitis B, was admitted to the emergency
room of Imam hospital for severe abdominal
pain in the right upper quadrant, jaundice, fever
and chills. He had an eight-year history of
vague intermittent pain in the upper abdomen
for which, he had been evaluated in his home-
town, and diagnosed with hepatic hydatid cyst
but refused the prescribed treatment of Alben-
dazol and surgery. He was never evaluated or
treated for hepatitis B. He was otherwise
asymptomatic, until twenty days before admis-
sion, when he noticed dark discoloration of
urine and pale stool leading to overt jaundice
within 10 days, followed by a constant pain in
the right upper quadrant, mild abdominal dis-
tension, fever and chills for two days prior to
admission.

The social history revealed him as a 48-year-
old married farmer, father of three, who was
born and lived in the city of Gonbad, Golestan
province. He consumed alcohol occasionally,
did not smoke and denied use of any medica-
tions or illicit drugs. His family history was un-
remarkable.

Investigation
Physical Examination at admission showed a
fully conscious middle-aged male with an oral
temperature of 38.9 °C, a pulse rate of 120 per
minute, and blood pressure of 110/70 mmHg.
The conjunctiva appeared pale and the face and
trunk were icteric. Multiple spider hemangi-
omas of varying sizes were found on the up-
per chest and back. The abdomen was mildly
distended with moderate tenderness on palpa-
tion of the right upper quadrant. The spleen was
apalpable about 4 centimeters below the left
costal margin. The liver span measured about 7
centimeters on percussion. Other physical ex-
aminations were normal.

Laboratory findings indicated anemia (Hb:
10.5, MCV: 87), marked leukocytosis (WBC:
20300/mm³, 75% PMNs) and thrombocytope-
nia (68000). Liver function tests were abnormal
with elevated alkaline phosphatase (545), mild-
ly elevated Aspartate and Alanine transaminase
(49.3 and 51 respectively), conjugated hyper-
bilirubinemia (total bilirubin: 13.7 and direct
bilirubin: 8.19), prolonged prothrombin time
(19, INR: 1.9) and hypoalbuminemia (2.7). Ul-
trasonography performed in the emergency
room showed decreased liver size, with dilated
CBD (14mm). The gallbladder had normal wall
thickness and contained no stones. A cystic mass with mixed echogenicity was noted in the right liver lobe, measured 73.58 mm which was highly suggestive of hydatid cyst. Spleen had a diameter of 20 cm. Minimal ascites fluid was present in the pelvis. The ascites fluid was drained and the result of laboratory examinations was indicative of spontaneous bacterial peritonitis with white blood cell count of 700 (50% PMN), total protein of 3 gr/dl, negative smear and culture for bacterial infection.

**Differential diagnosis**

HepatoCellular Carcinoma, because the patient was cirrhotic but AFP was normal and cystic mass.

**Treatment**

Due to acute suppurative cholangitis and spontaneous bacterial peritonitis, intravenously antibiotic regimen of Ceftriaxon and Metronidazole was administered. 48-hours after initiation of treatment, the clinical picture and test results deteriorated toward overt sepsis and decompensated cirrhosis. The patient was slightly lethargic, remained febrile (oral temperature: 39.2 °C), and developed massive tense ascites leathing to abnormal breathing with generalized abdominal tenderness. Biochemical tests showed leukocytosis (WBC: 36800, PMN: 80%) and eosinophilia (15%), total and direct levels of bilirubin reached 30.6 and 12.5 respectively. The patient was then transferred to the gastroenterology department of Imam hospital. The ultrasonography was repeated which revealed the following data. Liver appeared smaller than normal, with increased echogenicity and nodular surface. A cystic lesion with an echogenic center, containing multiple small cysts was noted in the right liver lobe, measuring 62.58 mms, which was strongly suggestive of hydatid cyst. The Common bile duct (CBD) was dilated (14 mm), and multiple echogenic centers without posterior shadow were noted in the common bile duct suggestive of biliary
sludge. The spleen was measured 187 mm, and abundant ascites fluid presented in the pelvis and abdomen. The ascites fluid was drained therapeutically and the result of ascites fluid analysis persistently met the criteria of peritonitis (WBC count: 1200, PMN:80%) with negative bacterial smear and culture. Therefore, in the absence of clinical and laboratory improvement, the intravenous antibiotic regimen was changed to Imipenem and Vancomycin. An emergent endoscopic retrograde cholangiopancreatography (ERCP) was scheduled and then cancelled due to prolonged pro-thrombin time (INR: 2.9). We performed endoscopic ultrasonography (EUS), while the patient received fresh frozen plasma to correct the coagulation status. The EUS results showed a dilated CBD (13mm), containing sludge debris and multiple cystic images without acoustic shadow, which may represent daughter cysts. In addition, the serology test for hydatid cyst antibody (IgG) was positive. These findings raised a very high suspicion of frank intrabiliary rupture of hydatid cyst, which was ignored. When INR reached 1.4, ERCP and sphincterotomy were done and the findings showed multiple irregularly-shaped filling defects in the CBD and a round cystic lesion which was may communicated with the biliary system, with mild distal stricture and dilatation of intrahepatic ducts. The comprehensive information provided by ERCP, presented conclusive evidence of the presence of daughter cysts in the CBD, demonstrated the cystobiliary fistula and above all,
served as an excellent therapeutic modality. Within 24 hours after ERCP, the patient became afebrile and his general condition improved significantly. Due to a hepatitis B viral load of more than 100 and decompensated cirrhosis, we started Lamivudin treatment. A surgery consult was done and the patient chose to have the surgery in a hospital in his hometown. Therefore with the existing Albendazol treatment, the patient was referred to a surgery centre and then discharged him based on his personal consent.

Discussion

Although hydatidosis is a benign disease, it can produce serious complications as portal hypertension in countries where the disease is endemic. Portal hypertension has been reported secondary to obstruction of inferior vena cava and hepatic outflow tract or extrahepatic presinusoidal portal hypertension caused by extrinsic compression of the liver by an hydatid cyst. Hydatosis in the previously cirrhotic patients is rare because liver parenchyma is fibrotic and growing cyst within the liver is difficult although if infection occurred may be severe due to several factors including: impaired humoral and cellular immune responses to the organism, nutritional deficiency and decompensation of cirrhosis [3].

In this case intrabiliary rupture of large hydatid cyst in cirrhotic liver was occurred which is a rare complication. The endoscopic sphincterotomy is the procedure of choice that resolves biliary obstruction and cholangitis. The cholangitis treated by liver transplantation might produce a good long-term outcome since the patient was child c but yet presented a technical challenge related to mortality and risk of procedure. Other challenge in prescribing albendazole for cirrhotic patients with hydatosis elevated aminotransferase levels and leucopenia, may required discontinuing albendazole.

In review of published cases no case of hydatid cyst in previously cirrhotic patient was reported

Learning points

"Frank intrabiliary rupture of hydatid cyst is a relatively rare, but very serious cause of obstructive jaundice and/or cholangitis and must be among differential diagnosis especially in endemic areas or in proper clinical settings.

"The diagnosis of intrabiliary rupture of hydatid cyst, may be challenging. Ultrasonography may suggest the diagnosis of a frank intrabiliary rupture in many of the cases. As evidenced in our patient, the dilatation of the biliary tree and the presence of small cystic images in CBD which may represented daughter cyst, a very specific sign of rupture, but not sensitive. Nonshadowing material in the CBD is more often seen and most probably with the expression of hydatid sand; however is not very specific being present in case of biliary sludge, blood clot or inflammatory sludge. The CT-scan, MRI, MRCP, EUS and ERCP provide more comprehensive information for patients with biliary hydatid cyst.

"Endoscopic sphincterotomy has proven to be an excellent alternative treatment for patients with biliary hydatid cyst, especially if risk of surgery is high.

"The optimal treatment, especially in the presence of a cystobiliary fistula, is surgery.

References


