Resolution Of Hepatic Hydatid Cyst Following Spontaneous Rupture Into The Biliary Tree And Subsequent Endoscopic Extraction
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Citation

Abstract
We report an interesting case of hepatic hydatid cyst which imaging modalities misinterpreted as amoebic liver abscess and later presented with rupture into the common hepatic duct and caused obstructive jaundice. The cyst resolved completely following endoscopic papillotomy and clearance of the membranes endoscopically. The role of magnetic resonance cholangiography and photo-documentation of the course of the disease is reported. A review of literature is also presented.

INTRODUCTION
The most common site of hydatid cyst formation is the liver. Hydatid cysts of the liver exert pressure on the surrounding parenchyma, and in approximately one-fourth of the cases, due to higher pressure in the cyst, the cysts eventually leak into small bile ducts or perforate into large ones. Thus the most common complication of hydatid cyst of the liver is spontaneous rupture into the biliary tract with biliary obstruction being reported to occur in 5% to 17% of cases. Intrabiliary rupture occurs into the right duct in 55–60% of cases, into the left duct in 25–30% and rarely into the confluence or gall bladder. We present a case of a hydatid cyst of the liver which ruptured spontaneously into the common hepatic duct and resolved completely following endoscopic papillotomy and repeated endoscopic clearance of the membranes in the common bile duct.

CASE REPORT
A twenty six year old male patient presented to the surgical emergency with history of high-grade fever and right hypochondrial pain of one week duration. The patient was icteric and had a tender hepatomegaly. Investigations showed a deranged liver function test (LFT) with a total bilirubin of 7.0mg/dl (direct of 6.5 mg/dl) and raised serum alkaline phosphatase (43KA units). An ultrasound of abdomen reported a heterogeneous hypodense lesion measuring 8.7 x 7 cm in right lobe of liver suggestive of liver abscess. CAT scan revealed a moderate hepatomegaly with large well defined abscess in the right liver lobe with an air-fluid level and pneumobilia suggestive of a communication with the non dilated intrahepatic biliary radicals (Fig 1).

Figure 1
Figure 1: CAT scan - axial section shows a thin walled cavity in the right upper lobe with an air fluid level

A small right pleural effusion was also reported. Attempt at needle aspiration of the liver abscess failed, while aspiration of the pleural effusion yielded 70 ml. of straw colored exudative fluid. Amoebic serology was strongly positive. The patient responded to antiamoebic drugs and serum bilirubin level fell to 2.1gm/dl. The patient was subsequently discharged.

The patient again reported to the surgical out patient department after fifteen days with history of fever, upper
abdominal pain and increased jaundice of five days duration. Investigations revealed a total bilirubin of 9.5 mg/dl (direct bilirubin 5.2 mg/dl) and serum alkaline phosphatase levels 38 KA units. An abdominal ultrasound revealed a hypoechoic lesion with multiple internal hyperechoic bands suggestive of hydatid cyst with ruptured membranes measuring 12 x 13 cm in right lobe of liver segment 7 and 8 extending into distal common bile duct (CBD) till the pancreatic head. The CBD measured 13 mm and there was intrahepatic bile duct dilatation. Magnetic resonance cholangiography (MRC) revealed an oval 12 x 10 cm well marginated thin walled cystic lesion in right lobe of liver suggestive of hydatid cyst (Fig 2).

**Figure 2**
Figure 2: MRI coronal section (T2W MRI) shows linear serpinginous shadows within the fluid filled cyst in the right upper lobe, suggestive of collapsed membranes (arrow). The dilated left main hepatic duct can be seen.

The cyst was seen to communicate inferiorly with common hepatic duct in the region of ductal confluence with extension of the hydatid membranes into common hepatic duct (CHD) and proximal CBD with upstream dilatation of the right and left hepatic ducts and intra-hepatic biliary radicles (Fig 3).

Hydatid Serology was found to be positive. The patient underwent endoscopic retrograde cholangiography (ERCP). During the procedure membranes were seen to be coming out of papilla of Vater and on injection of the contrast, communication of the bile duct with the cyst was established (Fig 4).

**Figure 3**
Figure 3: In the MRC scan the cyst can be seen to communicate with the common hepatic duct. The membranes can be seen entering the biliary duct.
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Figure 4

Figure 4: ERCP film showing the communication of the cyst with the biliary tract. The common bile duct is full of membranes and could not be delineated completely. Some intrahepatic radicles are also seen to fill up.

An endoscopic papillotomy was performed and a large amount of the membranes were removed from the CBD. The procedure was completed with CBD stenting and nasobiliary drainage (NBD). A repeat ERCP performed after four days showed the cavity to have reduced to around 2 cm and during the procedure more membranes were removed from the CBD. The resolving status of the cyst was also documented by MRCP. (Fig 5)

(LBD: left bile duct; CBD: common bile duct; GB: gall bladder; Duo: duodenum)

The patient recovered subsequently with decrease in jaundice, fall of total serum bilirubin to 2.8 mg/dl and decrease in size of cavity sonographically. During this period the NBD was draining around 300-400 ml. The patient refused further in-patient treatment and left the hospital with the biliary stent still in situ.

After three weeks the patient again reported to the out patient department with a decreased NBD output and high grade fever, increasing jaundice and a tender 10 x 10 cm lump in the right hypochondrium, extending on to right lumbar region and continuous with liver dullness. Investigations revealed total leucocyte count of 16500/mm3, total bilirubin of 5.5 gm/dl, and serum alkaline phosphatase of 33 KA units. A repeat ultrasound showed the cyst cavity
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to have increased in size to 12 x 13 cm with the stent still inside the CBD. An ultrasound guided pig tail catheter was inserted into the cavity and approximately 400 ml of pus was drained and the patient was started on broad spectrum antibiotics. The patient improved markedly and became asymptomatic within five days. A cavitogram after 7 days revealed a small 3x4 cm cavity communicating with CHD with membranes inside the CBD and stent in situ. (Fig 6)

**Figure 6**
Figure 6: Cavitogram contrast injected through the pigtail catheter into the cavity shows communication with the dilated biliary tree. Linear filling defects can be seen within the dilated bile duct due to the membranes within it. (RHD: right hepatic duct; LHD: left hepatic duct; CHD: common hepatic duct; GB: gall bladder; CBD: common bile duct)

The patient underwent an endoscopic clearance of the CBD 15 days after admission when the pigtail drainage was nil. A completion ERCP established a complete clearance of the CBD with no demonstrable membranes in the cyst cavity. The biliary stent was removed and the patient was discharged after removal of the pigtail catheter. The patient was doing well in the follow up period with no residual cavity sonographically four weeks after discharge. The patient remained asymptomatic during nine months of follow up.

**DISCUSSION**

Hepatic hydatid disease (HHD) is a major endemic problem in sheep-rearing regions of the world. The liver acts as a filter for hydatid larvae, making it the most commonly affected organ. The liver is involved in 50% to 70% of patients with hydatid disease. Symptoms arise either when the cyst has grown enough to cause pressure on adjacent organs or when a complication occurs. Rupture, secondary infection, and suppuration are the most common complications. Up to one-third of patients with HHD present with complications such as rupture (into the biliary tree, thorax or peritoneum), secondary infection, anaphylactic shock, sepsis and liver replacement. Cysts of the liver exert pressure on the surrounding parenchyma, and in most cases the cysts eventually rupture into the biliary tree in 1% to 25% of cases although an incidence of 64.75% has been reported from a multicentric study in Tunisia. It occurs in two forms: an occult rupture, resulting from small tears between the cyst wall and small biliary radicles, seen in 10% to 37% of the patients, or a frank rupture, involving large bile ducts, seen in 3% to 17% of the patients.

Obstructive jaundice occurs when cyst elements empty into the biliary tree. Rupture is most likely to occur in centrally located cyst with a high intracystic pressure up to 80 cm H2O. According to Lewall and McCorkell the cyst rupture can occur in three clinical forms: contained, communicating and direct. Contained rupture occurs when the cyst contents are confined within the pericyst. Communicating rupture defines tearing of the pericyst and evacuation of cyst contents into the biliary tract or bronchioles. Direct rupture describes complete tear of the cyst wall and spillage of the cyst contents into the peritoneal or pleural cavity. Although biliary obstruction is reported to occur in only 5% to 17% of cases, obstructive jaundice occurs in 57% to 100% of cases following intrabiliary rupture, especially when the rupture occurs into the large bile ducts thus emptying the contents into the biliary tract. It has been reported that if the cystobiliary opening is larger than 5 mm, cystic content migration into the biliary tract would occur in 65% of the cases. Other presentations are right upper abdominal pain (82%), fever (70–90%), acute cholangitis (20–37%), abdominal lump (22–39%), and rarely with acute pancreatitis, liver abscess or septicaemia; or in 5% to 6% it may be asymptomatic.

Early diagnosis and proper management are mandatory in these patients, since serious clinical complications with an increased mortality may ensue. The reported morbidity and mortality rates of all patients in literature are 19.44-43.03% and 1.8- 4.5% respectively. The most common causes of deaths were sepsis and hepatic failure. Ultrasound is the most commonly employed initial investigation. Gharbi et al. have described five types of echinococcal cysts on USG: purely cystic except for hydatid sand; detached membrane; multiseptated; peripheral or
diffuse distribution of coarse echoes in a complex heterogeneous mass; and calcified wall. These five types are believed to correspond to evolutionary stages of the hydatid cyst.

Computed tomography and magnetic resonance imaging are useful in diagnosing biliary hydatid disease resulting from rupture of a liver hydatid cyst; MRI is a useful tool in difficult cases such as intrabiliary rupture, where CT and ultrasound are not conclusive. However, ERCP and, more recently, MRCP, are confirmatory for intrabiliary rupture of the hepatic hydatid cyst. In addition ERCP may be of therapeutic benefit in selected cases.

The classic treatment for hydatidosis with rupture into the biliary tract has been surgery. However, persistent postoperative external bile drainage is a serious complication, occurring in 3.8% to 27.5% of cases, that often necessitates reoperation. Other complications include infection within the residual cavity and sinus formation; recurrence or dissemination can also occur. However, since al-Karawi et al reported the use of ERCP with endoscopic sphincterotomy and extraction of retained echinococcal daughter cysts from the common bile duct, endoscopic sphincterotomy with extraction of the cysts from the CBD has emerged as a safe and an effective alternative treatment for patients with intrabiliary rupture of hepatic hydatid cysts. It has been suggested that two to three courses of mebendazole may play an important role in prophylaxis after surgery or endoscopic evacuation of a ruptured cyst into the biliary tree.

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