A non-klatskin tumor: A case report and review of intrabiliary hydatid cyst rupture

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ABSTRACT

INTRODUCTION: A 64-year-old lady was diagnosed with having a klatskin type 3A tumor based on imaging, however, an alternative diagnosis was achieved during surgery.

PRESENTATION OF CASE: We present a case of a 64-year-old lady who presented for new-onset jaundice and was diagnosed with type 3A klatskin tumor based on MRCP findings. During surgery, it was revealed that the obstruction was caused by a frank intrabiliary hydatid cyst perforation. Cholecystoscopy with irrigation, cholangiography, and removal of the mother cyst were performed, and an end-to-end biliary anastomosis over a t-tube was then done. The patient tolerated the intervention and recovered well.

DISCUSSION: Hydatid cyst disease of the liver usually follows a benign course, however, intrabiliary rupture is one of the common complications associated with this disease. Intrabiliary rupture is classified into either frank or occult. Frank perforation, which is more common, is when hydatid material passes into the biliary ducts, and it may cause biliary obstruction and cholangitis with a high mortality rate. Occult perforation is when the hydatid cyst becomes infected itself, which usually leads to a silent presentation, and may only cause signs of suppuration. Diagnosis is usually achieved by imaging and relevant history. Treatment consists of medical and surgical intervention. Intraoperative cholangiography, cholecystoscopy, and t-tube drainage are recommended during surgery for frank rupture.

CONCLUSION: Intrabiliary hydatid cyst perforation can mimic cholangiocarcinoma and must be considered as an alternative diagnosis in these patients prior to surgery.

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1. Introduction

Hydatidosis of the liver usually causes no grave complications. Nevertheless, Intrabiliary rupture is not an uncommon occurrence [1]. Previous authors have classified intrabiliary rupture into either frank rupture, with possible obstruction and cholangitis, or occult rupture, which causes minimal symptoms. We report a case that was originally diagnosed with an obstructive klatskin tumor based on the general presentation and the MRCP results. However, during surgery, an alternative diagnosis was made.

This case was reported in accordance with the SCARE criteria [2].

2. Case presentation

A 64-year-old lady with no significant medical history presented to our ED complaining of new-onset jaundice. She reported recurrent dull right upper quadrant pain for the past 3 months. Upon examination, she was afebrile, jaundiced, and the abdominal exam was positive for Murphy’s sign. Biliary obstruction was suspected and labs and imaging were ordered accordingly. Her white count was 13,700 /µL and CRP was 81 mg/L. Bilirubin was elevated to 6.97 mg/dL (Direct 5.07 mg/dL), and liver enzymes followed a cholestatic pattern, with an Alkaline phosphatase of 939 IU/L, γ-GT 1245 IU/L, and ALT 1261 IU/L. MRCP was ordered to identify the obstruction, and it showed a 30 × 17.5 mm intraluminal lesion in the right hepatic duct, extending to the hepatic bifurcation. The finding was suggestive of a type 3A klatskin tumor (Fig. 1). The MRI also noted the presence of a simple 3 cm biliary cyst in segment V close to the right hepatic duct (Fig. 1). No evidence of metastasis or local invasion was seen on the MRI. A CT scan of the abdomen showed a localized mass in the biliary tree without any local invasion. Further metastatic workup included a PET scan and a chest CT scan showed no evidence of metastasis. Our surgery team was consulted, and the tumor was considered resectable. After a discussion between radiology, oncology, gastroenterology, and surgery teams, we decided not to perform an ERCP preoperatively due to the high likelihood of malignancy and lack of added benefit in that case. The patient was prepared for a right hepatectomy, right and common bile duct resection, and left roux-en-y hepatojunostomy.

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During surgery, a right subcostal incision was performed, followed by dissection until reaching the abdominal cavity, and identifying the hepatic duct. Full dissection of the right portal vein was difficult due to inflammation, so we decided to cut the common bile duct (CBD) close to the duodenum to have better exposure. Instantly, multiple daughter hydatid cysts, cystic fragments, and whitish fluid were ejected from the proximal part of the hepatic duct (Fig. 2).

We immediately alerted the anesthesia team to the occurrence and ordered ceftriaxone to wash the surgical field. After removal of all the debris and washing with ceftriaxone, further suction was done and more debris was removed from the proximal and distal parts of the resected CBD. Owing to the new findings, a diagnosis of intrahepatic hydatid cyst rupture was made, and we decided to abandon the hepatectomy after confirming the absence of a tumor. An intraoperative cholangiography was ordered and showed a small filling defect in the right hepatic duct. We decided to introduce a cholecystoscopy to confirm, and no tumoral lesions were seen. We were able to see some inflammatory debris in the right hepatic duct, which we removed by multiple lavages with normal saline and suction. In retrospect, the simple biliary cyst seen on MRI at admission was now considered the mother hydatid cyst, and we decided to remove it to prevent recurrence. The cyst was non-palpable and located deep in segment V. A hepatotomy was done, and only after a laborious search using ultrasonography were we able to find the cyst, puncture, and excise it. No obvious biliary communication was seen, and methylene blue test was negative. We decided to perform an end-to-end anastomosis of the CBD over a T-tube, even though the anastomosis was risky. We aimed to preserve the CBD in case future complications with the cyst were encountered and ERCP needed to be done. A drain was inserted in the cystic cavity, and another near the liver pedicle. The abdominal wall and skin were closed, and the patient was sent intubated to the ICU for monitoring.

The next day, the patient was extubated. The drains were draining a minimal amount of serosanguineous fluid, while the t-tube was dry. Bilirubin was 6.88 mg/dl on the first postoperative day, and the patient continued improving until it reached 2.39 mg/dl on the 6th postoperative day, and all drains except the t-tube were removed. A cholangiography through the t-tube revealed no obstruction and no leaks (Fig. 3), so we decided to discharge the patient on albendazol and remove the t-tube after 2 months.

3. Discussion

Liver hydatid cyst disease is a parasitic disease caused by the larval form of Echinococcus granulosus, mostly observed in endemic areas, such as the Mediterranean, New Zealand, Australia, South America, Central America, and Asia [3]. The organism can cause cyst formation in the kidneys, spleen, and other sites, however, the liver and lungs are the most commonly affected locations (55% and 45% respectively) [1].

Hydatid cyst disease usually has a benign course in the liver, however, some patients might develop unexpected complications. Anaphylaxis, cyst infection, thoracic extension, vascular complications, communication with skin or digestive tract, free intraperitoneal rupture, and Intrabiliary rupture have all been documented previously in the literature [1,3]. Intrabiliary rupture is reported in 6.1–17% of patients with liver hydatid cysts making it one of the common complications of this disease [4].
Hydatid cysts close to the biliary ducts can grow adjacent to the biliary ducts, eventually developing a communication. John R. Hankins in 1963 [5] was the first to classify intrabiliary rupture of hydatid cysts into either frank rupture or occult rupture. Frank rupture was described as the overt passage of hydatid material into the biliary tree, while occult rupture, on the other hand, is when the hydatid cyst itself develops a bacterial infection (suppurating hydatids) when bacteria gains access to the cyst through very small cysto-biliary communications. Although occult perforation entails a cysto-biliary communication, this communication is usually not enough to allow hydatid material to pass to the biliary tract and cause obstruction. Therefore, some authors like Dagher and Hovnanian [6] don’t consider occult rupture as true intrabiliary rupture of hydatid cyst, and believe that the diagnosis of intrabiliary rupture should be limited to cases where actual hydatid material is observed in the biliary tree [5]. This lack of consensus on classification and terminology has been (and will be) problematic in reporting and discussing intrabiliary rupture of hydatid cyst, where a lot of inconsistencies in definitions were observed when reviewing the literature related to this case.

According to Atli et al. [7], occult rupture seems to be more frequent than frank rupture. In both frank and occult rupture, nausea and vomiting are the most commonly reported complaints, and the duration of symptoms ranges between 1 month and 3 years with a median duration of 4 months before diagnosis.

Frank rupture can possibly cause symptoms of biliary obstruction over weeks to months [5] with obstructive jaundice and abdominal pain. This can, of course, progress into cholangitis and sepsis, with a high associated mortality rate [7]. Some authors have also reported pancreatitis and acute cholecystitis as complications of frank intrabiliary rupture. Occult rupture, on the other hand, has a less severe presentation, with patients usually complaining of pain, abdominal tenderness, and fever. The presentation can even be silent many times, however, it may progress to frank perforation at any stage [7]. Based on the presentation and our intraoperative findings, we believe that the case reported here is of a frank rupture causing acute biliary obstruction without cholangitis.

Ultrasonography and CT scanning are usually enough to diagnose liver hydatid cyst disease and identify its complications [1]. MRCP may be more valuable for diagnosis, however, the lack of experience in its use to identify hydatid cyst complications may limit its role [7]. On MRCP, radiologists may be able to see the cyst's intense rim, detachment of the membrane, daughter cysts, and hydatid material obstructing and dilating the biliary tree [4]. Nevertheless, in our case, MRCP was highly consistent (even in retrospect) with a simple liver cyst and the intrabiliary lesion could not be identified as a hydatid cyst.

ERCP can be useful for both diagnosis and treatment of intrabiliary rupture, especially in acute situations like cholangitis, allowing for proper surgical planning and elective definitive surgery later on. It was also shown that a complete cure can even be achieved if the biliary tract and the cyst are both evacuated during ERCP, eliminating the need for surgery [1]. It has also been reported that performing sphincterotomy during ERCP can reduce the occurrence of post-operative external fistulae.

According to Dziri and colleagues’ analysis [1], the current evidence supports the surgical approach as the best treatment for intrabiliary hydatid cyst rupture. The recommended procedure includes, besides removal of the mother cyst, an intraoperative cholangiography and choledoscopy followed by t-tube drainage after clearing the biliary tract. Based on the findings of Dagher and Hovnanian [6] and Hankins [5], removal of the mother cyst is essential for treatment, and if not done, there’s a high possibility of post-operative deterioration and recurrence. For occult rupture (suppurating hydatids), partial cystectomy with evacuation and external drainage seem to be enough [7]. In all cases, finding and closing the cysto-biliary communication should be attempted to avoid the formation of external biliary fistulae [7]. Medical treatment with albendazole is also recommended for 3 months after surgery [1].

Intrabiliary rupture is not an uncommon complication of liver hydatid cysts, however, it may be challenging to achieve the correct diagnosis in patients with no history of hydatid cyst disease. There’s no clear consensus in the literature regarding the classification of intrabiliary rupture, which raises the need for a clear definition of the original terminology to avoid future confusion. Frank perforation can obstruct the biliary ducts and mimic the presentation and appearance of klatskin tumor on MRCP and CT scanning. Based on this experience, we believe that intrabiliary hydatid cyst rupture should be suspected in every patient presenting with an obstructive biliary tumor, and who is found to have a biliary cyst, even if the cyst appears simple and non-hydatid on imaging. In such situations, ERCP might prove valuable in providing the correct diagnosis preoperatively and possibly treating the condition.

Declaration of Competing Interest

This article has no conflict of interest with any parties.

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Consent

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