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Rupture of a liver hydatid cyst into the right portal vein leading to right portal vein thrombosis: a case report and literature review

Houssem Ammar[1], Nouha Dammak[1], Mahdi Ben Latifa[1], Mohamed Amine Said[1], Marwa Azri[1], Rahul Gupta[2], Habiba Ben Hamada[3], Oussama Belkacem[4], Mohamed Ben Mabrouk[1] and Ali Ben Ali[1]

Abstract

Hydatid disease (HD) is a worldwide parasitic disease. HD is endemic in many sheep- and cattle-raising areas, with a high prevalence of 5% to 10% in the Mediterranean region. Fistulation of liver hydatid cysts (LHC) in the bile ducts is the most common complication, followed by rupture of cysts in the peritoneal and thoracic cavities. Vascular complications are a rare complication of HD. We describe the case of a 70-year-old woman who was admitted with the chief complaint of pain in the abdominal right upper quadrant for 6 months. Abdominal computed tomography revealed a large LHC in the right liver that had ruptured into the right portal vein branch, with venous thrombosis. Intraoperatively, the right portal vein was opened longitudinally, and the hydatid contents were evacuated. Right hepatectomy was performed to completely excise the LHC. The penetration of a cyst into an adjoining vessel is very rare, and portal vein invasion by HD is extremely rare, with only 10 cases published in the literature, to the best of our knowledge.

[1]Department of Gastrointestinal Surgery, Sahloul Hospital, University of Sousse, Sousse, Tunisia
[2]Department of Gastrointestinal Surgery, Synergy Institute of Medical Sciences, Dehradun, India
[3]Department of Anesthesiology, Sahloul Hospital, University of Sousse, Sousse, Tunisia
[4]Department of Pathology, Sahloul Hospital, University of Sousse, Sousse, Tunisia

Corresponding author:
Houssem Ammar, Department of Digestive Surgery, Sahloul Hospital, 11 Route Ceinture, cité Sahloul, 4054, Sousse, Tunisia.,
Email: Drhoussemammar@gmail.com

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Introduction

Hydatid disease (HD) is a zoonosis caused by infection with the larval stage of *Echinococcus granulosus*.[1] HD poses a significant public health problem in Tunisia.[2] HD may infest any organ but has a particular predilection for the liver and lungs and is frequently located in the right lobe of the liver (60%–70%).[1] Untreated or inadequately treated liver hydatid cysts (LHC) may lead to potentially serious complications. The most common complication of LHC is cystobiliary communication[2] followed by rupture, secondary infection, and suppuration. In rare cases, LHC can lead to vascular involvement. This complication is infrequent but is associated with high morbidity and mortality.[3] Penetration of the cyst into an adjoining vessel is very rare, and portal vein invasion by HD is extremely rare,[4] with only 10 cases published in the literature, to the best our knowledge. Here, we present a case of an LHC that ruptured into the right portal vein and was treated successfully by surgery. Verbal consent for the publication of this report was obtained from the patient mentioned in this case. The reporting of this case complies with the CARE guidelines.[5]

Case report

Overall summary

A woman in her early 70s with hypertension and type 2 diabetes mellitus was admitted with a chief complaint of pain in the abdominal right upper quadrant for 6 months. She had a history of surgery for LHC 7 years earlier. The LHC was located in the left liver lobe and treated by deroofing with removal of the cystic contents. The postoperative course was uneventful.

Case details

On physical examination, a healed right subcostal scar with an incisional hernia was found. Palpation revealed abdominal right upper quadrant pain without a fever. The results of laboratory liver tests were normal, and the complete blood count (CBC) results were within the normal ranges. Hydatid serology was positive. Abdominal computed tomography demonstrated a large LHC measuring 10 cm located in liver segments V, VI, and VII. The cyst had loss of fat planes with the right portal branch and exovesiculation (Figure 1).

Considering these findings, right hepatectomy was planned. Written and verbal informed consent for treatment was obtained from the patient and recorded. Intraoperatively, we identified atrophy of the right liver lobe with compensatory hypertrophy of the left lobe. Additionally, there was a cystic lesion in the right lobe with no obvious protrusion. Parenchymal transection was performed until the right branch of the portal vein was visualized. Then, the main portal vein and left portal vein were looped and clamped. The right portal vein was opened longitudinally, and the hydatid contents were evacuated. A large fistula between the hydatid cyst and

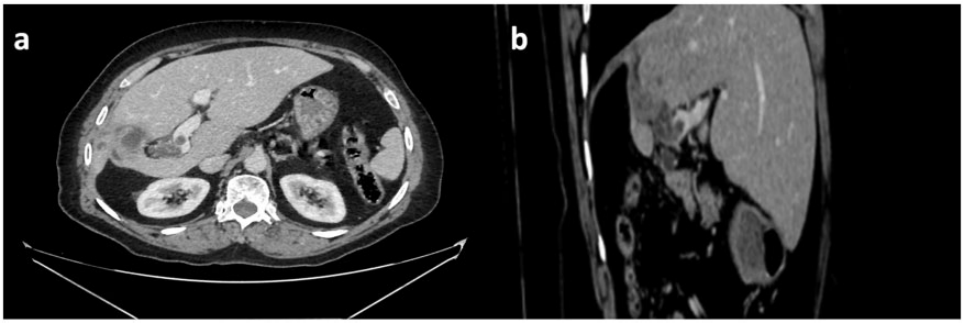

Figure 1. Abdominal CT showing a large LHC in the right liver. The cyst shows loss of fat planes with the right portal branch with exovesiculation on axial and sagittal sections (a and b)
CT, computed tomography; LHC, liver hydatid cyst.

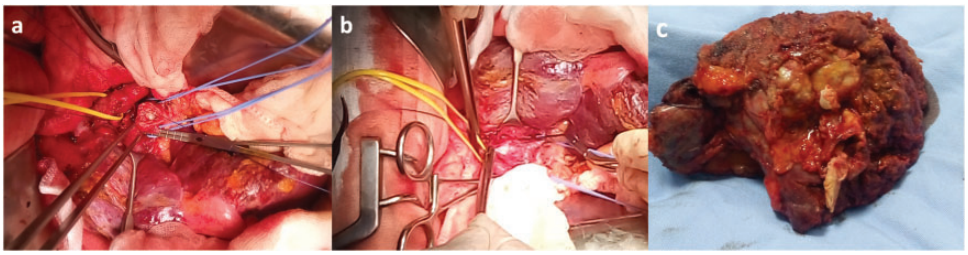

Figure 2. The main portal vein and left portal vein were looped and clamped. The right portal vein was opened longitudinally, which was thrombosed (a), and the hydatid contents were evacuated (b). Right hepatectomy with complete excision of the LHC, with exovesiculation (c). LHC, liver hydatid cyst.

the right portal vein was noted. Right hepatectomy was performed, which resulted in complete excision of the LHC with the exovesiculation, following injection of a 20% saline solution into the cyst (Figure 2). Transcystic biliary drainage was performed, and a drain was placed near the cut surface of the liver. The operative time and estimated blood loss were 130 minute and 400 mL, respectively.

Histopathological examination of the surgical specimen confirmed the diagnosis of LHC recurrence with cystobiliary fistula.

During the postoperative follow-up, the patient developed a right subphrenic hematoma, which was treated with antibiotics and image-guided percutaneous drainage. The patient was discharged on postoperative day 9. Subsequently, she was followed-up with abdominal CT and hydatid serology every 6 months. Currently, 2 years after the right hepatectomy, she is asymptomatic, with no sign of recurrence.

Discussion

HD is a worldwide parasitic disease. HD is endemic in many sheep- and cattle-raising areas, with a high prevalence of 5% to 10% in the Mediterranean region.[2] Fistulation of LHCs in the bile ducts is the most common complication, followed by rupture of cysts in the peritoneal and thoracic cavities.[2] Vascular complications are a rare complication of HC,[4] and very few cases of vascular involvement in LHC have been reported in the literature (Table 1). The spontaneous rupture of an LHC into the portal vein or

Table 1. Literature review of cases of liver hydatid cysts with portal vein involvement.

First author	Year	Country	Age (years)	Sex	Location	Treatment for the hydatid cyst	Treatment for portal hypertension	Follow-up	Recurrence
Gil-Egea (Gil-Egea et al., 1998)[14]	1998	Spain	84	Female	Hepatic Hilum	Albendazole	None	–	–
Akyürek (Akyürek et al., 2000)[12]	2000	Turkey	60	Male	Segment V	Surgery	None	1 month PO Asymptomatic	No recurrence
Kayacetin (Kayacetin et al., 2004)[16]	2004	Turkey	63	Male	Hepatic hilum	Surgery	None	–	–
Moisan (Moisan et al., 2012)[15]	2012	Chile	62	Female	Hepatic hilum	Albendazole	Propranolol	–	–
Haoues (Haoues et al., 2014)[3]	2014	Tunisia	49	Female	Segment VII	Albendazole	None	12 months PO Asymptomatic	No recurrence
Herek (Herek et al., 2015)[6]	2015	Turkey	52	Male	Hepatic hilum	None	None	–	–
Kirmizi (Kirmizi et al., 2016)[1]	2016	Turkey	33	Male	Hepatic hilum	None	Surgical; H-type mesocaval shunt	–	–
Ertan (Ertan et al., 2019)[9]	2019	Turkey	77	Male	Right posterior lobe	Albendazole	None	–	–
Berkane (Berkane et al., 2020)[11]	2020	Algeria	46	Male	Segment VII	Surgery + Albendazole	None	6 months PO Asymptomatic	No recurrence
Our case	2021	Tunisia	70	Female	Segments V, VI, and VII	Surgery	None	2 years PO Asymptomatic	No recurrence

PO, postoperatively.

its branches is an uncommon complication. To the best of our knowledge, there have been only 10 reported cases, and in all cases, the cysts were located near the hepatic hilum.[1,2] Vascular involvement is due to a mass effect on the adjacent vessels rather than vascular invasion leading to intravascular rupture.[6] The low incidence of intravascular rupture is owing to the elasticity of the vascular wall in contrast to the rigidity of the biliary ducts.[7,8] Venous compression is the result of two factors: mechanical and inflammatory, and its occurrence is determined by the cyst size and the perivascular topography.[3] Moreover, vascular compression may result in the development of a cystovascular fistula with passage of the hydatid contents into the blood vessels. The latter is considered one of the most serious complications of LHC.[3,9] This complication can lead to several problems, such as portal hypertension and anaphylactic shock due to the leakage of antigenic material from the cyst.[10,11] Therefore, it is important to make a rapid diagnosis of cystovascular fistulas and treat this condition as a surgical emergency.

The clinical presentation of HD is frequently nonspecific, and some cases may be completely asymptomatic.[10,12] Patients with portal hypertension and cavernomatosis often have right hypochondriac or epigastric pain, hepatomegaly, splenomegaly, and dilated, tortuous abdominal veins due to collateral venous circulation.[12] In all of these circumstances, clinical signs are not indicative of the rupture of the LHC into the portal venous system, and the diagnosis is based on radiological findings. Abdominal ultrasonography (US) and CT are the first-line and most readily available imaging methods for establishing the diagnosis of LHC as well as pinpointing the location of the cysts and their relationships with the surrounding structures.[7,13] In the case of rupture of the LHC into the portal vein, Doppler color US may show the thrombosis as echogenic content within the vessel with indirect signs of portal hypertension, such as splenomegaly, dysmorphic liver, varices, reversed portal venous flow, and multiple serpiginous vessels of variable size around the obstructed portal vein.[12] Contrast-enhanced CT is superior to US for the diagnosis of LHC. Contrast-enhanced CT is the imaging modality of choice for assessing complications and the relationships of the cysts with the neighboring structures.[14] In addition to the classic indirect signs of portal hypertension, contrast-enhanced CT may reveal communication between the cyst and the portal vein and the daughter vesicles, appearing as hypodense images without enhancement after injection of iodine contrast within a dilatated portal vessel.[12]

Treatment for HD comprises both surgical and medical modalities, requiring treatment for both the hydatid cyst and portal hypertension. However, in some cases, surgical treatment may be difficult owing to the higher risk of hemorrhage in patients with portal hypertension.[1] In previously reported cases, the hydatid cysts were treated surgically only in three cases.[1,11,12] One case was treated with endoscopic retrograde cholangiography, and five patients received albendazole.[3,10,12,15,16] Portal hypertension is treated with beta-blockers to reduce portal pressure, endoscopic ligation of esophageal varices, and percutaneous or surgical shunt surgery in some cases.[1,13] In previous cases, one patient was treated with propranolol[16] and another was treated surgically.[1] Radical hepatectomy helps prevent hydatid cyst recurrence and its complications, including portal vein thrombosis.[17,18]

Conclusion

Rupture of an LHC into the portal venous system is rare. Abdominal US and CT are the first-line and most readily available imaging methods for establishing the

diagnosis of LHC as well as pinpointing the location of the cysts and the relationships with the surrounding structures. The definitive treatment for LHC with portal vein involvement is radical surgery, including hepatic resection.

Ethics statement

This study was approved by the Sahloul Hospital Ethics Committee (approval no. U2349). All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Verbal informed consent (not recorded) was provided by the patient for the publication of her medical information and images.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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Author contributions

Editing and supervision of the manuscript writing: MBL. Editing of the manuscript, literature review, and drafting the manuscript: HA and RG. Editing of the manuscript and data collection: MAS, MA, OB, ND, and MBM. Manuscript revision, supervision of the manuscript writing, and approval of the version to be published: HBH. Supervision of the manuscript and approval of the version to be published: ABA.

ORCID iD

Nouha Dammak https://orcid.org/0000-0002-8447-4726

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