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Case Report

Giant Hydatid Cyst of the Liver Revealed by Portal Hypertension

*Abdelkrim Hablal[1], Mohand Amoura[1], Hakim Chater[1], Meriem Mokhbi[1], Lydie Bounoua[1], Selma Yahyaoui[1], Manel Amri[2], Rania-Sihem Boussa[2], Imene Soufli[2], Chafia Touil-Boukoffa[2] and Nassim Sid-Idris[1,3]**

[1]Department of Surgery, Djilali Belkhenchir hospital, Algeria

[2]Department of Biological Sciences, Laboratory of Cellular and Molecular Biology, University of Sciences and Technology Houari Boumediene, Algeria

[3]Department of Medicine, Benyoucef Benkhedda University, Algeria

Abstract

Echinococcosis is an endemic zoonosis in Algeria. It induces the formation of cysts in several organs with a predilection for the liver and the lungs. Hydatid cysts can grow to an extremely large size. In this case, they are often associated with additional symptoms due to organ compression. Although only a few cases have been reported in the literature, the vascular complications of a large liver cyst should be considered. The choice of treatment must be well studied to avoid any complications like cirrhosis. We describe here an unusual case of a 54-year-old patient with giant hydatid cyst of the liver associated with Portal Hypertension (PHT). Diagnosis was made by imaging and confirmed by serology and hydatid fluid examination. Treatment was performed by surgery.

Keywords: Echinococcosis; Giant liver cyst; Portal hypertension; Imagery; Surgery; Complications

Abbreviations

PHT: Portal Hypertension; LMWH: Low Molecular Weight Heparins

Introduction

Hydatid echinococcosis is a multi-visceral disease whose hepatic location is the most frequent (75% of all locations). It is an endemic cosmopolitan pathology in Africa, Asia, Latin America, and Eastern Europe [1]. In Algeria, it is most prevalent in rural areas and the highlands. According to INSP (national institute of public health) data, the incidence has decreased in recent years (from 2.2% in 2000 to 0.9% in 2017 per 100,000 inhabitants) [2].

Asymptomatic for a long time, hepatic echinococcosis is most often revealed fortuitously during an abdominal ultrasound. However abdominal Computed Tomography (CT) scan remains the key examination for the exhaustive assessment of the lesions in pre-therapy. Hydatid serology which has a dual diagnostic and monitoring interest; confirms the diagnosis in 95% of hepatic locations [3].

The hydatid cyst of hepatic location can be complicated by cracking towards neighboring organs or become infected. It can become voluminous and be responsible for compression, exceptionally causing Portal Hypertension (PHT) [4]. Thus aggravating the prognosis of an initially benign pathology, its management is multidisciplinary. Despite the contribution of medical treatment, surgery remains the only curative treatment [5].

We report in this article, the case of a large hydatid cyst of the liver revealed by PHT. We first describe patient's history, diagnostic methods, treatment modalities, and monitoring.

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***Corresponding author:** Nassim Sid-Idriss, Department of Surgery Djilali Belkhenchir hospital and Faculty of Medicine Benyoucef Benkhedda University, Algiers, Algeria, E-mail: nassim.sididris955@gmail.com

Case Presentation

A 54-year-old patient who consulted for pain in the right hypochondrium and chronic anemia. Clinical examination revealed the presence of a large mass going from the right hypochondrium to the right iliac fossa. This mass is firm, painful, 20 cm long axis, and associated with abundant ascites and collateral venous circulation.

Diagnostic methods

Biological examination revealed microcytic anemia (Hemoglobin at 9 g/dL), thrombocytopenia (57,000/mm^3), prothrombin level at 43%, and albumin at 48.8 g/L, associated with cytolysis syndrome without cholestasis.

Abdominal ultrasound (Figure 1) showed a large retro-peritoneal and retro-renal solid-cystic tumor mass with hepatic infiltration complicated by PHT in ascites decompensation.

The echography was supplemented by an abdominal/pelvis CT scan (Figure 2) showing a dysmorphic liver with atrophy of the right side and hypertrophy of the left side. It's the seat of five cystic formations including the largest multilocular cyst of 280/166/146 mm. The latest occupies the entire right liver causing a dilation of the intrahepatic bile ducts. The other cysts sitting at the level of Segment IV (55/45/38 mm and 44/29/19 mm), Segment III (67/59/36 mm), and Segment VII (80/54/35 mm). The portal trunk is increased in caliber to 19 mm, permeable, with splenomegaly and a very abundant fluid effusion occupying all the peritoneal recesses. There is also a multilithiasic gallbladder.

The hydatid origin of the cyst was confirmed by serology using ELISA and indirect hemagglutination test. In order to assess this PHT, a hepatic Doppler ultrasound (Figure 3) was performed. This test

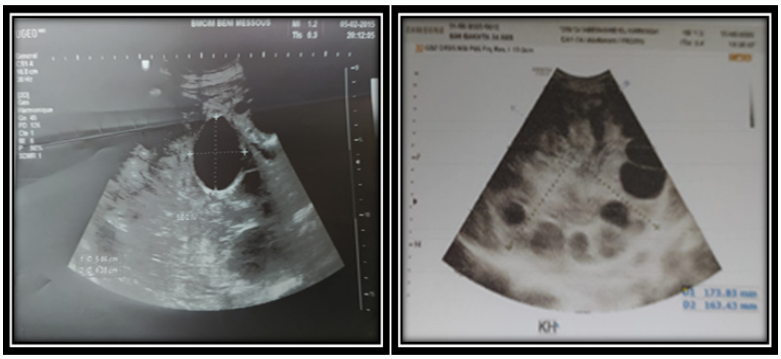

Figure 1: Abdominal ultrasound showing giant hepatic cyst.

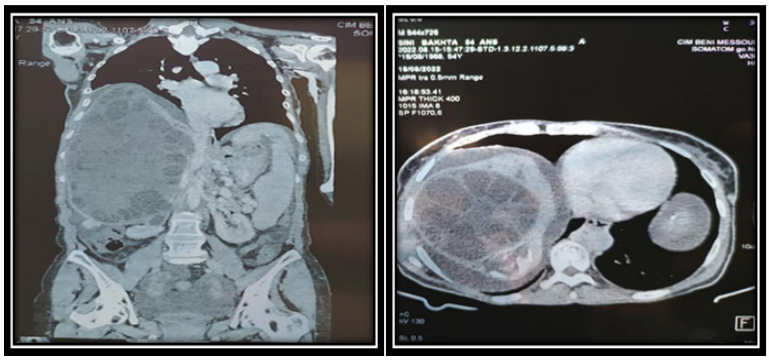

Figure 2: Abdomen and pelvis CT scan revealing a giant solid-cystic mass in the liver (arrowhead), characterized by several daughter-cysts (asterisks), and associated with four other cysts. Axial section (left), coronal section (right).

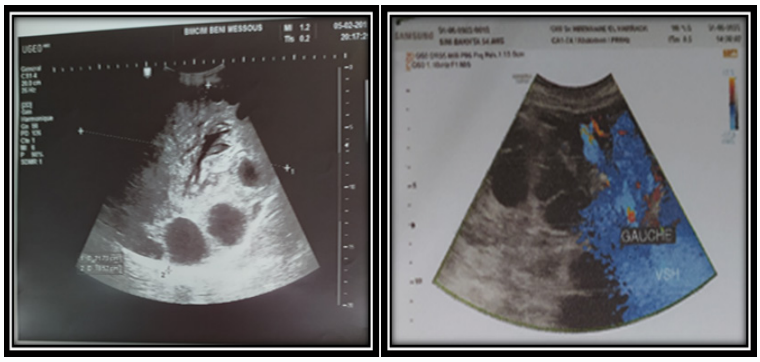

Figure 3: Hepatic Doppler ultrasound showing venous dilatation.

revealed a thrombosis of the right hepatic vein and a dysmorphic liver at the stage of cirrhosis. Moreover, an Esophagogastroduodenoscopy (EGD) showed grade 1 and 2 esophageal varices.

Treatment modalities

Based on characteristic images, in addition to a positive serology, diagnosis of PHT secondary to a bulky compressive hydatid cyst of the liver causing cirrhosis (Child-Pugh A5) was laid. Four other smaller cysts were also found. Following this, the therapeutic protocol which includes both medical and surgical acts was established.

The preoperative preparation includes the correction of various disorders caused mainly by coagulation. In this sense, patient benefited from a transfusion of red blood cells, platelets, fresh frozen plasma, and vitamin K. Moreover, she received an anticoagulant type Low Molecular Weight Heparins (LMWH) at a hypo-coagulant dose with discontinuation the day before surgery. After a last preoperative evaluation, the patient was admitted to the operating room.

Intraoperative exploration found abundant ascites and a large cyst occupying the entire right liver which appeared dysmorphic and nodular with signs of PHT, with venous dilation type and a second 5 cm cyst straddling segment III and IV with hypertrophic left liver.

After protection of the operating site by drapes soaked in scolicidal agent, we proceeded to an aspiration of the cyst after cystotomy which brought back a thick content made up of innumerable daughter vesicles (more than 500 vesicles of variable diameters) and germinal membrane (Figure 4); followed by resection of the protruding dome. The second cyst of the left liver was treated using the same process, followed by hemostasis and bilistasis. The procedure was completed with a cholecystectomy with drainage of the residual cavities and the

abdominal cavity.

During the operation, the patient presented hemodynamic instability requiring a transfusion of two packed red blood cells and three fresh frozen plasmas.

The aspiration of the fluid from intact cysts revealed either clear fluid showing some live protoscoleces characteristic of Echinococcus granulosus, or trouble fluid containing free hooks, as shown in (Figure 5). These observations confirm the initial diagnosis of hydatid cyst.

Monitoring

The postoperative days was marked by the appearance of medical jaundice on the 2nd day followed by an external biliary fistula directed at a high flow evolving favorably after an endoscopic sphincterotomy performed on the 7th day. The patient was declared discharged on the 21st postoperative day with a clean wound and prescription for Albendazole.

The follow-up at 3 months found a patient in a good general condition and anicteric. The clinical examination showed the persistence of PHT sequels (ascites and collateral venous circulation). She was entrusted to hepatologists for the management of her cirrhosis.

Discussion

Liver hydatid disease is initially asymptomatic, especially when the cysts are small, well-encapsulated or calcified [6,7]. However, complications are common in advanced disease. Cyst rupture, infections or compression from the growing cyst are the most common complications [3]. Actually, the disease is complicated by compression in 15% of cases, of which less than 1% are the cause of a vascular complication. The vascular complications are secondary to calcifications of the vessels causing ischemia involving the portal trunk and its branches, the inferior vena cava and the supra-hepatic veins [5].

Giant hydatid cysts as well as portal vein involvement are therefore not common with only few published cases [4,8,9]. A cohort analysis study reported that cystic echinococcosis of the liver is an infrequent cause of Portal Hypertension (PHT). However, larger cyst size and male sex are the main risk factors of this vascular complication [10]. In this context, we report here the case of a female patient with a large abdominal mass with signs of PHT.

Diagnosis of hydatid cyst is based on imaging. The ultrasound and Doppler association specify its characteristics: the seat, the number, the cyst contents, the vascular cartography (relationships with the vessels: supra-hepatic veins, portal vein, inferior vena cava, splenic vein), any flow abnormalities, splenomegaly, ascites, and the biliary mapping [11,3]. In fact, Doppler ultrasound is the most important technique used in the assessment of PHT allowing measurement of the pressure at the level of the portal trunk and showing the contact of the cyst with the different vascular axes. This method looks also for the presence of ascites, varices, or both [12]. Finally, Doppler ultrasound allows classifying the cyst according to the Gharbi classification [13]. In our case, Doppler ultrasound enabled us to classify the hydatid cyst

Figure 4: Intraoperative photograph showing the entire cysts and daughter cysts removed, which developed inside the primary giant cyst (left). Some are ruptured (left) and other intact and filled with hydatid fluid (right).

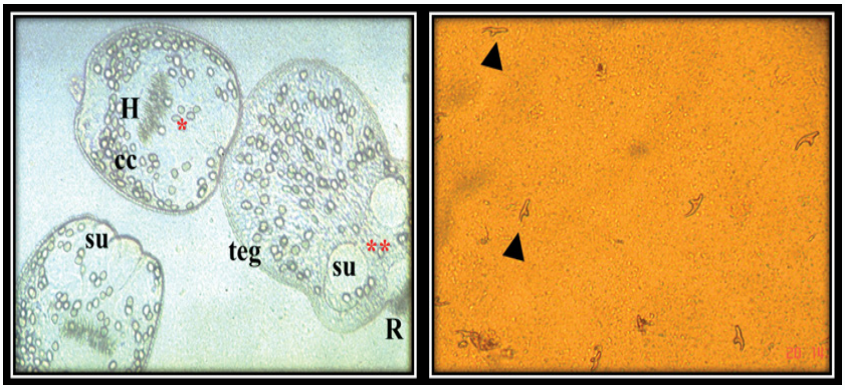

Figure 5: Aspirated hydatid fluid observation under light microscopy (Gx269). Live invaginated (*) and evaginated (**) protoscoleces (left). Hooks (arrowhead) (right).
cc: Calcareous Corpuscles; H: Hooks; R: Rostellum; su: Suckers. Teg: Tegument

as CE2 (multilocular with daughter cysts).

The scanner is not the first-line examination. However, it's indicated in front of a bulky cyst and a type IV cyst (according to the Gharbi classification) or if there is doubt about the diagnosis. MRI on the other hand does not bring more information but is the best tool to demonstrate a biliary tree involvement [14,3].

The biological tests do not allow the positive diagnosis of the vascular complications of liver hydatid cyst. However, if there is PHT, there is a peripheral cytopenia mainly due to hypersplenism and cytolysis in hepatic assessment [12].

Hydatid serology is an essential step in the diagnosis. Indeed, it can guide the diagnosis in 80% to 95% of cases, provided however that a qualitative and a quantitative technique are used [15]. However, a major problem arises is the existence of false positive and false negative results. The first are due to cross reactions with other parasites or presence of cancer or chronic immune disorders. The second are due to cyst calcification. Have found that serology is positively correlated with cyst size but within fertile cysts only. That mean that serology is useful in the case of multiple or large fertile cyst because of the quantities of parasitic antigens exposed to host immune response [16].

Direct diagnosis by searching for the pathogen is therefore impossible. Indeed, it is strictly contraindicated to perform a puncture of the cyst, because of the risks of dissemination and then the risk of a secondary echinococcosis. Finally, the definitive diagnosis of echinococcosis is based on the microscopic and anatomopathologic study of the hydatid fluid from an operating specimen [17] as performed here.

The place of surgery in the treatment of the hydatid cyst is reduced in the case of an uncomplicated cyst, but it remains essential in the case of complications (such as a giant cyst which contains many daughter vesicles) preventing percutaneous treatment as in our case here [18,3].

Surgical treatment depends on the parasite load at the time of diagnosis, the early detection of complications and the medical treatment undertaken [3]. Several studies have demonstrated the superiority of radical treatment compared to conservative treatment (resection of the protruding dome, partial pericystectomy, PAIR: Puncture, Aspiration, Injection, Reaspiration). Indeed, the radical treatment has the advantage of eliminating the residual cavity and therefore avoiding septic complications, mainly external biliary fistulas. Nevertheless, the conservative treatment, which can be carried out quickly, easily and without significant blood loss, is associated with low mortality. It finds its place in certain emergency cases [19,20]. Moreover, surgical management must be early before the appearance of irreversible sequels, in particular biliary fistulas and cirrhosis.

The management of preoperative and postoperative external biliary fistulas is multimodal. It's essentially based on endoscopic sphincterotomy, which is the treatment of choice. Indeed, this therapeutic approach avoids the consequences of excessive loss of bile and shortens the postoperative stay. However, it is not free of risk with an overall morbidity rate between 4 and 10% represented in order of frequency by acute pancreatitis (5.4%), haemorrhage (2%), biliary infection (1.5%) and perforation (0.3%) and a mortality rate of 0.5% [21-23].

Cirrhosis increases the poor prognosis and can be responsible for complications linked to chronic liver disease, namely cardiovascular and respiratory insufficiency and especially hemostasis disorders. Portal hypertension and its irreversible complications also constitute an important prognosis factor in the management of hepatic echinococcosis. The latter being increased by hepatic destruction at the origin of cirrhosis requiring at the terminal stage the use of liver transplantation [24]. In our patient, sequels of PHT and cirrhosis persisted after surgery due to late hospital admission and treatment.

Conclusion

Hepatic echinococcosis is a relatively frequent benign pathology. It is revealed exceptionally by portal hypertension increasing the vital prognosis immediately and remotely. Its diagnosis is based on ultrasound coupled with Doppler most often at the stage of complications.

Its management is difficult and required preparation before surgery. Conservative treatment is an emergency life-saving gesture but may be responsible for postoperative septic complications. Cirrhosis remains a possible and frequent complication based on the vital prognosis and sometimes requiring the use of transplantation.

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