Case report

Hydatid cyst of the liver fistulized into the inferior vena cava

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ABSTRACT

Introduction: Fistulization or rupture of hydatid liver cysts to the inferior vena cava (IVC) is an extremely rare and life-threatening condition.

Presentation of case: We report the case of a 70-year-old patient who presented with right-upper-quadrant pain and fullness evolving for 03 months. Physical examination showed dilated veins over the anterior abdominal wall and the flanks associated with lower-extremity swelling. Computed tomodograph of the abdomen showed a hydatid cyst invading segments VI and VII of the liver fistulized into the inferior vena cava. The IVC was partially thrombosed. The diagnosis of a possibly ruptured hydatid cyst in the inferior vena cava was then made. The patient underwent surgical management. Per-operatively the cystic cavity had bloody content but the cysto-vascular communication was not identified. Partial cystectomy was performed leaving a fairly extensive contact between the calcified pericyst and the IVC. The postoperative course was uneventful.

Discussion: Rupture of the hepatic hydatid cyst into the IVC is very rare and may lead to fatal pulmonary embolism secondary to the migration of vesicles in the pulmonary artery or haemorrhagic shock. CT scan remains the best investigation method to assess the vascular links of the hepatic hydatid cyst especially with the IVC. Surgical treatment of the hepatic hydatid cyst ruptured into the IVC mandates vascular control before the hydatid cyst is punctured or removed.

Conclusion: Fistulized hydatid cysts into the IVC should be operated on in centres equipped for extracorporeal bypass techniques, and experienced in the surgery of hepatic echinococcosis.

1. Introduction

Cystic echinococcosis is a parasitic disease highly endemic in the Mediterranean countries [1].

Echinococcal cysts involve predominately the liver (60-70%) and the lungs (15%). It arises in other abdominal organs in 10% of cases [2].

Compression of the inferior vena cava by hydatid cysts is commonly seen, however, fistulization or rupture of hydatid liver cysts to the inferior vena cava (IVC) is an extremely rare and life-threatening condition [3]. It may cause the development of hydatid cysts either in pulmonary parenchyma or rarely pulmonary arteries [4].

It can manifest with an acute fatal or subacute or chronic clinical presentation. Massive pulmonary embolization is the most dreadful event and should indicate an emergency treatment [5].

We report herein a case of a 70-year-old man with a spontaneous fistulization of a hepatic hydatid cyst in the inferior vena cava.

This work has been reported in line with the SCARE 2020 criteria [6].

2. Case presentation

A 70-year-old male patient, without past medical history, presented with right-upper-quadrant pain and fullness evolving for 03 months. On physical examination, the vitals of the patient were stable. Inspection showed dilated veins over the anterior abdominal wall and the flanks associated with lower-extremity swelling. The laboratory findings were normal. Serological tests detected the presence of anti-hydatid cyst antibodies. Abdominal sonography showed a 7 cm type III hydatid cyst (as CE 2 cyst following the WHO classification) involving the hepatic fundus. Computed tomography reported that the cyst was 9 cm × 7 cm in diameter invading segments VI and VII of the liver and fistulized in the retrohepatic inferior vena cava. The infrahepatic segment of the IVC was not totally filled with contrast agent (Fig. 1). This flow defect suggested direct extension into the lumen of the vessel. There was no hydatid cyst in the lung parenchyma. The diagnosis of a possibly ruptured...
Hydatid cyst in the inferior vena cava was then made. Curative anticoagulation was initiated. Accounting for the risk of dissemination of daughter cysts and pulmonary embolization, surgery was considered. Peroperatively the cyst was opened with caution, the cyst cavity had bloody content. Meticulous examination of the inner surface of the cyst does not find any macroscopic cysto-vascular communication. There was no need to clamp the IVC in its upper and lower parts. Partial cystectomy was performed leaving a fairly extensive contact between the calcified pericyst and the IVC (Fig. 2). The procedure was concluded with an omentopexy into the remaining cavity. The histological examination of the resected specimen confirmed the presence of a laminated acellular hydatid membrane (Fig. 3). The postoperative course was uneventful and the patient was discharged on day five. During follow-up, the patient was treated with albendazole (10 mg/kg/d in two divided oral doses) for three months. A radiological check-up is envisaged at 6 months postoperatively.

3. Discussion

Hydatid disease remains a public health problem in highly endemic countries such as Tunisia [1].

Infectious, biliary and thoracic complications are the standard complications and are present in 40% of cases [7]. Spontaneous rupture of the hepatic hydatid cyst into the IVC is very rare, with only a few cases reported in the literature [3].

The rupture is typically small and not visible on contrast-enhanced CT. If the rupture site enlarges, a fatal pulmonary embolism can result secondary to the migration of vesicles or “daughter” cysts in the venous to the right atrium as well as the right ventricle and finally to the pulmonary artery [8].

Clinically hydatid cyst pulmonary embolism has three possible reported outcomes: (1) sudden death due to acute embolism, (2) subacute pulmonary hypertension, and (3) chronic pulmonary hypertensive cases [9].

In addition to the pulmonary embolism, theoretically spontaneous rupture of the hepatic hydatid cyst into the IVC can trigger a haemorrhagic shock. However, this condition has never been described in the literature and can probably be explained by the fact that these patients die before they reach the hospital [10].

In our case, CT and per-operative exploration did not visualize the exact cysto-vascular communication. This fact can be attributed to the higher intracystic pressure and the valvular functions of the pericystic folds which usually prevent blood from the vena cava from filling the cyst [11].

Radiologically, the CT scan remains the best investigation method to assess the vascular links of the hepatic hydatid cyst especially with the IVC [12].

The rupture in the IVC concerns generally hydatid cysts located in the posterior segments of the liver (VII, VIII and I) and in contact with the IVC. The latter may be compressed or laminated by the cyst, and sometimes it can be partially or totally thrombosed due to the presence of vesicles in its lumen.

Surgical treatment of the hepatic hydatid cyst ruptured into the IVC is mandatory to prevent a fatal pulmonary embolism and hemorrhagic shock [11]. Per-operatively the hydatid cyst must be handled with care, the cystic-IVC communication must be checked and then shut.

According to some reports total vascular occlusion with direct access to the subphrenic or even intrapericardial vena cava is compulsory, before the hydatid cyst is punctured or removed. The cavo-caval bypass should be available, if necessary [8].

When lesions of the liver significantly violate the retrohepatic IVC

Fig. 1. (A) Abdominal contrast-enhanced CT scan revealing the communication between the hydatid liver cyst and the IVC (Red arrow). (B) Visualisation of a 19 mm thrombus in the inferior vena cava (white arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Fig. 2. Intraoperative photograph demonstrating the adhesion between the calcified pericyst and the anterior surface of the inferior vena cava.
Pulmonary embolism due to a ruptured hepatic hydatid cyst: clinical and radiologic imaging findings, Emerg. Radial. 18 (2011) 437–439.

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