Budd-Chiari syndrome secondary to hepatic echinococcosis

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SUMMARY

Objectives — Hydatid cyst of liver is a parasitosis which is an endemic state in Tunisia and is a very rare cause of Budd-Chiari syndrome. The purpose of this study was to report the clinical features, radiological investigations and therapeutic management.

Patients and methods — A retrospective analysis of 12 patients who underwent surgery for Budd-Chiari syndrome secondary to hepatic echinococcosis between January 1990 and December 2004 was performed.

Results — The series included ten females and two males with a mean age of 36 years. Budd-Chiari syndrome was subacute in 75% of cases. Ultrasound showed a compression of hepatic veins by cysts with a mean diameter of 13 cm situated in at least two hepatic segments. US Doppler and CT-scan of the liver provided the diagnosis in all cases. Laparotomy was performed in all cases. Operative mortality was 8% and morbidity 66% due to biliary fistula and deep abscess formation. Hepatic vein outflow was successfully re-established in four patients.

Conclusion — Budd-Chiari syndrome is a rare but severe complication of hydatid cyst of the liver. Early diagnosis is necessary to improve prognosis.

SYNDROME DE BUDD-CHIARI

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Objectifs — Le syndrome de Budd-Chiari est une complication rare du kyste hydatique du foie. Le but de cette étude était de décrire les particularités cliniques et radiologiques ainsi que la prise en charge thérapeutique de cette entité.

Malades et méthodes — Rétrospectivement, 12 cas de syndrome de Budd-Chiari secondaire à une compression hydatique ont été colligés sur une période allant de janvier 1990 à décembre 2004.

Résultats — Dix femmes et 2 hommes, d’âge moyen de 36 ans, ont été étudiés. Le syndrome de Budd-Chiari était cliniquement subaigu dans 75 % des cas. À l’échographie abdominale, les kystes hydatiques du foie comprimant le carrefour cavo-sus-hépatique occupaient au moins deux segments du dôme hépatique et avaient une taille moyenne de 13 cm. L’échodoppler et l’angioscanner ont permis de poser le diagnostic de kyste hydatique du foie compliqué d’un syndrome de Budd-Chiari. Tous les malades ont été opérés par voie conventionnelle. Vis-à-vis du kyste, le geste a été conservateur dans tous les cas. La mortalité opératoire a été de 8 % et la morbidité de 66 %, principalement par fistule biliaire et suppuration de la cavité résiduelle. Le traitement du kyste a permis une reperméabilisation des veines sus-hépatiques dans 4 cas.

Conclusion — Le syndrome de Budd-Chiari est une complication rare mais grave du kyste hydatique du foie pouvant mettre en jeu le pronostic vital. Le pronostic peut être amélioré par un diagnostic précoce.

Introduction

Hydatid disease of the liver is a particularly frequent in Tunisia where echinococcosis is endemic. Several complications have been described. Vascular complications are rare, generally involving vessel rupture or Budd-Chiari syndrome (BCS).

The purpose of this retrospective analysis of twelve cases of BCS secondary to hepatic echinococcosis was to describe the particular aspects of this specific complication. Ten women and two men, mean age 36 years, were included in the analysis. Nine presented subacute BCS. The abdominal ultrasound (US) revealed venous outflow obstruction caused by cystic formations occupying at least two segments of the hepatic dome and measuring 13 cm on average. US Doppler and computed tomographic (CT) angiography led to the diagnosis of hydatid disease of the liver with secondary BCS. A standard surgical procedure was performed with conservative management of the cyst(s) in all cases. There was one operative death and nine patients developed complications, mainly biliary fistulae and deep abscess formation. Surgical treatment successfully restored venous outflow in four patients.
operative findings were recorded as well as follow-up data. The Gharbi classification of hydatid liver cysts was noted [1] and a standard battery of laboratory tests was performed to assess liver function. After a period of medical management of variable duration, all patients underwent surgical treatment. Clinical, radiological, operative and post-operative findings were recorded as well as follow-up data.

Results

This retrospective series included twelve cases of BCS secondary to hydatid liver disease in ten women and two men, mean age 36 years. Mean time from symptom onset to consultation was 30 days. All patients resided in a rural area. The clinical manifestations were acute in three patients (25%) and subacute in nine (75%). The predominant symptom was pain in the right hypochondrium (10 patients). Other symptoms were jaundice (2 patients) and swelling of the lower limbs (2 patients) (table I). The physical examination revealed hepatomegaly in ten patients, collateral venous circulation in seven, ascites in five, splenomegaly in three, and cutaneous cutaneous jaundice in two (table I). Laboratory tests disclosed inflammatory anemia in eight patients, cholestasis in six, and elevated serum transaminase levels in all twelve patients. The transaminase level was moderately elevated in the nine patients with subacute disease (on average 3 x upper limit of normal), and very elevated in the three with acute disease (9, 11 and 14 times the upper limit of normal). The three patients with acute BCS also presented coagulation disorders with a decreased serum level of factor V. The mean prothrombin time was 30% of the normal control (range: 72-100%). Echinococcus serology was positive in all patients.

A first-intention abdominal US was performed in all patients and consistently demonstrated the presence of a hydatid cyst in the hepatic dome, in contact with the suprahepatic venous confluence. Other hepatic cysts were also identified in nine patients. Mean cyst diameter was 13 cm (range 7-19 cm). The cyst occupied at least two segments of the hepatic dome in all patients. According to the Gharbi classification the cysts were: type I (univesicular) in five patients, type III (multi-vesicular) in five and type IV (pseudo-tumoral) in two. Hypertrophy of segment I was noted in two patients. Ascites was minimally abundant in seven patients, moderately abundant in three and very abundant in two.

US Doppler was performed in seven patients and disclosed extrinsic compression of at least two hepatic veins in all seven, with presence of an associated thrombus in five. Portal hypertension was noted in all seven patients with a US Doppler examination. A CT angiogram was available for the five other patients and demonstrated compression of the hepatic veins in all five, and a thrombus in the inferior vena cava in one (figures 1 and 2). Hypertrophy of segment I was noted in two patients. Upper gastrointestinal endoscopy was performed in all patients and demonstrated grade I esophageal varices in one. There were no cases with pulmonary localization of the hydatid disease and no thoracic complications were noted.

Preoperative care was necessary for anti-coagulation (heparin 3-5 mg/kg/d) (12 patients), large-volume ascites paracentesis (2 patients), and correction of electrolyte imbalance (1 patient) and anemia (1 patient). To control hemostasis, patients were given an infusion of fresh-frozen plasma the day before surgery and a provision of at least three units of packed red cells was available at surgery in all patients.

A standard surgical procedure was used in all patients. None of the patients were given albendazole before surgery due to the risk of hepatic toxicity. A right subcostal approach was used in seven patients and a bilateral subcostal approach in five. The liver appeared swollen and congestive in all patients. Hypertrophy of segment I was noted in four patients. Ascites was a constant finding. Three patients had unique cysts and nine had multiple cysts. The cysts were located in the hepatic dome in all patients with a univesicular aspect in five and a multivesicular aspect in five. In two patients, the cysts were described as bilipurulent. A small biliary fistula was noted in five patients.

The surgical procedure consisted in resection of the protruding hepatic tissue in seven patients and puncture aspiration of the cyst with sterilization and removal of the proliferous membrane or daughter vesicles in five. This simple minimalistic procedure was dictated in these five patients because of the risk of bleeding and the associated portal hypertension. Cholecystectomy was associated in all patients with intraoperative cholangiography, which was normal in seven patients and revealed extravasation of the contrast agent into the residual cavity in the others. This was due to the presence of a biliary fistula which had to be closed with an X suture. The residual cavity was drained in all patients using two n° 18 drains. A liver biopsy was obtained in eight patients and provided histological proof of BCS in all of them (fibrosis of the portal spaces without regeneration nodules). One liver biopsy demonstrated cirrhosis.

The postoperative period was complicated in eight patients (66%). Abscess formation in the residual cavity required drainage in five patients. This complication aggravated the BCS in three patients and led to the formation of a thrombus in the inferior vena cava in one patient who was successfully treated with curative-dose anticoagulants. A biliary fistula to the skin was noted in three patients and resolved successfully with aspiration drainage. One 60-year-old diabetic patient died from septic shock on the fourth postoperative day. This patient had acute BCS and presented pneumonia on admission.

Mean follow-up was six months. During the postoperative follow-up, ascites regressed in 86% of patients, within ten days on average. Hepatomegaly resolved in 43%, within 45 days on average. Venous outflow was successfully restored with anticoagulant treatment in four patients, 72 days after surgery on average. There were two cases of venous thrombosis, with successful recovery of a patent inferior vena cava 90 and 110 days postoperatively. At the third postoperative week, liver tests had

### Table I. – Clinical signs at diagnosis.  
*Signes cliniques lors du diagnostic.*

<table>
<thead>
<tr>
<th>Signs</th>
<th>N (%)</th>
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<tbody>
<tr>
<td>Right hypochondrial pain</td>
<td>10 (83)</td>
</tr>
<tr>
<td>Hepatomegaly</td>
<td>10 (83)</td>
</tr>
<tr>
<td>Collateral venous circulation</td>
<td>7 (58)</td>
</tr>
<tr>
<td>Ascites</td>
<td>5 (42)</td>
</tr>
<tr>
<td>Splenomegaly</td>
<td>3 (25)</td>
</tr>
<tr>
<td>Edema of the lower limbs</td>
<td>2 (17)</td>
</tr>
<tr>
<td>Jaundice</td>
<td>2 (17)</td>
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</tbody>
</table>
Budd-Chiari syndrome secondary to hepatic echinococcosis

returned to normal in ten patients and remained perturbed in the patient with cirrhosis. Anticoagulant treatment was continued for six months in patients with venous thrombosis.

Discussion

Obstruction of hepatic venous outflow is the characteristic feature of BCS. The obstruction has to involve at least two of the main suprahepatic veins to produce BCS [2]. Many causes have been described, predominantly myeloproliferative syndromes, coagulation factor deficiencies, and antiphospholipid syndrome. Hydatid causes are exceptional [3, 4]. In our series, none of the patients had a personal or family history of venous thrombosis and hematology tests were normal (excepting one patient with inflammatory anemia).

BCS secondary to hepatic echinococcosis can occur if voluminous hydatid cysts in the hepatic compress the hepatic veins [5, 6]. To produce BCS, cysts have to be very large and lie in an appropriate position [5]. Average cyst diameter in our series was 13 cm. Advanced-stage disease and the status of the pericyst are aggravating factors [5]. As it develops, the cyst produces a mass effect, with compression of the suprahepatic veins and/or the inferior vena cava. This mechanical effect is associated with an inflammatory effect resulting from the chemical and bacteriological aggression of the cyst contents producing endovenitis and subsequently venous thrombosis which in turn aggravates the BCS and, in the event of a postoperative deep abscess, favors thrombus formation in the vena cava.

Three pathological stages of BSC are distinguished. In stage I, the hepatic parenchyma presents reversible congestion and hemorrhagic infiltration. The development of centrallobular fibrosis is the hallmark of stage II and in stage III the damage becomes irreversible, with fibrosis of the portal spaces and inevitable progression to cirrhosis [7] was observed in one of our patients. The clinical triad of rapidly developing hepatomegaly, abdominal pain and abundant ascites is the typical presentation of acute BSC, observed in one-quarter of the patients in this series. These manifestations are secondary to acute obstruction of all three hepatic veins [2]. A subacute presentation is much more frequent and was observed in three-quarters of our patients. Occasionally upper gastrointestinal bleeding subsequent to rupture of esophageal varices is the inaugural sign [2]. The presence of esophageal varices is however exceptional in BCS despite the portal hypertension. Among the seven patients in our series with US Doppler proven portal hypertension, only one had esophageal varices at upper gastrointestinal endoscopy. Laboratory tests contribute little to diagnosis, with the exception of echinococcosis serology.

Before the development of US Doppler techniques coupled with abdominal CT, the diagnosis of BCS required invasive vascular explorations. Because of its specificity and sensitivity, abdominal US is now the first-intention exploration for suspected hydatid cysts of the liver. In advanced-stage disease, the caudal lobe is hypertrophic with generalized atrophy of the remainder of the hepatic parenchyma [2, 8]. Pulsed Doppler can establish the diagnosis of BCS by revealing the loss of the triphasic flow pattern or absence of color filling of the hepatic veins, sometimes associated with thrombosis of the inferior vena cava [9]. In our series, all of the cysts affected at least two segments of the hepatic dome. In this situation, US Doppler is required to search for BCS.

Abdominal CT provides a better analysis of hepatic dysmorphism and, with contrast injection, an assessment of blood flow patterns. The cyst can be localized with a description of the vascular relations. Signs suggestive of BCS include: defective hepatic perfusion, hypertrophy of segment I (variable, observed in 40% in our series), and thrombus formation in the inferior vena cava or the hepatic veins [10]. Magnetic resonance imaging is highly useful for analyzing thrombi in the inferior vena cava [11].

Surgery is required for the treatment of BCS secondary to extrinsic compression of the hepatic veins by hydatid cysts. The goal is to remove the obstacle and restore effective hepatic blood outflow, and thus eliminate portal hypertension and its consequences. Preoperative management is necessary: heparin in the event of venous thrombosis, treatment of ascites, and treatment of anemia [2].

Laparotomy via a large subcostal incision on the right or bilaterally is required to treat the cyst and cure the biliary com-
plications. Laparotomic exploration confirms the diagnosis of BCS and details the atrophic or hypertrophic aspect of each lobe as well as the presence or not of ascites and signs of portal hypertension. A conservative procedure is required for the cyst (resection of the protruding dome or puncture-aspiration) because of the significant risk of hemorrhage and damage to the hepatic veins exposed to the inflammatory reaction to the pericyst [5]. Drainage of the residual cavity is crucial to prevent abscess formation. In patients with BCS, epiploplasty has been proposed to prevent deep abscess formation, but can also prevent the cystic cavity from collapsing and thus actually sustain the compression of the hepatic veins [13]. This is why this technique was not used in the patients in this series. Liver biopsy should be systematic to evaluate disease progression. In our series, five patients were treated by puncture aspiration alone. A strictly percutaneous procedure could have been used but was contraindicated because of the ascites. Most of the cysts in this series were type III or IV, contraindicating percutaneous treatment [14].

Mortality is high for surgical treatment of hydatid cysts of the liver in patients with BCS (8.3% in our series). Morbidity (66% in our series) is generally related to deep abscess formation. The thick inflammatory pericyst also complicates suture of biliary fistulae [12]. This complication occurred in 41% of our patients. Retention of purulent collections is a serious complication not only because of the risk of infection but also because of the increased risk of BCS and the formation of thrombi, particularly in the vena cava. When associated with abscess drainage, anticoagulant therapy is usually successful for the treatment of vascular thrombi (one case in our series).

**Conclusion**

BCS is a rare but sometimes life-threatening complication of echinococcosis of the liver. This exceptional condition should be suspected when the hydatid cyst affects two (or more) hepatic segments of the dome. Prognosis can be improved by careful preoperative preparation. Prevention of echinococcosis in endemic zones is the best approach for improved treatment.

**REFERENCES**

7. Maillard JN, Hay JM. Syndrome de Budd-Chiari. EMC 7042 A12. Foie, tome II.