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Surgical Treatment for Splenic Hydatidosis

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Abstract. Splenic involvement is rare in patients with hydatid disease even in endemic countries. The spleen is the third most commonly involved organ after the liver and the lung. In our series splenic echinococcosis represents 5.8% of abdominal hydatid disease. During the last 22 years, 14 patients were operated on for splenic hydatid cysts in our department. In 10 patients the spleen was the only location of hydatid disease; in 2 patients there was concomitant liver hydatid disease; one patient had disseminated intraabdominal disease; and one patient had a coexisting hydatid cyst in the quadriceps femoris muscle. Plain abdominal films, ultrasonography, and computed tomography scans were most useful for establishing the diagnosis. All patients underwent splenectomy alone or combined with management of cysts at other sites, except for two patients who underwent omentoplasty and one patient who underwent external drainage. One patient died during the early postoperative period (mortality rate 7%), and three patients had minor complications. Splenic hydatid disease should be included in the differential diagnosis when a splenic cyst is identified, especially in patients with a history of hydatid disease. Surgery remains the treatment of choice to avoid serious complications.

Hydatid disease of the spleen is a rare clinical entity even in endemic countries. A few authors have conducted specific studies for splenic hydatidosis. The first case of a hydatid cyst of the spleen was reported by Bertelot in 1790 from an autopsy [1]. The first cumulative report on splenic hydatidosis was by Sabadini, who presented a review of 300 cases from the international literature [2]. Most reports on this matter are based on small numbers or even sporadic cases [3]. This brief report concerns 14 patients who underwent surgery for splenic hydatid disease in our center. We analyze the pathogenesis, clinical symptomatology, imaging studies, and surgical therapy of the disease.

Methods

Between January 1972 and May 1995 a total of 14 patients with hydatid disease of the spleen were treated surgically in the Second Department of Propedeutic Surgery of the University of Athens. During this period, a total of 241 patients with abdominal hydatid disease underwent operation. In consequence, splenic hydatid disease represents 5.8% of abdominal hydatid disease treated in our center. There were 10 male and 4 female patients, with ages ranging between 27 and 82 years (mean 58.4 years).

The spleen was the only location of the disease in 10 patients, whereas 2 patients had associated liver hydatid cysts, 1 patient had multiple peritoneal cysts together with hepatic involvement, and 1 patient had a concomitant hydatid cyst of the quadriceps femoris muscle (Table 1). Associated diseases in five patients were choledocholithiasis with a recent episode of acute pancreatitis, asymptomatic cholelithiasis, hairy cell leukemia with excessive splenomegaly and hypersplenism, a recent attack of hepatitis A, and rectal adenocarcinoma.

Most of our patients presented no overt clinical symptomatology. Mild abdominal discomfort or pain in the left upper quadrant was the main complaint in some cases. A palpable mass in the left hypochondriac region was observed in five patients with large spleen hydatid cysts.

Preoperative diagnosis was based on imaging techniques and immunologic tests. Upper gastrointestinal radiographs usually indicated calcification of the cyst, an elevated left hemidiaphragm, and displacement of the stomach to the right. The liver scintiscan was a useful diagnostic tool for the early cases (before 1980) but is no longer employed. With the introduction of ultrasonography (Fig. 1) and computed tomography (CT) (Fig. 2) other diagnostic imaging methods have been abandoned. The Casoni skin test was used for diagnosis in early cases but after 1980 was completely replaced by immunologic tests of higher sensitivity, such as counter immunoelectrophoresis (CIE).

All but three patients with splenic hydatid cysts underwent splenectomy. In six cases a median supraumbilical incision was used and in the other eight cases a left subcostal incision. Concomitant liver hydatid disease or multiple peritoneal cysts were managed surgically at the same time. Omentoplasty was used for two splenic cysts and most of the liver cysts; multiple peritoneal cysts were excised.

Our patients were followed every 12 months with a physical examination, liver function tests, serologic tests, and ultrasonography. Every second year an abdominal CT scan was also performed to rule out recurrent disease.

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Table 1. Patient characteristics.

Pt. no.	Age/sex	Associated diseases and abdominal echinococcosis	Symptomatology	Diagnostic studies	Immunologic studies	Size (cm)	Surgery
1	52/M	Hepatic cyst (right lobe)	Mild abd discomfort, palpable mass	AbdXr, GDXr	Casoni (−)	20 × 15 × 10	Drainage, capitonage (liver cyst), splenectomy
2	45/M	—	Left upper quadrant mass	AbdXr, spl scintigram	C asoni (+)	20 × 12 × 8	Splenectomy, caudal pancreatectomy
3	59/M	—	—	GDXr, spl scintigram	—	10 × 10	Splenectomy
4	64/M	Superficial hepatic cysts (right lobe), lesser sac	Mild abd discomfort	AbdXr, US	Casoni (+)	15 × 9 × 5	Resection, omentoplasty, splenectomy
5	58/M	Hepatic cysts, multiple peritoneal cysts	Abd pain	AbdXr, US, CT	Casoni (+)	10 × 8 × 6	Splenectomy-omentoplasty, excision
6	68/F	—	Left upper quadrant pain, palpable mass	US, CT	Serology (+)	32 × 15 × 8	Splenectomy
7	74/M	Choledocholithiasis, pancreatitis	Mild abd discomfort, dyspepsia	US, CT	Serology (+)	20 × 10 × 8	Splenectomy, choledochoduodenal anastomosis
8	52/F	Cholelithiasis	—	AbdXr, US, CT	Serology (+)	15 × 12 × 10	Splenectomy, cholecystectomy
9	70/F	Splenomegaly, hairy-cell leukemia	Hypersplenism, palpable mass	AbdXr, US, CT	Serology (−)	7 × 6 × 5	Splenectomy
10	65/M	Hepatitis A	Mild abd discomfort	Spl scintigram, US, CT	Serology (+)	22 × 15 × 7.5	Splenectomy
11	60/M	Adenocarcinoma of rectum	Rectal bleeding, asymptomatic cyst	Incidental finding at laparotomy	—	10 × 8 × 4	Low anterior resection, splenectomy
12	82/F	—	Palpable mass, left upper quadrant pain	US, CT	Serology (+)	Approximately 33 × 20 × 8	Partial excision, omentoplasty
13	27/M	—	—	CT	Eosinophilia	6 × 5 × 4	Cyst evacuation, omentoplasty
14	42/M	Hydatid cyst of quadriceps femoris muscle	—	CT	—	—	Cyst evacuation, external drainage

Abd: abdominal; AbdXr: abdominal radiograph; GDXr: gastroduodenal radiograph; Spl: spleen; US: ultrasonography; CT: computed tomography.

Results

In 11 cases the preoperative diagnosis was established using diagnostic imaging studies; it was confirmed at surgery. In the other cases the diagnosis was confirmed at surgery performed for liver hydatid disease in two cases and during laparotomy for a low anterior resection for a rectal adenocarcinoma in one case.

The most common radiographic finding was splenomegaly (in 6 of 10 cases) or a soft tissue mass in the left hypochondrium. The cysts were partly calcified in eight patients and completely calcified in two. A single cyst was observed in all cases. In one case associated liver hydatid disease was revealed by ultrasonography, and in another case intraperitoneal hydatid spread was seen on both ultrasound and CT scans. In one case CT scanning revealed a large hydatid cyst of the left quadriceps femoris muscle.

The spleen was removed in 10 cases; in 4 cases splenectomy was laborious because of multiple adhesions. The size of the resected cysts ranged from 7 to 32 cm (Table 1). In one patient a caudal pancreatectomy was required. Other intraabdominal cysts were treated during the operation in three cases. In one patient with concomitant choledocholithiasis a choledochoduodenal anastomosis was required, whereas in another patient with cholelithiasis cholecystectomy was performed. In one patient a large splenic cyst occupied the entire space from the posterior peritoneum to the anterior abdominal wall, displacing the esophagus, stomach, and colon. Excision of the cyst was not feasible, and partial excision of the cyst with omentoplasty was performed. Cyst excision was not feasible in two other patients as well, and they underwent cyst evacuation with omentoplasty and external drainage, respectively.

In one patient there was diffuse hemorrhage from the spleen during operation. A small hole (0.5 cm diameter) remained in the left hemidiaphragm after removal of the spleen due to infiltration of the diaphragm from the cyst wall, which was closed with interrupted sutures. This patient was reoperated on the first postoperative day for recurrent hemorrhage. Her condition deteriorated, and she died from multiorgan insufficiency 15 days later, for a perioperative mortality rate of 7% in our study.

Three other patients developed minor complications during the early postoperative period: One had a left pleural effusion, one developed bronchopneumonia, and one had a wound infection (morbidity rate 21.4%).

The duration of hospitalization ranged from 10 to 45 days. In all cases the postoperative outcome was satisfactory. After surgery three patients were lost to long-term follow-up. The remaining 10 patients were followed every year according to our protocol for hydatid disease. Follow-up ranged from 17 months to 14 years (mean follow-up 5 years 4 months). One patient required a second operation 6 months postoperatively for removal of a femoral cyst. Another patient required a second operation for recurrent peritoneal hydatidosis 3 years later. The remaining patients are in a good general condition with no signs of recurrence.

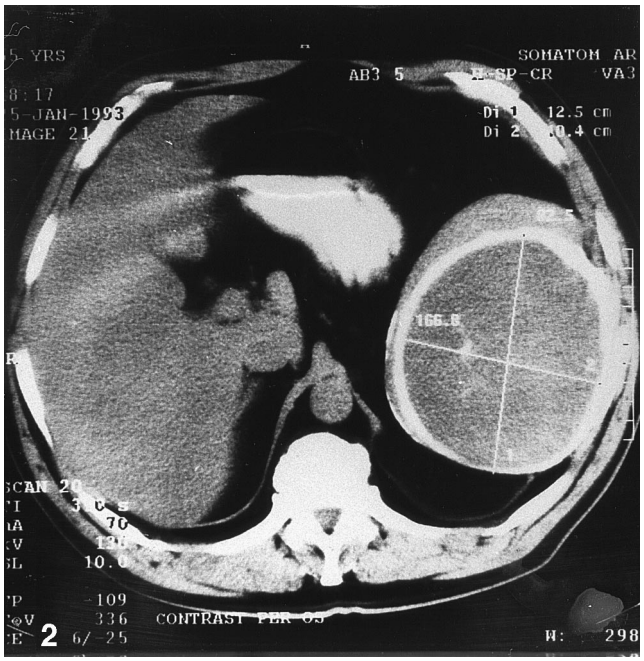

Fig. 1. Longitudinal sonogram of the spleen of a patient with splenic hydatidosis showing a large, round, anechoic lesion.

Fig. 2. Computed tomography of the upper abdomen indicating a large calcified cystic lesion of the spleen with a density similar to that of liver and spleen.

Discussion

Echinococcal infection is caused by the larval form of the cestode *Echinococcus*, with *E. granulosus* accounting for 95% of all cases of hydatid disease [4]. In our center and in most reports in the literature, splenic hydatidosis occupies the third place behind hepatic and pulmonary hydatid disease. Its frequency has varied between 0.9% and 8.0% in several reports [1, 3, 5]; in our series it is calculated as 4.64%. Associated hepatic or peritoneal disease occurs in 20% to 30% of cases (21.4% in our series) and constitutes a significant prognostic factor [5, 6].

Bourgeon et al. were the first to present a pathophysiologic and pathologoanatomic study for splenic hydatidosis [7]. The hexacanth embryo is embolized in the periphery of splenic capillaries. The incompressible mass of the cyst gradually crushes the segmentary vessels, with extensive pericystic atrophy and regional necrotic zones resulting. This aseptic inflammation of the splenic parenchyma extends to the outside of the splenic capsule, resulting in formation of adhesions to adjacent anatomic structures, (i.e., stomach, colon, and diaphragm). This situation predisposes to accidental rupture of the cyst to adjacent organs. We had only one case with infiltration of the diaphragm from the cystic wall. Cases with cyst rupture to adjacent viscera and most commonly the colon are also reported [1, 7–9].

In our series a single large hydatid cyst comprising most of the splenic parenchyma existed in all cases. Cases with multiple hydatid cysts of the spleen have been reported in the literature [3, 7].

The latent period of cyst development is usually long: 5 to 20 years [5, 6]. Splenic hydatid cysts develop insidiously and may reach a large size.

Clinical manifestations of splenic hydatid disease are usually mild and nonspecific. Half of our patients presented with mild abdominal discomfort as the only symptom. More severe pain in the left hypochondriac region may develop in patients with large cysts. A hard, round mass can be palpated under the left hypochondrium and may be considered a kidney tumour [7]. Complications such as secondary infection, cyst rupture into the abdominal cavity, and anaphylactic shock have been reported [1, 3, 10].

Splenic hydatidosis can be diagnosed preoperatively with radiologic studies. Before the development of cross-sectional imaging, radiographic evaluation of splenic cysts was limited to gastrointestinal radiographs and isotopic studies [1, 3]. The diagnosis of splenic cysts was greatly aided by the advent of ultrasonography and CT. However, sonographic and CT findings of splenic hydatidosis are not specific; and other splenic cystic lesions such as an epidermoid cyst, a splenic abscess, a pseudocyst, or a cystic neoplasm of the spleen may present with similar appearances on sonography and CT [3, 11]. The differential diagnosis is greatly helped by the history, the presence of calcification of the cyst wall, and especially the presence of daughter cysts in a large cystic lesion or concomitant cystic lesions in the liver or other organs [12].

Several immunologic tests also help in the diagnosis; the presence of the arc 5 of Capron et al. [13] on immunoelectrophoresis is the most reliable laboratory diagnostic tool for human hydatid disease.

As for hepatic hydatidosis, several surgical techniques have been applied to splenic hydatid disease. Many authors support conservative surgery, as partial splenectomy, cyst enucleation, or unroofing of the cyst wall with omentoplasty [2, 5, 10, 14–16]. The splenic cyst is evacuated using a large-gauge sump drain, and the residual cavity is filled with a 15% saline solution. These techniques are used for superficial cysts, cysts localized in one pole of the spleen, or cysts that are unresectable due to extensive adhesions. Partial splenectomy is a hemorrhagic operation because it is difficult to have vascular control when incising the splenic tissue, whereas unroofing the cyst wall leaves behind the

pericystic layer and a residual cavity, which carries the risk of postoperative infection [7]. For the above reasons and the possibility of multiple splenic cysts, we are in favor of splenectomy for the treatment of spleen hydatid disease, reserving partial excision with omentoplasty for cases with unresectable cysts tightly adherent to adjacent structures. Splenic salvage is also justified for pediatric cases to avoid septic complications [15, 17, 18]. In the case of associated hepatic involvement, splenectomy is followed by surgical management of hepatic cysts, usually omentoplasty [4, 18]. If there are multiple peritoneal cysts or the cysts exist in uncommon locations (e.g., in soft tissues), all cysts should be excised. After splenectomy individuals are susceptible to infectious complications and postsplenectomy sepsis [19]. Therefore all appropriate prophylactic measures should be undertaken to prevent them. In our series splenectomy had low morbidity and mortality rates (21.4% and 7.0%, respectively), similar to those observed after surgery for hepatic hydatidosis [12]. All patients received prophylactic vaccinations and a 2-year course of prophylactic penicillin. None developed serious septic complications.

In conclusion, splenectomy remains the therapeutic procedure of choice for splenic hydatidosis, offering complete cure of splenic and perisplenic disease with a low mortality rate.

Résumé

La rate est rarement touchée par la malade hydatique, même dans les pays endémiques. L'atteinte de la rate vient en troisième position, après celle du foie et du poumon. Dans notre série, l'échinoccocose splénique représente 5.8% des maladies hydatiques abdominales. Pendant les 22 dernières années, 14 patients ont été opérés pour des kystes hydatiques de la rate. Chez 10 patients, l'atteinte splénique était unique alors que chez deux patients, l'atteinte était concomitante avec celle du foie. Un patient avait une maladie abdominale disséminée et un dernier malade avait un kyste hydatique du muscle quadriceps. L'abdomen sans préparation, l'échographie et la tomodensitométrie ont été les examens les plus utiles pour établir le diagnostic. Tous les patients ont eu une splénectomie, soit seule soit en combinaison avec le traitement des autres kystes, exceptés deux patients qui ont eu une omentoplastie pour l'un et un drainage externe isolé pour l'autre. Un patient est décédé dans la période postopératoire immédiate (mortalité-7%) et trois patients ont eu des complications mineures. La maladie hydatique de la rate doit d'être considérée comme un diagnostic différentiel des kystes spléniques, particulièrement chez le patient ayant une maladie hydatique. La chirurgie reste la méthode thérapeutique de choix, de façon à éviter des complications.

Resumen

La ubicación esplénica es rara en pacientes con enfermedad hidatidica, aún en países donde la entidad es endémica. El bazo es el tercer órgano más comúnmente afectado luego del hígado y el pulmón. En nuestra serie la equinococosis esplénica representa el 5.8% de la enfermedad hidatídica abdominal. En el curso de los últimos 22 años, 14 pacientes fueron operados por quistes hidatídicos esplénicos en nuestro Departamento. En 10 pacientes el bazo fue el único lugar de ubicación de la enfermedad hidatídica hepática concomitante, en un paciente había enfermedad intraabdominal diseminada y en un paciente había un quiste hidatídico concomitante en el músculo cuadriceps femoral. Las radiografías simples de abdomen, el ultrasonido y la TAC fueron los exámenes de mayor utilidad en el establecimiento del diagnóstico. Todos los pacientes fueron sometidos a esplenectomía sola o combinada con el manejo de quistes ubicados en otro sitios, excepto dos pacientes que fueron sometidos a esplenectomía y un paciente que recibió drenaje externo. Un paciente murió en el postoperatorio immediato (tasa de mortalidad: 7%) y tres pacientes exhibieron complicaciones menores. La enfermedad hidatídica esplénica debe ser incluida en el diagnóstico diferencial en presencia de un quiste esplénico, especialmente en pacientes con historia de enfermedad hidatídica. La cirugía sigue siendo el tratamiento de escogencia, y se debe realizar con el objeto de evitar complicaciones graves.

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Invited Commentary

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All of the reported literature on hydatid (echinococcal) disease stresses the rarity of splenic involvement, as related by the authors of this article. One is reminded of the rarity of splenic metastases in malignant disease, in which the spleen is also a reluctant host organ. Why the spleen is not vulnerable in these instances of hematogenously disseminated diseases remains a mystery.

As rare as hydatid disease of the spleen is in countries where it is endemic, it is virtually nonexistent in Western countries where conditions for infection are not optimal. Nevertheless, as the world "shrinks" through air travel and emigration, acquaintance with treatment of esoteric maladies such as hydatid disease should be part of every physician's armamentarium.

The authors relate an interesting spectrum of both isolated splenic hydatid disease and splenic involvement in disseminated disease. In contrast to nonparasitic splenic cysts, for which treatment for small cysts may be expectant, I agree that these parasitic splenic cysts require surgical intervention. Among the complications reported with hydatid cysts of the spleen, in addition to those mentioned by the authors, are splenothoracic prolapse, bronchosplenic fistula, and hypersplenism.

Although enucleation of splenic hydatid cysts and partial splenectomy for splenic hydatid disease have now been reported, I am in agreement with the authors that total splenectomy is the treatment of choice for this disease for the reasons mentioned in the article. Moreover, I believe it is even justifiable in older children, given the grave consequences of complications of this condition and the low incidence of postsplenectomy sepsis. I think these are cogent reasons for total splenectomy, rather than the technical difficulty associated with partial splenectomy. The latter need not be a "hemorrhagic" operation. Finally, it might be in order to mention drug therapy with albendazole or mebendazole as possible adjuncts to surgical therapy for severe cases of splenic hydatid disease.