

CASO CLÍNICO

Splenic hydatidosis: a rare differential diagnosis in a cystic lesion of the spleen

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SUMMARY. Cystic tumours of the spleen are generally rare, and a parasitic origin is relatively unlikely. The present case report shows, however, that when a splenic cyst is found, the differential diagnosis must always consider the possibility of echinococcosis. We report the case of a patient suffering from a cystic lesion of the spleen where surgery and histopathology yielded the diagnosis of splenic echinococcosis. Abdominal pain in the left upper quadrant and splenomegaly detected by simple abdominal radiology are the most commonly found indicators for this disease. The treatment should be surgical, attempting to preserve as much splenic tissue as possible, although conservative treatment is frequently unfeasible due to massive involvement of the spleen. Although rare, splenic hydatidosis should be included in the differential diagnosis when a cystic splenic lesion is identified with sonography or CT scan.

Key words: Splenic hydatid cyst, hydatidosis, echinococcosis, spleen.

RESUMEN. Los tumores del bazo son raros y más aún cuando son de origen parasitario. El caso que se presenta demuestra que cuando se encuentra un quiste en el bazo se debe realizar diagnóstico diferencial con equinococosis. Se informa el caso de una paciente que presentaba una lesión quística en el bazo, que tanto quirúrgicamente como histopatológicamente correspondió al diagnóstico de equinococosis esplénica. La sospecha diagnóstica se sustenta en la presencia de dolor abdominal en el cuadrante superior izquierdo y esplenomegalia detectada por medio de una placa simple de abdomen. El tratamiento es quirúrgico, procurando conservar la mayor parte de tejido esplénico, aunque el tratamiento conservador frecuentemente no es posible debido al involucro masivo del bazo. Aunque la equinococosis esplénica es rara, se debe incluir en el diagnóstico diferencial cuando se detectan lesiones esplénicas por ultrasonido o tomografía.

Palabras clave: quiste hidatídico del bazo, hidatidosis, equinococosis, bazo.

INTRODUCTION

Cystic lesions of the spleen include parasitic and nonparasitic cysts.^{1,2} Parasitic cysts represent 50-80% of the splenic cysts, most of them due almost exclusively to echinococcal disease.³ However, splenic involvement is rare in patients with hydatid disease, even in endemic countries, but represents the third most commonly involved organ after the liver and the lung.⁴⁻⁷ In 1790, Berthelot reported the first case of splenic hydatidosis and in 1954 Mills reported fifty cases found in the literature.⁸ In Mexico, hydatid disease is an uncommon finding and few cases have been informed.⁹⁻¹⁴ To our knowled-

ge, only a previous autochthonous case of splenic echinococcosis has been reported in Mexico.¹⁵

CASE REPORT

Female patient, 23 years old, born in Mexico City, with no previous traveling and belonging to the low socioeconomic class, had always owned dogs and frequently eaten lamb meat. The patient consulted us because of abdominal pain and the presence of a painful, slowly-growing mass in the left hypochondrium for the last five months. The patient denied chills, fever, weight or appetite loss, itching or other symptoms. Seven days

prior to admission she presented changes in bowel habits without blood or mucus in the stools. On admission her blood pressure was 110/70 mm Hg with a heart rate of 72 beats per minute and a rectal temperature of 36.2 °C. No jaundice or cutaneous malformations were found. A soft 5 mm cervical adenopathy was palpated. Physical exploration was limited to the abdomen where a firm, slightly tender to palpation mass was found 5 cm below the left costal border. No pulsations, nodules, guarding or rebound were found in this mass, apparently coming from the intraperitoneum. Laboratory data on admission included hematocrit 37.7%; WBC 3,400/mm³ with 1% eosinophils and 55% neutrophils; prothrombin time (control/patient) 13.9/13.3 seconds, total bilirubin 0.4 mg/dL, alkaline phosphatase 92 UI/L, AST 17 UI/L, ALT 12 UI/L, and seric albumin 4.1 g/dL. The chest X-ray was within normal limits. Plain abdominal films showed a high density mass in the upper left quadrant. Upper gastrointestinal series revealed an extrinsic mass compressing the greater curvature of the stomach.

Real-time sonography with a 3.5-MHz sector transducer (Sonoline AC, Siemens) showed a well-defined anechoic cystic lesion which was difficult to distinguish from the spleen. A CT scan showed a 12 x 14 cm cystic lesion of the spleen without wall calcification (*Figure 1*). At laparotomy, a 15 x 10 cm white cystic lesion on the spleen was found; the spleen weight was 389 grams; therefore splenectomy was performed. No other lesions were found in the abdominal cavity. Pathologic examination of this material revealed a turbid fluid (hydatid sand) which showed the presence of protoscolices (*Figure 2*) and loose hooklets, both of which confirmed the diagnosis. In histopathologic sections, the intermediate acellular laminated layer and the inner germinal layer of

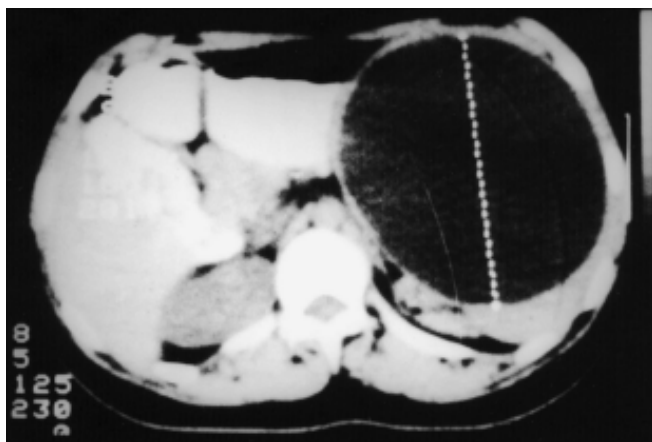


Figure 1. CT of the abdomen showing a large echinococcal cystic lesion of the spleen (12x14x8 cm) without wall calcification.

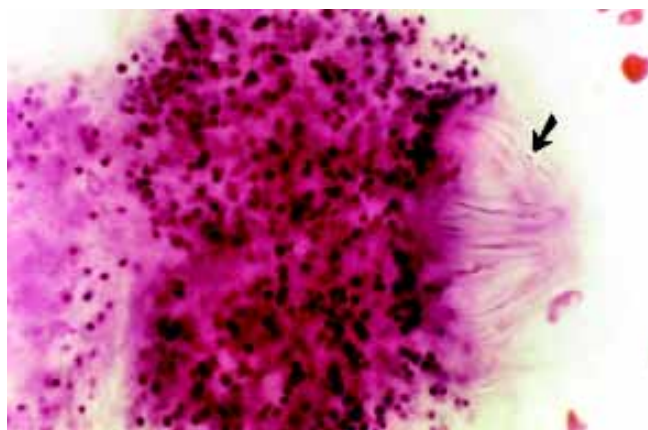


Figure 2. Exvaginated protoscolex of *Tenia echinococcus* showing hooklets (arrow). Hydatid sand sample taken during histopathologic procedure.

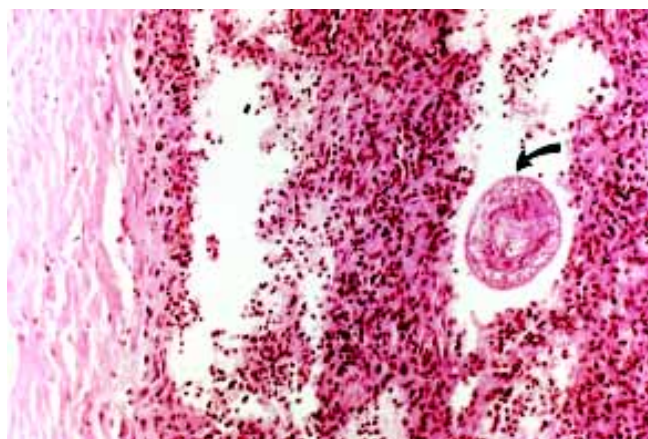


Figure 3. A histopathologic section of cyst wall show the intermediate acellular laminated layer and the inner germinal layer showing a viable protoscolex (arrow) (H & E 100x).

the cyst wall each had a characteristic appearance of their own (*Figure 3*). Albendazol at a dosage of 10/mg/kg/day during 28 days was started. Today, the patient remains asymptomatic after a nine-year follow-up.

DISCUSSION

The word “echinococcus” originates from the Greek meaning “hedgehog berry”, a descriptive term of the gross pathology of the lesion.⁴ Another descriptive Greek word applied to this disease is “hydatid”, meaning “a drop of water”.⁵ In the human host, metacestode cysts may inhabit virtually all anatomic sites following oral ingestion of eggs (primary echinococcosis). Secondary echinococcosis results from the spread of *E. granulosus* metacestodes from the primary sites via blood vessels to distant organs or by rupture of cysts into the peritoneum,

pleura, bronchial tree or biliary ducts. The majority of the patients have single-organ involvement and harbor a solitary cyst. The size of the cyst is variable but usually ranges from one to fifteen centimeters. The cysts growth rate is variable (one to 31 mm per year) and 16% of cysts do not expand or collapse.

The localization of the hexacanth embryo within the spleen is quite rare, with an incidence of 0.5-5%, although this organ is the third most frequently affected after the liver and the lung according to some series,⁸ or the fifth organ in adults and sixth in children in other series.^{16,17} The infestation of the spleen usually takes place by arterial route when the parasite has passed through the two other filters: the hepatic and pulmonary. The retrograde venous route, the portal circulation, the hemorrhoidal veins and the lymphatic channels may also be considered.¹⁸ For his rarity, signs and symptoms are difficult to define and most are case reports. Dieulafoy was the first physician to diagnose splenic hydatidosis.⁴ After a thorough review of the world literature, abdominal pain in the upper left quadrant can be considered as the most common finding.¹⁶⁻²⁰ Other symptoms are cough, dyspnea and changes in bowel habits, but patients may as well be asymptomatic.¹⁷ An abdominal mass is the most common finding during physical examination, usually without fever. The presence of hydatid fremitus has also been described.¹⁹ Routine laboratory tests as a rule do not show specific results; however, in most cases eosinophilia is low (<15%) or absent.⁸ The most common finding in X-rays is a hyperdense mass or calcifications in the splenic area.^{21,22} The typical ultrasonographic image is an anechoic lesion in 38%-48% of all cysts. Mixed echoic (about 10%) or echodense structures (about 7%-8%) are observed in cysts filled with folded membranes or more solid masses. Internal septation (cart wheel sign) is characteristic for multiple daughter cysts within a larger mother cyst. A spherical rim (eggshell pattern) is a typical sign of a usually inactive cyst. Computed tomography²¹ permits better documentation of its site, size, and structure. Magnetic resonance imaging²¹ has no major advantage over CT in the diagnosis, but is superior in identifying changes of the intrahepatic and extrahepatic venous system. Immunodiagnostic tests as Casoni's skin test and Weinberg's hemagglutination test⁸ for the detection of specific serum antibodies or circulating antigens are highly relevant for the diagnosis but once a cyst is formed, 10%-40% do not produce detectable serum antibodies and exhibit false-negative testing results that may be misleading. In our patient no immunodiagnostic tests were performed.

Treatment options for patients infected with *E. granulosus* are usually categorized into surgical or chemotherapeutic approaches.⁸ Frequently, a combination of both treatments is used. Selection of the appropriate therapy may depend on several factors including the age and health status of the patient, the severity of the symptoms, the skills of the clinician in charge of the patient, the availability of sophisticated imaging procedures, and the size, number, location, and viability of the hydatid cysts.

Surgical management continues to be the most appropriate therapeutic approach. Total splenectomy is advocated by the majority of the surgeons, since it provides a minimal risk of recurrence.^{23,24} Splenectomy however is associated with sepsis-related deaths in 1.9% in adults and 4% in children.²⁴ Thus, conservative surgical procedures have increasingly been proposed including partial splenectomy, enucleation, de-roofing with omentoplasty, internal drainage with cystojejunal anastomosis or external drainage.²⁴ Laparoscopic treatment²⁵⁻²⁷ has been performed, but reluctance to perform laparoscopic surgery is probable because of the concern of spillage of the fluid into the peritoneal cavity, with the possibility of anaphylactic reaction and recurrence.²⁷ An alternative to surgery is percutaneous drainage and administration of sclerosing agents such as alcohol 96% under sonographic guidance.²⁸ Medical treatment with albendazole, mebendazole and praziquantel has been shown to be effective in nonsurgical cases, in patients who refuse surgery or as a prophylactic measure before, during or after surgery if spillage occurs, so as to minimise recurrence.²⁹ In our case, the patient had been asymptomatic for nine years; peritoneal recurrence can present 4 to 15 years after splenectomy.³⁰

True cysts of the spleen are very rare and include epidermoid and dermoid cysts, cystic hemangiomas and cystic lymphangiomas.²

In Mexico, most of the hydatid disease cases have been reported in immigrants from Spain or South America,¹³ in whom *E. granulosus* was common but, in turn, few had autochthonous hydatid disease; however, the frequency for Mexican citizens is unknown, although infection from domestic animals such as cattle, sheep and pigs in some degree has been detected in many parts of the country, particularly in Culiacan (Sinaloa), where 6.3% of the slaughtered pigs examined were positive for *Echinococcus*.^{31,32} In 1880 Bandera⁹ informed the first case in Mexico affecting the liver. Few case reports have been informed affecting other organs. Our case is the second autochthonous splenic hydatid cyst informed in Mexico. The other case was informed by Menendez-

Arzac, et al. in 2002 in a pregnant woman.¹⁵ Hydatid disease should be looked for in patients with cysts or with presumptive abdominal images of this disease.

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