

Spontaneous Rupture of a Splenic Hydatid Cyst with Anaphylaxis in a Patient with Multi-Organ Hydatid Disease

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Rezumat

Ruptură spontană a unui chist hidatic splenic și anafilaxie la un bolnav cu boală hidatică multi-organ

Chisturile hidatice ale splinei sunt rare, splina fiind al treilea viscer din punct de vedere al frecvenței afectat de Echinococcus Granulosus. Chisturile hidatice splenice sunt, de cele mai multe ori, însoțite de boală hidatică multi-organ. Diagnosticul se face des la investigarea unei splenomegalii sau întâmplător, în timpul unui alt consult. Diagnosticul se poate face și după ruptura chistului fie datorită unui traumatism (cel mai frecvent), fie spontan. Ruptura chistului hidatic splenic necesită terapie imediată și este un eveniment cu potențial fatal. De cele mai multe ori se soldează cu splenectomie. Prezentăm cazul unui pacient cu boală hidatică pluri-viscerală care s-a prezentat cu un chist hidatic splenic rupt și a dezvoltat fenomene de șoc anafilactic. Cazul a fost tratat prin splenectomie și a avut o evoluție favorabilă.

Cuvinte cheie: chist hidatic splenic, boală multi-organ, anafilaxie

Abstract

Hydatid cysts of the spleen are a rare occurrence, the spleen being the third most common organ for the development of Echinococcus Granulosus. Splenic hydatid cysts are commonly part of multi-organ hydatid disease. Diagnosis is often established when investigating a splenomegaly or by chance during an unrelated consult. It can also be diagnosed after rupture, be it following trauma (the most common occurrence) or spontaneous. Splenic hydatid cyst rupture requires immediate action and is a life-threatening condition. It results, most often, in splenectomy. We present the case of a patient with multi-organ hydatid disease that presented with a ruptured splenic cyst and developed anaphylaxis. The case was resolved by splenectomy and recovered well.

Key words: splenic hidatid cyst, multi-organ hidatid disease, anaphylaxis

Introduction

Cystic echinococcosis (CE) is the larval cystic stage of the tape worm Echinococcus granulosus and can cause illness in intermediate hosts like humans. It is slightly more common in women than men in endemic areas, and can affect all ages. Southeast Europe, and thus Romania, is considered a highly endemic area in which the infection can pose serious public health problems.

Splenic involvement is the third most common location of the cystic disease. It follows liver and lung sites. Only 10 to 20% of the hexacanth embryos escape the double liver-pulmonary circulation (Lemman's filters) and can spread to any other organ

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or apparatus (1). Other pathogenic pathways like contiguity, lymphatic and retrograde venous contamination have been proposed in order to explain the cases with isolated splenic disease. In 20 to 65% of cases the splenic condition is accompanied by other organs, most commonly the liver or peritoneum (2).

Early stages are usually asymptomatic and the diagnosis may often be incidental, associated with abdominal ultrasound performed for other clinical reasons. In endemic areas, the presence of symptoms suggestive of CE in a person with a history of exposure to animal carriers, supports the suspicion of hydatidosis. Immunodiagnosis is used to confirm radiological findings. Ultrasound is the gold standard for defining number, site, dimensions and vitality of the cysts and it also helps to plan treatment. A standardized ultrasonographic classification has been developed by the World Health Organization (3), updating the older Gharbi classification (4).

Splenic hydatid cysts can be complicated by secondary infection and fistulization to adjacent organs, even above the diaphragm. Systemic anaphylaxis can occur following rupture of the cyst in the peritoneal cavity (5). The rupture is traumatic in most cases but can also occur spontaneously. Radical surgery is the mainstay of treatment and chemotherapy is added using antihelminthic regimens to avoid recurrence (6).

Case Report

We present the case of I.C. a female patient of 43 years old that presents to the emergency department of our institution for pain in the lower abdomen, malaise, nausea and emesis. She presents no relevant family history. The pain started 2-3 hours before presentation; its debut was sudden and was quickly followed by nausea and emesis. She reports no history of trauma, denies alcohol, cigarettes or drug consumption. She reports no significant medical history outside an appendectomy at age 12. She also reports no contact with sheep or dogs at home or at work.

Clinical examination reveals a patient of normal weight (BMI – 23.5). She is pale, tachycardic (135 bpm) with a BP of 80/50 mmHg. On general inspection no lesions suggestive of trauma are found. She has a tender abdomen, especially in the lower quadrants with localized muscle guarding. Peritonitic signs are present in the lower abdomen (Blumberg, Mandell). Palpation reveals a round tumor in the lower abdomen that extends 10 cm above the pubic bone, is mobile and has a firm consistency. Rectal examination reveals pain at the palpation of the cul-de-sac Douglas. Gynecological consult reveals no pathological findings; the patient is found to have a normal uterus with healthy adnexa.

Complete blood count reveals leucocytosis (WBC – 13500/uL) with an increase of the neutrophils (NEU – 11730/uL). Blood chemistry records a glycemia of 255 mg/dl and is otherwise insignificant. Coagulation assays are normal. Simple abdominal x-ray reveals no pathological findings. Ultrasound of the abdomen records a large fluid collection in the peritoneal cavity (Douglas pouch, Morisson, left upper quadrant) and a cystic lesion (62/45 mm) in the lower abdomen

left of the uterus; the spleen cannot be visualized and the rest of the investigation is normal.

During these investigations the patient is administered i.v. fluids during which there is no increase in blood pressure and she remains tachycardic. The presence of fluid in the peritoneal cavity, peritonitic signs and low blood pressure prompted surgical intervention. The patient is taken to the OR for exploratory laparotomy.

Exploratory laparotomy is performed under general anesthesia through a midline incision above and below the umbilicus. We recorded about 300 ml clear liquid in the peritoneal cavity on opening. A large cystic lesion of the spleen was found (Fig. 1) that takes up about 2/3 of the organ. The cyst is ruptured and has multiple, smaller, cystic lesions inside. We found a second, intact cystic lesion with a diameter of about 6 cm in the sigmoid mesocolon (Fig. 2). Numerous, small cysts were found disseminated in the peritoneal cavity (Fig. 3). We found no cysts of the liver. Splenectomy followed by mesosigmoid cystectomy was performed. The peritoneal cavity

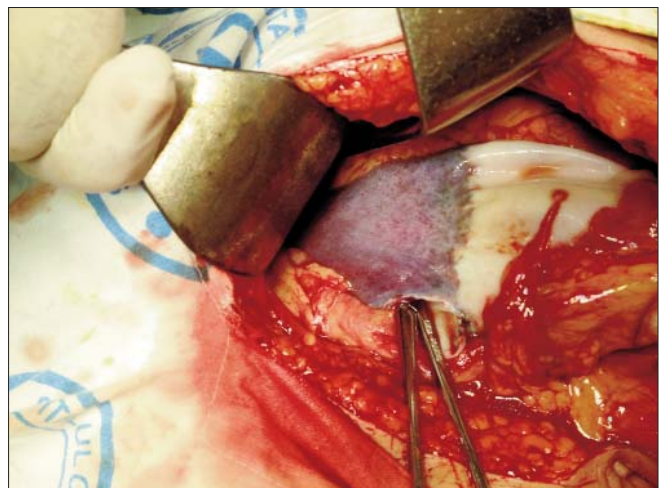


Figure 1. Ruptured cystic lesion of the spleen

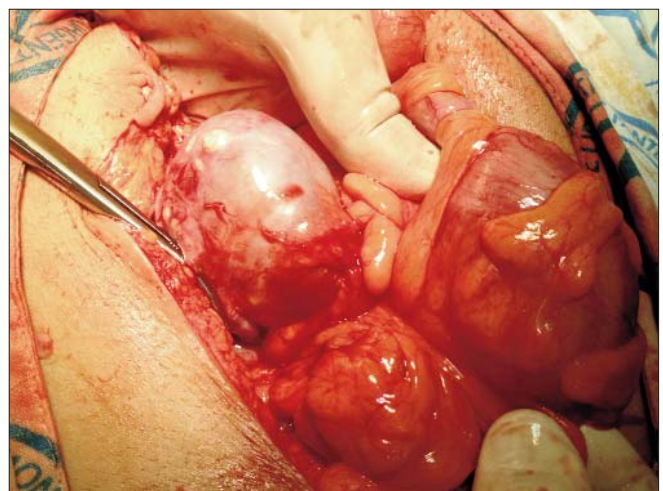


Figure 2. Intact cystic lesion of the sigmoid mesocolon

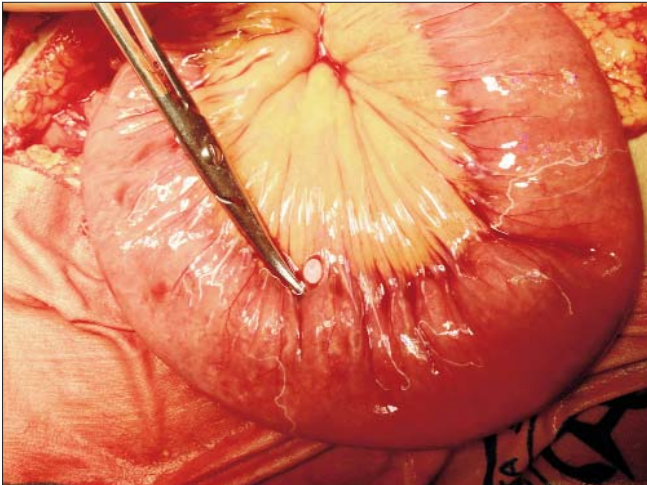


Figure 3. Small cystic lesion on the serosa of the small bowel



Figure 4. Cystic lesion of the spleen; resection specimen

was irrigated with hypertonic saline and two drains were placed (one in the cul-de-sac Douglas and one in the left paracolic gutter). Pathological studies confirmed the hydatid nature of all the cystic lesions.

Postoperative course was uneventful. The patient was started on i.v. sulperazone (4g/day) and received analgesics as needed. Drainage was suppressed on the fourth postoperative day and bowel movements were recorded on the fifth. She is discharged after a six day hospital stay with total digestive tolerance, minimal pain and a good general health state. She received albendazole (400 mg twice daily) for 3 months postoperatively. Anti-pneumococcal vaccine was administered one week after surgery. Clinical and ultrasound examination at 3 months postoperatively revealed no pathological findings.

Discussion

The case we presented showed the management of hydatid disease of the spleen with spontaneous rupture of the cyst and anaphylaxis. Spontaneous cyst rupture is rare and the lack of traumatic history can make the diagnosis difficult.

The presentation is often dramatic because of the gross spillage of the contents that not only cause chemical peritonitis but can cause anaphylaxis. Postoperative diagnosis can be missed if suggestive history is absent. The combination of clinical exam and ultrasound can, however, suggest the diagnosis.

Treatment has to be done expeditiously to prevent spreading of chemical peritonitis and to prevent disseminated peritoneal echinococcosis. Surgical therapy is the mainstay but the exact approach, radical or conservative is still not defined. Total splenectomy is probably better suited for large cysts that take up most of the splenic parenchyma. It also has the advantage that removing the diseased organ prevents local recurrence. Atmatzidis et al reported no significant difference between patients in the splenectomy and spleen-preserving groups with regard to hospital median stay, recurrence rate and postoperative complication rate (7).

To prevent recurrences, albendazole therapy is mandatory. It should be started immediately and given for a prolonged period of time (8). Some authors recommend that follow-up should be confined to a parasitologist or infectious disease specialist (9). While we agree that their participation is mandatory, we also feel that surgeon involvement in long-term follow-up is beneficial to patients. Also, given the infectious nature of the disease, we feel that correct reports should be mandatory for every case encountered.

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