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Total Splenectomy for a Giant Isolated Splenic Hydatid Cyst Compressing the Abdominal Viscera: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABDEF 1 **Dimitrios Chatzifotiou**
BEF 1 **Christian Wolf**
BEF 1 **Arthurs Baibakovs**
CD 2 **Harald Werthebach**
BEF 1 **Bogdan Lupascu**
BEF 1 **Martin Schnell**

1 Department of General, Visceral and Thorax Surgery, Hegau Bodensee Clinic, Singen (Hohentwiel), Germany
2 Department of Diagnostic and Interventional Radiology, Hegau Bodensee Clinic, Singen (Hohentwiel), Germany

Corresponding Author: Dimitrios Chatzifotiou, e-mail: dimitrakis2811@outlook.com.gr
Conflict of interest: None declared

Patient: Female, 49-year-old
Final Diagnosis: Echinococcus infection
Symptoms: Left upper quadrant abdominal pain
Medication: —
Clinical Procedure: —
Specialty: Surgery





Objective: Rare disease
Background: Extrahepatic and extrapulmonary localizations of cystic echinococcosis (CE) are rare and the incidence of splenic involvement is seen in 1.0% to 3.3% of all cases in the endemic areas of the Middle East and Asia. The diagnostic pathway consists of a detailed travel history, physical examination, abdominal ultrasonography, computed tomography, and serological tests. The efficacy of perioperative administration of albendazole (400 mg twice a day) has been proven; however, the appropriate surgical procedure for the treatment of giant, centrally located splenic hydatid cysts remains controversial.

Case Report: We present the case of a 49-year-old woman referred to our hospital for a suspected isolated splenic hydatid cyst causing a compression of the right kidney, stomach, and the tail of the pancreas. She reported chronic pain in the left upper quadrant and a history of contact with animals. She underwent open splenectomy via a medial to lateral approach to minimize manipulation of the spleen. In addition, she received perioperative parasitostatic drug therapy with albendazole. The postoperative period was uneventful and the histologic analysis confirmed the diagnosis.

Conclusions: The spleen is a rare location for isolated CE, especially in non-endemic areas and must be considered in the differential diagnosis of splenic cystic masses. Surgical resection remains the most effective treatment that completely resolves this condition. A comparison of randomized trials is needed to compare the recurrence rates between splenectomy and spleen-preserving procedures in the treatment of giant splenic hydatid cysts.

Keywords: Echinococcosis • Splenectomy • Splenomegaly

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Background

A hydatid cyst is the most common form of echinococcosis in humans. Although it presents as an endemic transmission in some areas of South America, Asia, and Africa, it is rare in European countries. The disease is mostly localized in the liver and lungs, although other organs can be affected due to systemic dissemination. An isolated spleen involvement is rare and presents only in 0.7% of the infected patients [1]. Large cysts can be symptomatic causing a compression of the adjacent abdominal organs. Cystic echinococcosis (CE) has a good prognosis in the majority of cases. The perioperative mortality rate is 0.8% and anaphylactic shock is the main cause of death [2]. This case report presents the rare case of an isolated splenic hydatid cyst, which should be considered in the differential diagnosis of cystic lesions of the left upper abdomen, especially in non-endemic regions.

Case Report

A 49-year-old Balkan woman living in a rural area presented to her general practitioner reporting 6 months of left upper quadrant pain without tenderness. Her past medical, pharmacological, and surgical history was unremarkable. On physical examination, a mass occupying the left upper abdomen was palpated. Ultrasonography revealed an uncertain cystic mass with solid components. Computed tomography (CT) of the abdomen and pelvis with an intravenous contrast showed a 22×18×13 cm septated splenic cyst with scattered calcifications and transient enhancement of the wall after contrast administration (Figure 1). The voluminous cyst had displaced the right kidney, stomach, and pancreas (Figure 2). No liver cysts were identified and the inflammatory markers were normal. On suspicion of CE, the patient was referred to our clinic. She mentioned contact with animals in her hometown. A CT scan

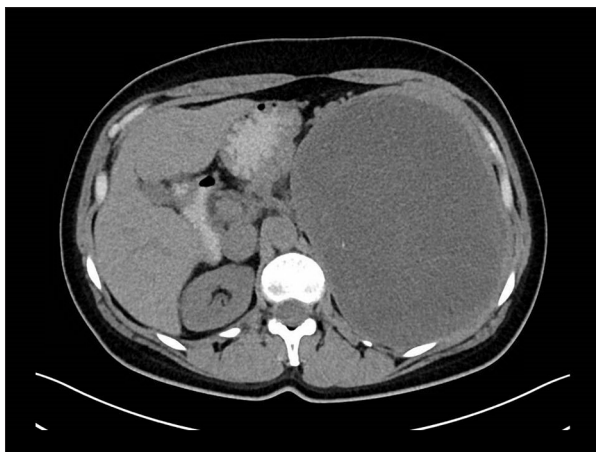


Figure 1. Computed tomographic scan of the patient in an axial plane.

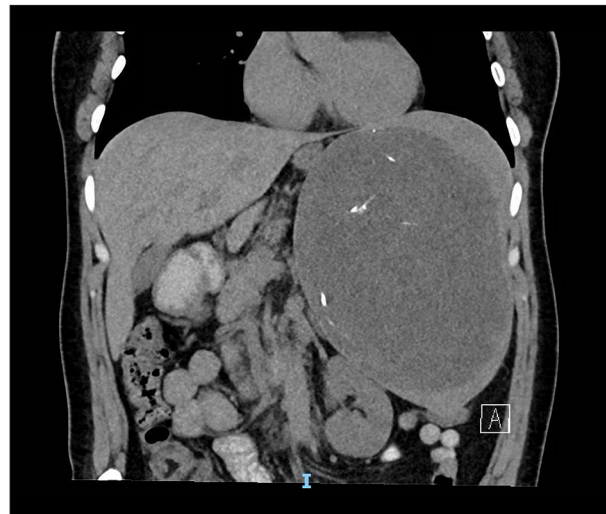


Figure 2. Computed tomographic scan of the patient in a coronal plane.



Figure 3. The specimen after the resection.

of the lungs and a magnetic resonance imaging of the brain did not detect any additional lesions. The *Echinococcus* IgG enzyme-linked immunosorbent assay (ELISA) test was negative. A splenectomy was recommended due to the cyst's size and the compression of the splenic parenchyma.

Two weeks before the hospital admission, she was vaccinated against *Pneumococcus*, *Influenza*, and *Meningococcus*. We ordered an anti-infective albendazole (ABZ) treatment. She underwent open splenectomy via a medial to lateral approach with a supraumbilical midline incision. The medial to lateral approach was preferred to minimize specimen manipulation and the risk of cyst perforation (Figure 3). The postoperative period was uneventful and she was discharged on the 8th postoperative day. She received an adjuvant treatment based on ABZ for 1 month (400 mg twice a day). The histological analysis

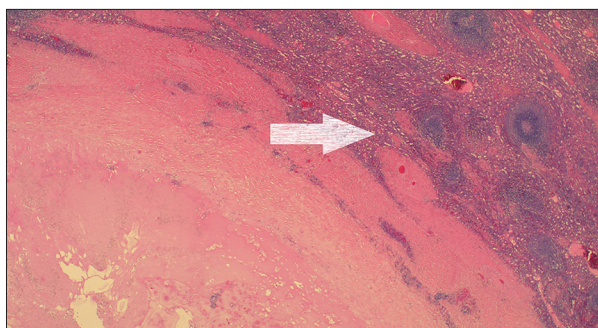


Figure 4. Histologic slide: The calcified cyst wall (white arrow). Scolices cannot be found in inactive cysts.

revealed a parasitic cyst with an outer pericyst (**Figure 4**). The weight of the specimen was 3.05 kg. Cyst fluid cytology confirmed the diagnosis of the CE. The first follow-up 6 months after surgery consisted of a clinical evaluation, abdominal ultrasound, echocardiogram, and a chest X-ray, which did not detect any signs of infection recurrence. The patient gave informed consent for the publication of this case report.

Discussion

CE due to *E. granulosus* is a rare parasitic disease in European countries. The majority of cases are seen in patients who immigrated from areas of high endemicity. According to the 2018 annual report of the Robert Koch Institute, 83 cases of CE were identified in Germany without any isolated splenic echinococcosis [3].

Hydatid disease can affect any organ, although the most common localization sites are the liver and the lungs. According to a systematic review, a splenic echinococcal involvement develops in 1% to 3.3% of the patients [4]. Other studies report this percentage to be between 2% and 5% [5,6]. However, most reported cases had concomitant hepatic hydatidosis, showing

a systematic dissemination. According to a study from China, only 0.7% of 3003 patients with CE had an isolated splenic involvement [3]. There is no universally accepted size to define a hydatid cyst as ‘giant.’ A large hydatid cyst causing a significant enlargement of the affected organ or with exophytic growth through the natural routes provided by the organ capsule is defined as a giant hydatid cyst [7].

Diagnosis is based on clinical examination, imaging techniques, and serological tests. Hydatid cysts are mostly asymptomatic or can present with mild, dull, and non-specific abdominal pain, justifying the diagnostic delay [8]. The differential diagnosis incorporates other cystic formations of the spleen, including an epidermoid cyst, a pseudocyst, an abscess, a hematoma, and a cystic neoplasm of the spleen [9].

Ultrasonography is the diagnostic tool of choice for the detection of cystic lesions. Based on ultrasound imaging, the World Health Organization’s Informal Working Group for Echinococcosis (WHO-IWGE) has classified CE into 5 types. A CT is indicated for the subdiaphragmatic localizations of disseminated disease and presurgical evaluations [10]. The main radiological characteristics for hepatic and splenic hydatid cysts can vary from simple cysts to a solid appearance. However, solitary components are mainly found in splenic cysts [8,11]. Calcifications, a daughter cyst sign, hydatid membranes, and hydatid sand are other typical findings of the disease. In the present case, the CT revealed a type 4/5 CE hydatid cyst according to the WHO-IWGE classification (**Table 1**).

Serology has a complementary role in the primary diagnosis of the disease, although it is also useful for the follow-up. The ELISA tests have progressively replaced the tests with low sensitivity and specificity, including the Cassoni intradermal test, latex agglutination test, and indirect hemagglutination test. The enzyme-immunoassay system for the determination of IgG antibodies against *E. granulosus* provides high

Table 1. Summary of the WHO classification of hydatid cysts adapted from the 2010 WHO Informal Working Group on Echinococcosis.

Cyst type	Stage	Imaging features
CE1	Active	Unilocular simple cyst
CE2	Active	Multivesicular, multiseptate cyst. Daughter cysts partly or totally fill mother cyst. “Wheel” or “honeycomb” appearance
CE3a	Transitional	Detached laminated membrane floats in cyst (water lily sign). Anechoic content
CE3b	Transitional	Complex mass. Mother cyst contains both anechoic daughter cysts and echoic areas of disrupted membranes or degenerating daughter cysts
CE4	Inactive	Heterogenous hypoechoic cyst without daughter cysts. Degenerating membranes may appear like “ball of wool”
CE5	Inactive	Thick calcified wall

sensitivity with limited specificity because of cross-reactions due to the possible coexistence of *Taenia solium* infection, infections of other helminth diseases, malignancies, and liver cirrhosis [10,12]. According to a systematic review, the sensitivity of the *Echinococcus* IgG ELISA test varies from 72% to 96%, with a specificity between 40.8% and 97% [13]. The use of recombinant antigens based on *E. granulosus* antigen B and recombinant antigen 5 can increase the diagnostic value of the IgG ELISA test [14]. Even in these tests, the diagnostic accuracy is limited in case of inactive cysts, cysts with extrahepatic localization, and in patients who have already received ABZ chemotherapy [15].

Surgical treatment is the main therapeutic option for patients with splenic hydatid cysts. The aim of the surgical procedure is cyst resection without intraoperative contamination of the abdominal cavity [10]. A percutaneous treatment with puncture, aspiration, injection of the protoscolicidal agent and reaspiration (PAIR), a watch and wait approach, or a long-term parasitostatic chemotherapy cannot be considered as alternative treatments for patients without any significant contraindications for surgery [10]. For patients with splenic hydatid cysts, a splenectomy is considered the treatment of choice providing a complete removal of the cyst with a low recurrence rate. On the other hand, it is associated with an overwhelming sepsis syndrome after the splenectomy. Thus, many authors suggested a spleen-preserving procedure for younger patients or in cases of peripherally located cysts, which occupy <50% to 75% of the parenchyma [3,16]. Spleen-preserving procedures can be a partial splenectomy, a partial cystectomy, or a cystotomy using flushing with 20% saline solution, and an omentoplasty [14]. Further studies with longer follow-ups are needed to estimate the recurrence rates in patients who undergo a partial cystectomy or cystotomy. In the present case, we performed an open splenectomy to avert a cyst perforation by manipulation of the specimen. A laparoscopic approach could be an optional procedure in patients with small cysts due to the low perforation risk. If a laparoscopic operation is performed, it is essential to adhere to the same technical standards that apply to corresponding open procedures. To the best of our knowledge, there are several case reports and case series in the bibliography, using various surgical procedures [17-21].

The surgical treatment must be combined with the perioperative administration of benzimidazole-carbamate compounds (BMZ). Among the BMZ compounds, ABZ and mebendazole are currently the only licensed drugs for the systemic treatment of echinococcosis [10]. ABZ has better bioavailability and is the preferred first-choice parasitostatic drug with a daily dose of 15 mg/kg divided in 2 doses [22]. The suggested duration of ABZ administration for patients after a radical and uncomplicated resection of the hydatid cyst is 4 weeks to 8 weeks [23]. The histological and cytological examinations are mandatory to confirm the presence of a parasitic cyst.

Conclusions

Although the spleen is a rare location for an isolated hydatid cyst, it should be included in the differential diagnosis of splenic cysts, especially in non-endemic countries. Despite the fact that surgery is the standard therapeutic option, there is no established consensus regarding the type of surgical procedure. We support the opinion of most surgeons that the main aim of the surgical procedure is complete resection of the hydatid cyst without any intraoperative peritoneal spillage. Therefore, we prefer a total splenectomy performed with a medial to lateral approach for giant hydatid cysts with a central localization in the parenchyma.

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Department and Institution Where Work Was Done

Department of General, Visceral and Thorax Surgery, Hegau Bodensee Clinic, Singen, Germany.

Conflicts of interest

None.

Declaration of Figures Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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