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

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

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

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/931195
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 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 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.

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.
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 sensitivity, specificity, and good performance for diagnosis [8].

### 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| Dated laminated membrane floats in cyst (water lily sign). Anechoic content |
| CE3b      | Transitional| Complex mass. Mother cyst contains both anechic daughter cysts and echoic areas of disrupted membranes or degenerating daughter cysts |
| CE4       | Inactive| Heterogenous hypoechogenic cyst without daughter cysts. Degenerating membranes may appear like “ball of wool” |
| CE5       | Inactive| Thick calcified wall |
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.

Acknowledgements

We are grateful to Dr. Ulrich Oehler and Dr. Christian Szalai (Department of Diagnostic Pathology, HBK Singen, Baden-Wurttemberg, Germany) for their pathological assessments.

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.
References:

1. Ran B, Shao Y, Yimiti Y, et al. Spleen-preserving surgery is effective for the treatment of spleen cystic echinococcosis. Int J Infect Dis. 2014;29:181-83
2. El Malki HO, El Mejdoubi Y, Souadka A, et al. Predictive factors of deep abdominal complications after operation for hydatid cyst of the liver: 15 years of experience with 672 patients. J Am Coll Surg. 2008;206(4):629-37
3. Robert-Koch-Institut Infektionsepidemiologisches Jahrbuch meldepflichtiger Krankheiten für 2018. [cited 2019 March 1]. https://www.rki.de/DE/Content/Infekt/Jahrbuch/Jahrbuch_2018.pdf?__blob=publicationFile [in German]
4. Eckert J, Deplazes P. Biological, epidemiological, and clinical aspects of echinococcosis, a zoonosis of increasing concern. Clin Microbiol Rev. 2004;17(1):107-35
5. Akcam AT, Ulku A, Koltas IS, et al. Clinical characterization of unusual cystic echinococcosis in southern part of Turkey. Ann Saudi Med. 2014;34(6):308-16
6. Mahmoudi S, Mamishi S, Banar M, et al. Epidemiology of echinococcosis in Iran: A systematic review and meta-analysis. BMC Infect Dis. 2019;19:929
7. Zheleznovsky D, Addis MF, Biosa G, et al. Diagnostic accuracy of antigen 5-Based ELISAs for human cystic echinococcosis. PLoS Negl Trop Dis. 2016;10(3):e0004585
8. Ettorre GM, Vennarecci G, Santoro R, et al. Giant hydatid cyst of the liver with a retroperitoneal growth: A case report. J Med Case Rep. 2012;6:298
9. Franquet T, Montes M, Lecumberri FJ, et al. Hydatid disease of the spleen: Imaging findings in nine patients. Am J Roentgenol. 1990;154(3):525-28
10. Brunetti E, Kern P, Vuitton L, et al. Hydatid disease of the spleen: Imaging findings in nine patients. Am J Roentgenol. 1990;154(3):525-28
11. Ilica AT, Kocaoglu M, Zeybek N, et al. Extrahepatic abdominal hydatid disease caused by Echinococcus granulosus: Imaging findings. Am J Roentgenol. 2007;189(2):337-43
12. Manzana-Román R, Sánchez-Ovejero C, Hernández-González A, et al. Serological diagnosis and follow-up of human cystic echinococcosis: A new hope for the future? Biomed Res Int. 2015;2015:428205
13. Zhang W, Li J, McManus DP. Concepts in immunity and diagnosis of hydatid disease. Clin Microbiol Rev. 2003;16(1):18-36
14. Wen H, Vuitton L, Tuoxun T, et al. Echinococcosis: Advances in the 21st Century. Clin Microbiol Rev. 2019;32(2):e00075-18
15. Pagnozzi D, Addis MF, Biosa G, et al. Diagnostic accuracy of antigen 5-Based ELISAs for human cystic echinococcosis. PLoS Negl Trop Dis. 2016;10(3):e0004585
16. Ameur HB, Affes N, Abdelvedi C, et al. Hydatid cyst of the spleen: Tunisian series of 21 cases. Indian J Surg. 2015;77(Suppl.2):S15-19
17. Ozogul B, Kisaoglu A, Atamanalp SS, et al. Splenic hydatid cysts: 17 cases. Indian J Surg. 2015;77(Suppl.2):257-60
18. Eris C, Akbulut S, Yildiz MK, et al. Surgical approach to splenic hydatid cyst: Single center experience. Int Surg. 2013;98(4):346-53
19. Singal R, Singh SK, Mittal A, et al. A giant splenic hydatid cyst. Proc (Bayl Univ Med Cent). 2016;29(1):55-57
20. Zhuoli Z, Yu Z, Liya X, et al. Case report: laparoscopic excision of a primary giant splenic hydatid cyst: Literature review. Am J Trop Med Hyg. 2019;101(4):821-27
21. Pukar MM, Pukar SM. Giant solitary hydatid cyst of spleen – a case report. Int J Surg Case Rep. 2013;4(4):435-37
22. Velasco-Tirado V, Alonso-Sardón M, Lopez-Bernus A, et al. Medical treatment of cystic echinococcosis: systematic review and meta-analysis. BMC Infect Dis. 2018;18(1):306
23. Arif SH, Ul-Bari S, Wani NA, et al. Albendazole as an adjuvant to the standard surgical management of hydatid cyst liver. Int J Surg. 2008;6(6):448-51