Multiple Hepatic Hydatid Cysts - Review of the Literature and Case Report

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Rezumat

Chiste hidatice hepatice multiple, prezentare de caz și review al literaturii de specialitate

Introducere: Boala hidatică este o afecțiune parazitară produsă de *Echinococcus granulosus* care este des întâlnită în practica clinică. Cel mai frecvent chistele hidatice sunt localizate la nivelul ficatului, iar plămânul reprezintă cel de-al doilea organ în ceea ce privește frecvența localizării la acest nivel.

Prezentare de caz: Prezentăm cazul unei paciente în vârstă de 40 de ani care s-a prezentat în serviciul nostru cu diagnosticul de chiste hidatice pulmonare (2 chiste hidatice localizate la nivelul lobului superior și inferior stâng) și chiste hidatice hepatice multiple (10 chiste hidatice localizate la nivelul ambilor lobi hepatici. Inițial s-a intervenit chirurgical la nivel toracic, practicându-se rezecții pulmonare atipice de lob superior și inferior stâng. La 6 luni de la intervenția chirurgicală toracică s-a prezentat pentru tratamentul chirurgical al chistelor hidatice hepatice practicându-se chistectomii hepatice parțiale multiple, colecistectomie, drenaj Kehr (2 chiste hidatice hepatice prezentau fistule biliare). Evoluția postoperatorie a fost favorabilă, pacienta fiind externată în ziua a 10-a postoperator.

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Concluzii: Chiar dacă, chistul hidatic hepatic reprezintă o boală aparent benignă, există în practica clinică cazuri complexe de ehinococoză diseminată care pot necesita tratament complex. Tratamentul chirurgical rămâne în continuare cea mai bună opțiune terapeutică în aceste cazuri. La acești bolnavi se impune o urmărire postoperatorie atentă pentru depistarea cazurilor de recidivă a chistului hidatic la nivel hepatic sau extrahepatic.

Cuvinte cheie: chiste hidatice hepatice multiple, tratament chirurgical

Abstract

Introduction: Hydatid disease is a parasitic disease caused by *Echinococcus granulosus* and is commonly met in clinical practice. The most common location for hydatid cysts is the liver, while the lung is the second organ in terms of localization frequency.

Case report: We present the case of a 40-year-old patient with pulmonary hydatid cysts (two hydatid cysts located in the upper and lower pulmonary left lobes), and multiple hepatic hydatid cysts (ten cysts located in both hepatic lobes). Initially, the patient underwent thoracic surgery and was subjected to atypical lung resection of the upper and lower left pulmonary lobes. The patient underwent surgical treatment of the hepatic hydatid cysts 6 months after the thoracic surgery. The patient underwent multiple partial cystectomies, cholecystectomy, Kehr drainage with two hepatic hydatid cysts showing biliary fistulas. The postoperative evolution was favorable with patient discharge 10 days following surgery.

Conclusions: Although the hepatic hydatid cyst is a seemingly benign disease, there are complex cases of disseminated echinococcosis in clinical practice that may require complex treatment. Surgical treatment remains the best therapeutic option in these cases. Thus, for these patients, a careful postoperative follow-up is required to detect recurrence of hydatid disease.

Key words: multiple hepatic hydatid cysts, surgical treatment

Introduction

Hydatid disease is a parasitic illness caused by *Ecchinococcus granulosus*. Most often the parasite multiplies in the liver, after the absorption of its eggs from the intestine through portal circulation. If the eggs pass the liver filter and reach the systemic circulation, they can develop in any organ in the body. The lung is the second most affected organ by this disease, but hydatid cysts can be located in any organ in the human body (1). In some parts of the world, such as North America, Eastern Europe, China and the Middle East, the disease is endemic, affecting more than 50 per 100,000 people annually in certain areas (2). Untreated, hepatic hydatid

cysts (LHC) can cause multiple complications, the most common being cyst infection, cyst rupture in the bile ducts with bile fistulas, rupture of the cyst in the peritoneal cavity with peritonitis, allergic manifestations that can even cause anaphylactic shock, or the appearance of hydatid cysts disseminated in several organs. Severe cases can even lead to patient death. Therefore, at the time of diagnosis even in asymptomatic patients, the indication for treatment of these patients is absolute (3).

The purpose of this manuscript is to present the particular case of a young patient with multiple hydatid cysts (two in the lungs and 10 in the liver) as well as to conduct a review of the literature.

Case Report

We present the case of a 40-year-old female patient who, on admission, had 2 hydatid cysts located in the left lung (upper lung lobe and lower left lung lobe) as well as multiple hepatic hydatid cysts (ten cysts). Imaging studies with contrast medium revealed 2 round/ oval lesions in the left lung field, one with a solid, inhomogeneous structure located in the anterior segment of the left upper lobe, with an axial diameter of 35/28 mm AP/LL, the second lesion presenting with typical radiological appearance of detached proliferative membranes, located at the level of the posterior segment of the lower lobe with an axial plane diameter of 30/39 mm AP/ LL. The computed tomography scans pleaded for hydatid cysts stage III/ IV (*Fig. 1*).

Abdominal slices showed the presence of 10 native and post-contrast hypodense lesions at the level of the liver, diffused in the entire hepatic parenchyma, mainly in the right hepatic lobe, with dimensions between 5-37 mm (antero-posterior diameter in axial plane) pleading for hepatic hydatid cysts stage II/ III. Also, a native hypodense lesion with a hemangioma aspect was highlighted, located in the subcapsular 7th segment with dimensions of 27/24 mm (AP/ LL) in the axial plane (*Fig. 2*).

Initially, the patient underwent surgery for

the two pulmonary hydatid cysts for which atypical pulmonary resection of the upper lobe and pulmonary resection of the left lower lobe were performed. Preoperatively for 4 weeks and postoperatively for 4 weeks, the patient was treated with Albendazole. The postoperative evolution was favorable. At 6 months following pulmonary surgery, the patient returned for the treatment of the hepatic cysts. Before the second surgery, the patient underwent treatment with Albendazole for four weeks again. Surgery was performed and multiple partial hepatic cysts were removed. The intraoperative appearance confirmed the diagnosis of the imaging studies (*Fig. 3*).

Given the fact that two hepatic cysts presented with a biliary fistula, cholecystectomy and Kehr drainage of the main bile were performed. The postoperative evolution was favorable, the patient being discharged on the 10th postoperative day. The peculiarity of the present case is mainly represented by the large number of hydatid cysts in the liver (ten cysts).

Discussions

Hepatic hydatid cysts are often asymptomatic, and in the case of symptomatic LHCs (liver hydatid cysts), the most common symptom is pain in the right hypochondrium (4).

Figure 1. Thoracic Angio-CT image

Figure 2. Abdominal Angio-CT image

Figure 3. Intraoperative image

Currently, the most used imaging methods for LHC diagnosis are abdominal ultrasound and computed tomography (5). LHCs can be located both superficially or deep in the liver parenchyma.

LHCs located deep within the liver parenchyma can often be associated with biliary fistulas or may have complications due to the existence of liver abscesses (or other similar cavities in the liver parenchyma). Therefore, their surgical approach is often difficult (6).

The main therapeutic options for hepatic hydatid cysts are: antiparasitic treatment, conservative surgical treatment (aspiration of the cyst, partial cystectomy), and radical surgical treatment (total cystectomy, liver resections) (7).

Regarding antiparasitic drug treatment, the first drug used in the treatment of LHCs was mebendazole. Currently, due to the fact that it has a higher intestinal absorption, the most used is Albendazole. There are authors who claim that, administered for 4 weeks in doses of 10 mg/kg, Albendazole destroys almost all parasites, but its maximum effectiveness occurs after 3 months of uninterrupted treatment. It is also particularly useful in the preoperative preparation of these patients, being administered for 4 weeks before surgery, then, after a hiatus of 2 weeks, another 4 weeks of per os treatment (5,8). Praziquantel also has a toxic effect on the parasite, however,

it presents a disadvantage over albendazole with its lower efficacy to the germination membrane (9).

In patients with surgical contraindications, it has been noted that drug treatment is most effective in the case of small cysts (less than 4 cm) with thin walls, while in young patients it sometimes completely destroys the parasite (10). Albendazole treatment is usually well-tolerated by these patients, the main side effects being a temporary increase in liver enzymes (11).

Percutaneous treatment of LHCs is usually recommended only in patients who have contraindications to surgery with small cysts that are unilocular or in patients who refuse surgery. In these cases, the existence of any degenerative changes in the pericyst, documented with the help of abdominal CT, is a favorable prognostic factor. Usually, in these cases, antiparasitic treatment is associated (12,13). These therapeutic maneuvers are performed under ultrasound guidance that allows a precise localization of the cyst, its relationship with the surrounding liver parenchyma, and the diagnosis of potential complications. In these cases, under ultrasound guidance, the puncture, aspiration or re-aspiration of the hepatic hydatid cyst is performed. Under ultrasound guidance, hypertonic saline solution (30% NaCl) is introduced into the cyst cavity to inactivate the parasite. A volume of solution approximately 30-50% of

the cyst volume is usually introduced, the solution being left inside the cyst for 5-10 minutes, and then aspirated. A major advantage of percutaneous treatment in these cases is that it requires a shorter hospital stay, as well as a lower morbidity compared with open surgical treatment (14).

Surgical treatment remains the most effective therapeutic method in the treatment of patients with LHC. In clinical practice, both open and laparoscopic surgical approach is used. It can be done both conservatively and radically. Radical surgical treatment includes resection of the cyst together with the pericyst, while conservative treatment involves sterilization of the cyst cavity followed by evacuation of its contents, followed by partial resection of the cyst. The main purpose of surgical treatment is to inactivate the cyst, evacuate the contents of the cyst together with the germinal membrane, and treat the remaining cavity (15).

Radical surgical treatment of LHC can be performed either by cysto-pericystectomy or by liver resections. Cysto-pericystectomy is recommended for superficially located LHCs while liver resections are indicated for patients with LHCs located deep in the liver parenchyma (15). Cysto-pericystectomy can be performed either in the manner of the open cyst or by block resection of the hepatic hydatid cyst together with the pericyst. The open approach is preferred in the case of thin-walled LHCs located in the vicinity of important anatomical structures (16). Regarding the indication for liver resection in these patients, it has been shown that, at the time of diagnosis, 70% of LHC patients are not suitable for liver resection, mainly due to the widespread location of hydatid cysts in the liver parenchyma as well as due to cyst invasion of the retroperitoneal space, inferior vena cava, or hepatic hilum. A limiting factor for the practice of liver resections in these cases is done by the amount of liver volume remaining following surgery (17).

Among the endemic areas, the most used therapeutic method is the conservative surgical treatment. This involves inactivating the cyst, followed by evacuating its contents

and treating the remaining cavity. Most authors recommend inactivating the hydatid cyst with hypertonic saline. External drainage, marsupialization, internal drainage or filling of the remaining cavity with omentum can be performed for the treatment of the remaining cavity (16). Unlike radical treatment, conservative treatment has a higher postoperative morbidity, the main complications that may occur in these cases are biliary fistula, hemorrhages, liver abscesses, peritonitis, or anaphylactic shock. Conservative treatment also involves a higher recurrence rate compared with radical surgical treatment (18,19). On the other hand, radical treatment predisposes to a higher risk of intraoperative incidents and accidents, therefore it is recommended that this type of surgical approach be practiced only in centers specialized in this type of surgery (20).

A common complication of LHCs is the rupture of bile ducts. This occurs in 3-17% of cases. In the case of small fistulas with a diameter of less than 5 mm, patients are often asymptomatic, but in the case of fistulas larger than 5 mm, there is a risk of migration of daughter vesicles into the bile duct and even fragments of proliferative membrane. These patients often develop obstructive jaundice, cholangitis, or even sepsis (21,22). In these cases, in addition to the suture of the biliary fistula, if possible, it is recommended to explore the main biliary tract, perform Kher drainage, or the practice of a choledocho-duodenal anastomosis, ERCP (endoscopic retrograde cholangiopancreatography). Choledochoduodenal anastomosis is recommended especially in patients with marked dilation of the main bile duct or if the persistence of daughter blisters or prolific membrane residues in the bile duct is suspected (23).

One of the most dangerous complications that can occur in patients with LHCs is its rupture in the peritoneal cavity. This usually occurs spontaneously, post-traumatically, or following percutaneous LHC drainage. In these cases, either radical treatment or conservative treatment can be practiced. Abundant lavage of

the peritoneal cavity with hypertonic saline is also mandatory (24). The major disadvantage in these cases is a very high postoperative morbidity that can reach up to 60% as well as a postoperative mortality that can reach up to 12% of cases (25).

Both in the case of conservative treatment and in the case of radical treatment, the most common postoperative complication that may occur is biliary fistula. If the amount of bile discharged daily is less than 100 ml, these fistulas most often close spontaneously (26). If the fistula persists for more than 10 days, it is recommended to perform an endoscopic sphincterotomy followed or not by the implantation of a stent in the bile duct (27). As for the remaining cavity, the most common postoperative complication that may occur is bleeding or the formation of abscesses. In the case of liver abscesses, it is recommended to initiate antibiotic treatment and performing a percutaneous drainage under ultrasound guidance if possible. In case of failure of percutaneous drainage or in case of impossibility to perform it, the surgical approach comes into question (25).

Lately, more and more surgeons have been turning to the laparoscopic approach of LHCs. Initially, the method was not unanimously accepted by surgeons, its opponents claiming that there was a very high risk of spreading the contents of the LHCs in the peritoneal cavity (28). Recently, with the use of new LHC isolation devices from the rest of the peritoneal cavity, as well as the use of wide-angle laparoscopes, it has been shown that the risk of peritoneal dissemination in these cases is much lower than suspected, and the risk of recurrence is comparable to that of open surgery (29,30).

One of the major advantages of laparoscopic surgery is the possibility for a very thorough exploration of the inside of the cyst cavity by inserting the laparoscope inside the cyst cavity (28). On the other hand, the disadvantages of laparoscopic surgery are the technical difficulty of performing pericystic-digestive anastomoses or liver resections. This is the

reason why this type of surgical approach is used especially in high volume centers (31). In surgical centers with experience of the laparoscopic approach, the main contraindication to the laparoscopic approach is the location of the LHC in the posterior segments (especially in the 7th or 1st segment) or the existence of calcified cysts located deep in the liver parenchyma (32).

In clinical practice, there are cases that are unfortunately not suitable for surgical treatment, especially in patients with alveolar echinococcosis or patients with indication for liver transplant. At present, the indication for a liver transplant is given to patients with alveolar echinococcosis with liver failure, recurrent cholangitis, or in patients who are not suitable for liver resection and do not show extrahepatic hydatid disease (33). On the other hand, the mortality of patients undergoing a liver transplant for alveolar echinococcosis has been shown to be higher than that of patients undergoing liver transplantation for liver cirrhosis. One of the possible causes is the septic syndrome due to liver abscesses, as well as recurrent episodes of cholangitis (34). In these cases, one of the main risks for these patients is the recurrence of extrahepatic hydatid cysts due to immunosuppressive status secondary to immunosuppressive medication (35).

Conclusions

Although hepatic hydatid cyst is a seemingly benign disease, there are known complex cases of disseminated echinococcosis in clinical practice that may require complex treatment. Nevertheless, surgical treatment remains the best therapeutic option in these cases. In these patients, a careful postoperative follow-up is required to detect cases of recurrence of hydatid cyst in the hepatic or extrahepatic area.

Conflict of Interest

The authors declare no conflicts of interests.

Ethics Approval

The authors obtained the patient's consent for the publication of this case presentation.

References

1. Pakala T, Molina M, Wu GY. Hepatic Echinococcal Cysts: A Review. *J Clin Transl Hepatol* 2016;4(1):39–46.
2. Lyske J, Hutchinson C, Manolea F, Patel VH, Low G., Case 275: Multiple Hepatic Hydatid Cysts., *Radiology*. 2020;294(3):716–719.
3. Syllaios A, Schizas D, Koutiras A, Zotos PA, Davakis S, Vailas M, et al. Abdominal pain caused by a giant liver hydatid cyst and the management of the residual cavity in a tertiary public hospital in Greece. *Pan Afr Med J*. 2019;34:11.
4. Sherlock S. Hydatid disease: Disease of liver and biliary system. Oxford: Blackwell Scientific Publications; 1981. p. 247–452.
5. Shams-UI-Bari, Arif SH, Malik AA, Khaja AR, Dass TA, Naikoo ZA. Role of albendazole in the management of hydatid cyst liver, *Saudi J Gastroenterol*. 2011;17(5):343–7.
6. Kayaalp C, Bostanci B, Yol S, Akoglu M. Distribution of hydatid cysts into the liver with reference to cystobiliary communications and cavity-related complications. *Am. J. Surg*. 2003;185:175–9.
7. Julien C, Le Treut YP, Bourgouin S, Palen A, Hardwigsen J. Closed Cyst Resection for Liver Hydatid Disease: a New Standard., *J Gastrointest Surg*. 2021;25(2):436–446.
8. Chinnery JB, Morris DL. Effect of albendazole on viability of hydatid protoscolexes. *Trans R Soc Trop Med Hyg* 1986;80:815–17.
9. Horton RJ. Chemotherapy of Echinococcus infection in man with albendazole. *Trans of Roy Soc of Trop Med Hyg* 1989;83:97–102.
10. Arif SH, Shams-UI-Bari, Wani NA, Zargar SA, Wani MA, Tabassum R, et al. Albendazole as an adjuvant to the standard surgical management of hydatid cyst liver, *Int J Surg*. 2008;6(6):448–51.
11. Türkçapar AG, Ersöz S, Güngör C, Aydinuraz K, Yerdil MA, Aras N. Surgical treatment of hepatic hydatidosis combined with perioperative treatment with albendazole. *Eur J Surg*. 1997;163:923–928.
12. Gomez I Gavara C, López-Andújar R, Belda Ibáñez T, Ramia Ángel JM, Moya Herraiz Á, Orbis Castellanos F, et al. Review of the treatment of liver hydatid cysts., *World J Gastroenterol*. 2015;21(1):124–31.
13. Saidi F, Habibzadeh F. The Non-operative Management of Asymptomatic Liver Hydatids: Ending Echinococcophobia. *J Gastrointest Surg*. 2018; 22(3):486–495.
14. Akhan O, Yildiz AE, Akinci D, Yildiz BD, Ciftci T. Is the Adjuvant Albendazole Treatment Really Needed with PAIR in the Management of Liver Hydatid Cysts? A Prospective, Randomized Trial with Short-Term Follow-Up Results., *Cardiovasc Intervent Radiol*. 2014;37(6):1568–74.
15. Gupta N, Javed A, Puri S, Jain S, Singh S, Agarwal AK. Hepatic hydatid: PAIR, drain or resect?, *J Gastrointest Surg*. 2011;15(10):1829–36.
16. Sozuer E, Akyuz M, Akbulut S, Open Surgery for Hepatic Hydatid Disease, *Int Surg*. 2014;99(6):764–9.
17. Kamiyama T. Recent advances in surgical strategies for alveolar echinococcosis of the liver. *Surg Today*. 2020;50(11):1360–1367.
18. Birnbaum DJ, Hardwigsen J, Barbier L, Bouchiba N, Le Treut YP. Is hepatic resection the best treatment for hydatid cyst? *J Gastrointest Surg* 2012; 16(11):2086–2093.
19. Kucuk C, Yilmaz N, Akyildiz H, Sozuer E. Surgical treatment of liver cyst hydatid cases: analysis of 276 patients. *Erciyes Medical J* 2008;30(3): 170–174.
20. Cirenei A, Bertoldi I. Evolution of surgery for liver hydatidosis from 1950 to today: analysis of a personal experience. *World J Surg* 2001;25(1):87–92.
21. Atli M, Kama NA, Yuksek YN, Doganay M, Gozalan U, Kologlu M. Intrahepatic rupture of a hepatic hydatid cyst: associated clinical factors and proper management. *Arch Surg* 2001;136(11):1249–1255.
22. Yildirman MI, Basoglu M, Atamanalp SS, Aydinli B, Balik AA, Celebi F. Intrahepatic rupture in liver hydatid cysts: results of 20 years' experience. *Acta Chir Belg* 2003;103(6):621–625.
23. Elbir O, Gundogdu H, Caglikulekci M, Kayaalp C, Atalay F, Savkilioglu M. Surgical treatment of intrahepatic rupture of hydatid cysts of liver: comparison of choledochoduodenostomy with T-tube drainage. *Dig Surg* 2001;18(4): 289–293.
24. Akcan A, Akyildiz H, Artis T, Ozturk A, Deneme MA, Ok E. Peritoneal perforation of liver hydatid cysts: clinical presentation, predisposing factors, and surgical outcome. *World J Surg*. 2007;31(6):1284–1291.
25. Sozuer E, Ok E, Arslan M. The perforation problem in hydatid disease. *Am J Trop Med Hyg*. 2002;66(5):575–577.
26. Kayaalp C, Bzeizi K, Demirbag AE, Akoglu M. Biliary complications after hydatid liver surgery: incidence and risk factors. *J Gastrointest Surg* 2002; 6(5):706–712.
27. Akcakaya A, Sahin M, Karakelleoglu A, Okan I. Endoscopic stenting for selected cases of biliary fistula after hepatic hydatid surgery. *Surg Endosc* 2006;20(9):1415–1418.
28. Zaharie F, Bartos D, Mocan L, Zaharie R, Iancu C, Tomus C. Open or laparoscopic treatment for hydatid disease of the liver? A 10-year single-institution experience. *Surg Endosc*. 2013;27(6):2110–6.
29. Manterola C, Fernández O, Muñoz S, Vial M, Losada H, Carrasco R et al (2002) Laparoscopic pericystectomy for liver hydatid cysts. *Surg Endosc* . 2002;16:521–524.
30. Seven R, Berber E, Mercan S, Eminoglu L, Budak D. Laparoscopic treatment of hepatic hydatid disease. *Surgery*. 2002;128:36–40.
31. Boilève J B, De La Torre B, Coello PC, García Vallejo LA, Brenlla González J, Escudero Pérez B et al. Laparoscopic treatment of hepatic hydatid cysts: techniques and post-operative complications. *Cir Esp*. 2009;86:33–37.
32. Chautems R, Buhler L, Gold B, Chilcott M, Morel P, Mentha G. Long term results after complete or incomplete surgical resection of liver hydatid disease. *Swiss Med Wkly*. 2003;133:258–262.
33. Brunetti E, Kern P, Vuitton DA. Expert consensus for the diagnosis and treatment of cystic and alveolar echinococcosis in humans. *Acta Trop*. 2010;114:1–16.
34. Aydinli B, Ozturk G, Arslan S, Kantarci M, Tan O, Ahiskalioglu A, et al. Liver transplantation for alveolar echinococcosis in an endemic region. *Liver Transpl*. 2015;21:1096–102.
35. Geyer M, Wilpert J, Wiech T, Theilacker C, Stubanus M, Kramer-Zucker A, et al. Rapidly progressive hepatic alveolar echinococcosis in an ABO-incompatible renal transplant recipient. *Transpl Infect Dis*. 2011;13:278–84.