



Hepatic Hydatid Cysts. New Perspectives and Future Challenges: A Literature Review

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Abstract

Alveolar Echinococcosis (AE) persists endemically in various regions worldwide. Due to the scarce number of cases, it depicts the Achilles' heel regarding contemporary clinical and most of all surgical interventions. The aim of our study represents proper evaluation and establishment of assiduous therapeutic mapping in cases of hepatic hydatid cysts, emphasizing optimal surgical procedures and increasing optimal survival. Additionally, further extensive research will be presented, focusing on recent bibliography, using comprehensive references from databases such as PubMed, Embase, and Cochrane database, in order to depict proper surgical interventions.

Keywords: *Echinococcus granulosus*; Hepatic alveolar echinococcosis; Liver hydatid cysts; Disease-free survival

Introduction

Echinococcosis is caused by the larval tapeworm of the genus *Echinococcus*, with *E. granulosus* and *E. multilocularis* accounting for the greater number of infections worldwide reflecting as the cornerstone of cystic (CE) and Alveolar Echinococcosis (AE), respectively. This helminthic infection is transmitted to humans by direct contact with contaminated feces from animal hosts, which leads to ingestion of contaminated eggs and dissemination of the embryos into the circulation, being able to remain latent for years [1].

Geographical distribution differs between the two main types of echinococcosis. According to World Health Organization (WHO) incidence of CE accounts from <1 to 200 per 100,000 persons, while the incidence for AE is estimated from 0.03 yo to 1.2 per 100,000 people in endemic areas, respectively [2].

Thus, AE mainly affects the liver, resulting in abdominal pain, jaundice, weight loss, and other symptoms resembling a hepatic malignancy. Main locations regarding extra-hepatic disease consist of the lungs, followed by the spleen and the peritoneum [3].

A PNM system of classification for alveolar echinococcosis based on imaging findings was proposed by the World Health Organization (WHO), resembling the Tumor-Node-Metastasis (TNM) system for hepatic cancer. Thus, P represents a parasitic mass in the liver, N the involvement of neighboring organs, and M metastasis, combined into four stages, I to IV, as in the oncological model [4].

Different classification criteria for CE based on ultrasonography findings categorize active, fertile cysts as CE Type 1 and 2, cysts with compromised integrity either due to host response or chemotherapy as CE Type 3, and inactive and non-fertile cysts as CE Type 4 and 5 [5].

Discussion

Diagnosis

Laboratory diagnosis is broadly based on the use of specific antigens, which possess high diagnostic sensitivity and specificity allowing the discrimination between AE and CE, followed by antibody assays [2].

As gold standard of assiduous diagnostic mapping and post-treatment monitoring method remains ultra-sonographic scanning, with typical findings of hyper- and hypo-echogenic hepatic masses with irregular margins and calcification. Abdominal CT depicts more detailed information

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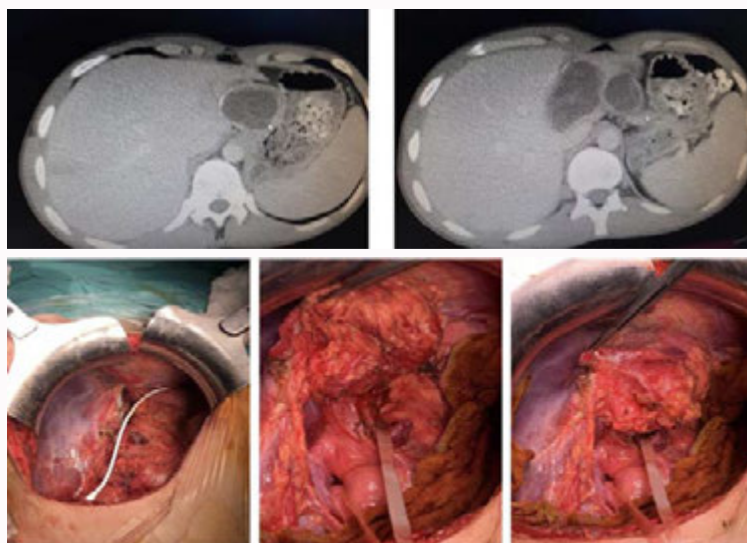


Figure 1: Patient with hepatic echinococcosis and thrombosis of the portal vein’s left ramus, who underwent left lateral hepatectomy and cholecystectomy (December 2021).

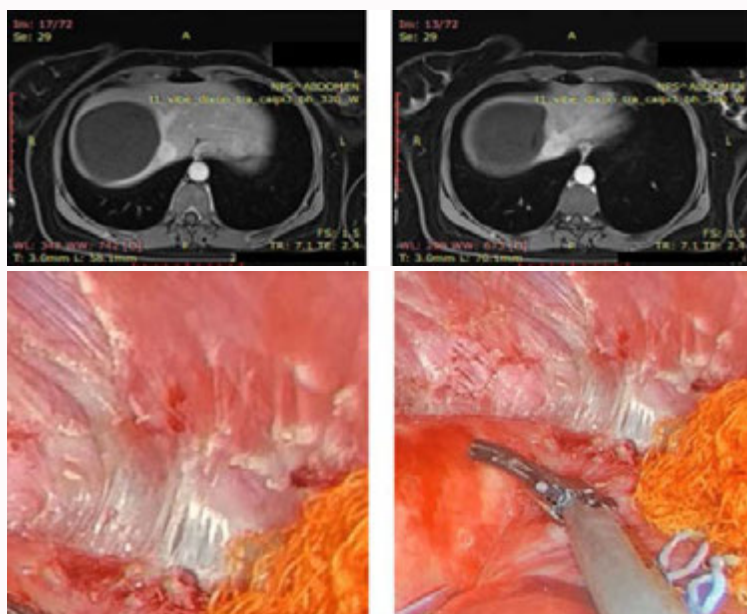


Figure 2: Laparoscopic unroofing of an extensive hepatic cyst. (August 2019).

about size, location, and depth of the cysts, as well as the hepatic parenchyma and the adjacent bile tree. Especially in cases of cerebral echinococcosis, Magnetic Resonance Imaging represents a cornerstone of therapeutic strategy. Direct X-rays possess low specificity, but could lead to infection suspicion [6].

Treatment

Timely intervention remains vital for the patient’s overall survival. Surgical procedures include per cystectomy and hepatic resection as radical measures, while unroofing, drainage, and marsupialization as conservative ones. Pre- and post-operative chemotherapy is administered [7].

The available surgical techniques can be performed either open or laparoscopically. The types of resections include a partial hepatectomy, per cystectomy, cystectomy with or without

omentoplasty, marsupialization and internal drainage. The unroofing of the hepatic simple cysts remains an acceptable option [8].

The Percutaneous Aspiration and Injection of sporicidal agents (PAIR) allows a less invasive choice. The acronym describes the four steps of the procedure: Puncture, Aspiration, Installation and Respiration. In other words, the cyst is punctured under vision and its content aspirated, the cavity is thoroughly irrigated with a sporicidal agent and repeatedly aspirated. Despite the danger of dissemination and anaphylactic shock when compared to a classical surgical approach, the PAIR technique has proven to be a safe therapeutic choice of uncomplicated hydatid cysts [9].

Chemotherapy with benzimidazoles can be effective in specific cases alone or adjunctive to the surgical resection. Two drugs are used for this purpose: Albendazole (10-15 mg/kg/day) or Mebendazole



Figure 3: Hepatic echinococcosis followed by unroofing and cholecystectomy. Intraoperative cholangiography was performed to exclude direct contact with the biliary tree. (March 2019).



Figure 4: Drainage of a hydatid cyst with the placement of a Kehr tube, due to a fistula formation with the biliary tree. (May 2018).

(40-50 mg/kg/day) [10].

The watchful waiting approach proved to be safe for naturally inactivated cysts, as the likelihood of their relapsing is low. However, some cysts inactivated through drug therapy carry the potential risk of reactivation and thus should be closely monitored with a follow-up up to 5 years [11].

Follow-up algorithm accounts for reexamination every 6 months for the first 2 years and later yearly up to ten years [12].

Complications

Disease complications can be potentially life-threatening and thus it is important to recognize and manage them promptly. Contained rupture, rupture communicating within the biliary tree or into adjacent viscera, superinfection, mass effect-related complications and anaphylactic shock are listed as the main ones [13].

Our single-center experience on surgically managing cases of hepatic echinococcosis encapsulates a plethora of the above-mentioned techniques, such as laparoscopic unroofing, intraoperative cholangiography for excluding complications, drainage through a Kehr tube due to fistulas, hepatectomy and others (Figures 1-4).

Conclusion

New treatment developments in approaching echinococcosis promise better outcomes with less complications, but are yet to be routinely used.

As with any surgical approach, laparoscopic versus open one reduces the operative time in trained surgical hands as well as the postoperative recovery time, not only allowing early mobilizations,

faster recovery and earlier hospital discharge, but also providing better aesthetical results and minimizing the risk of incisional hernia development.

A robot-assisted excision using the revolutionary Da Vinci surgical system has proven to be safe and less traumatic for the patient [14].

Alternative modified methods of the PAIR technique (Percutaneous Evacuation of Cyst content or PEVAC, indicated in patients with multivesicular cysts) have been proposed but not yet sufficiently studied for their long-term outcome and efficacy. These new techniques could set a new gold standard in the therapy of hepatic echinococcosis. We therefore encourage further research in this area.

Take-Home Message

Definitely understudied, due to the lesser amount of the affected population when compared to other clinical entities, hepatic echinococcosis still constitutes nowadays a challenge even to a skilled surgeon. Therefore, establishing clear guidelines based on contemporary evidence-based research is encouraged.

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