Hydatid Disease of the Liver Presenting as Spontaneous Cutaneous Fistula

A Case Report

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Abstract

Hydatid cyst (HD) disease is a parasitic infection produced by cysts containing the Echinococcus granulosus larval phase. Patients with HC are typically asymptomatic until incidentally diagnosed or when complications occur. A rare presentation of liver HC is spontaneous cutaneous fistulization; we report a 63-year-old female patient admitted in the hospital in 2019 (Al-Thora General Hospital, Ibb, Yemen) with an infected cutaneous fistula induced by a ruptured HC. The patient underwent laparotomy and partial cystectomy with excision of the fistula tract. The main purpose of this report is for physicians to consider this diagnosis when they face an unusual cutaneous fistula near HC common involved organs, especially in areas where the prevalence of this disease is high. We also briefly discuss the management and outcome of this disease.

Keywords: Case Report; Complications; Cutaneous Fistula; Echinococcosis; Liver; Surgery.

Introduction
Hydatid cyst (HC) is a serious zoonotic disease and a substantial health risk in endemic areas. The larval stage of Echinococcus tapeworm causes the infection. Dogs are the primary host of this worm, and sheep and cattle act as the intermediate hosts. Humans are accidental intermediate hosts in the parasite dog-sheep life. The most common primary infection site is the hepatobiliary system, and HC can remain asymptomatic in the liver for a long period of time. Most early cyst detections are caused by complications of fistula formation, which are rare manifestations. Only a few cases are reported in adults to present with spontaneous cutaneous fistula based on HC. The main purpose of this report is to consider this diagnosis by physicians when they face an unusual cutaneous fistula near the organs commonly involved in HC, especially in endemic areas.

Case Report

This report study is approved by the ethics committee of the university. A 63-year-old female patient referred to our hospital in 2019 with chronic right upper quadrant abdominal pain, started one year ago, and a recent small skin opening located in the right eighth intercostal space medial to the anterior axillary line. She also had complaints of nausea, vomiting, and fever. During history taking, the patient had no history of chronic illness; however, it was noted that the patient was living with a sheep in her house in a village. During a physical examination, the patient had a mass in her right hypochondrium, which was tender in palpation. A skin-opening was also noticed in this area (10 to 15 mm in size), with a whitish membranous structure and fluid discharge (Figure 1). Laboratory tests showed a leucocyte count of 19,000/mm³ with mild eosinophilia, an erythrocyte sedimentation rate of 100 mm/h, a hemoglobin level of 12 g/dL, total bilirubin of 2.1 mg/dL, and direct bilirubin of 1.1 mg/dL, while the other liver/kidney function tests were normal. Echinococcus latex hemagglutination test and indirect hemagglutination (IHA) test were reported was positive. Abdominal computed tomography (CT) scan revealed a 6×9-cm irregular hypodense mass in the right lobe of the liver with some calcific areas in favor of the diagnosis of hepatic HC (Figure 2).
Besides, diffuse inflammatory changes were detected around the trajectory of the fistula from the abdominal cyst toward the skin. Based on all findings, the patient was hospitalised based on "the diagnosis of" infected hepatic HC with fistualisation.

She underwent laparotomy as a part of treatment, which revealed multiple cysts in the remnants of the right hepatic lobe, inflammatory changes in the abdominal wall, and a fistula opening to the skin. Before dissecting the cyst, hypertonic saline (NaCl 20%) was used to wash the abdominal cavity and fistula tract to prevent any potential infection caused by contamination. All residual tissues involved in the right lobe of the liver and the cyst attached to the abdominal wall plus the fistula tract were excised (Figure 3). Some calcification areas were observed in the cyst wall, and the cyst consisted of heterogeneous degenerative contents along with numerous small daughter cysts spreading even in the fistula tract. Intraoperative cystic fluid examination showed protoscolices. Finally, a Fr 24 drain was inserted in the peritoneal cavity near the liver, and the abdominal wall was reconstructed. Histopathological report of the specimen revealed a laminated cyst with many scolices with a double layer of hooklets, a classic report of Echinococcus granulosus infection. The patient had a normal postoperative course in the hospital and was discharged on the 5th day on albendazole (6 mg/kg every twelve hours for four weeks) and ceftriaxone (1 gr every twelve hours for one week). A postoperative abdominal CT scan follow-up in the sixth month demonstrated the absence of recurrence. To participate in our study, written informed consent was obtained from the patient.

Discussion

HC is a common disease in agricultural communities, including the Middle Eastern countries and the Republic of Yemen. Cysts can be ruptured internally, which contaminate the biliary tract, gastrointestinal tract, or the peritoneal cavity, or externally, leading to cutaneous fistulization, which is a rare phenomenon. For cutaneous fistulization to occur, the cyst should protrude the intramuscular abdominal wall, contaminate the subcutaneous soft tissue, and make a path into the surface of the skin, forming a fistula as occurred in our patient. Clinical suspicion of such rare complications in endemic areas is essential for proper diagnosis when followed by proper radiological and serological investigations.
In humans, the symptomatic phase follows an early asymptomatic infection. Types and severity of symptoms vary based on the numbers, size, and location of the cyst and its pressure effect on the adjacent organs. For example, a non-complicated HC of the liver affects the liver locally, leading to hepatomegaly while putting pressure on the surrounding tissues to produce jaundice or abdominal pain symptoms. The overlying skin in patients with cutaneous fistulization of HC is usually normal in appearance but rarely erythematous.

The sign and symptoms of hepatic HC are not specific, and they may appear in a wide range of hepatic diseases. However, the most common symptom was the pain in the right upper quadrant (RUQ) or epigastric region, whereas the most frequent signs were hepatomegaly and a palpable mass. Additionally, radiological imaging methods may fail to show the direct communication between a hepatic cyst and the biliary system if hepatic HC is associated with a small fistula. When the serological test results, including enzyme-linked immunosorbent assay (ELISA), IHA test, and immunoelectrophoresis, are combined with imaging findings, hydatidosis can successfully be diagnosed in 90% of cases.

Cyst rupture into the abdominal cavity is an uncommon complication of HC. The amount of leaked material can introduce a wide range of allergic reactions from urticaria to life-threatening anaphylactic shock. Other complications include cyst communication with the biliary tree and secondary echinococciosis or symptoms caused by compression effects of the cyst on the biliary system.

According to Gharbi classification, our patient had a type V cyst, defined as typically inactive cysts with membrane calcification in imaging; however, on rare occasions, abscess formation within the calcified cyst cavity and occult abdominal trauma lead to cutaneous fistula, presenting with symptoms observed in our patient.

Reviewing previous research, we found one study that reported a patient with HC cutaneous fistula accompanied by rib erosion. Typically, cysts in the right lobe of the liver invade the right lateral abdominal wall, while those in the left lobe invade the anterior abdominal wall. Hematogenous metastasis is an unusual cause of subcutaneous HC, and this manifestation is
mostly caused by a primary invasion of hepatic echinococcosis or contamination during the previous surgery.¹²

Due to the rarity of HC and lack of prospective studies, there is no consensus on the gold-standard treatment modality. Treatment options include medical management, minimally invasive procedures, and invasive surgery. For minimally invasive procedures, cysts may be aspirated percutaneously, instilled, and reaspirated. In contrast, surgical options include deroofing, pericystectomy, and hepatic resection.⁵,¹³

In our case, the patient had a complicated right-lobe liver HC, which causes left-lobe hypertrophy and, by intruding the abdominal wall, leads to abdominal wall injuries and cutaneous fistula formation. For the patient, right-lobe hepatectomy was carried out, the fistula tract was excised, and the abdominal wall was reconstructed.

Radical surgical approaches include lobectomy, segmentectomy, and pericystectomy. Adjuvant therapy is not required before these procedures. Other conservative surgical approaches are considered less suitable to prevent the disease recurrence than radical approaches despite having lower morbidities.⁶ However, if conservative surgical approaches such as cystotomy and partial cystectomy are chosen in complicated cases, it is suggested that the cystic cavity should be evacuated, and the patient should receive benzimidazole treatment for 4 to 12 weeks to reduce the cyst size. However, due to the patient's low income, she could not afford it.

After resecting the skin and fistula during surgery, if fascial defects are observed in the abdominal or thoracic wall, they can be closed primarily. Even in case of larger defects, the surgeon can use synthetic graft materials.¹⁴ In case of superinfections or abscess formation, specific antibiotics should be administered after drainage.¹⁵ Unfortunately, no culture test was sent for the patient, and the patient was discharged with empirical antibiotic therapy.

Conclusion

A cutaneous fistula, as a presentation of a ruptured HC, is extremely rare. When drainage from the fistula lacks the cyst material, the clinical diagnosis would be extremely difficult. Raising
awareness around different HC complications, suggesting proper imaging modalities and serological tests are vital for accurate diagnosis of HC, especially in endemic areas.

**Authors’ contributions:**
All authors contributed to data analysis, drafting and revising the article, gave final approval of the version to be published and agree to be accountable for all aspects of the work.

**Acknowledgements:**
The authors would like to thank Shiraz University of Medical Sciences, Shiraz, Iran, and also Center for Development of Clinical Research of Nemazee Hospital and Dr. Nasrin Shokrpour for editorial assistance.

**References**


Figure 1: Whitish membranous structure protruding out from the opening of the fistula (arrow).

Figure 2: An abdominal computed tomography scan (a; axial and b; coronal view) showing calcified hydatid cyst in the right lobe of the liver (arrow).
Figure 3: The fistulous tract during excision (arrow).