

Cutaneous fistulization of the hydatid disease

A PRISMA-compliant systematic review

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Abstract

Aim: To provide an overview of the medical literature on cutaneous fistulization in patients with hydatid disease (HD).

Methods: According to PRISMA guidelines a literature search was made in PubMed, Medline, Google Scholar, and Google databases were searched using keywords to identify articles related to cutaneous fistulization of the HD. Keywords used were hydatid disease, hydatid cyst, cutaneous fistulization, cysto-cutaneous fistulization, external rupture, and external fistulization. The literature search included case reports, review articles, original articles, and meeting presentations published until July 2016 without restrictions on language, journal, or country. Articles and abstracts containing adequate information, such as age, sex, cyst size, cyst location, clinical presentation, fistula opening location, and management, were included in the study, whereas articles with insufficient clinical and demographic data were excluded. We also present a new case of cysto-cutaneous fistulization of a liver hydatid cyst.

Results: The literature review included 38 articles (32 full text, 2 abstracts, and 4 unavailable) on cutaneous fistulization in patients with HD. Among the 38 articles included in the study, 22 were written in English, 13 in French, 1 in German, 1 in Italian, and 1 in Spanish. Forty patients (21 males and 19 females; mean age \pm standard deviation, 54.0 ± 21.5 years; range, 7–93 years) were involved in the study. Twenty-four patients had cysto-cutaneous fistulization (*Echinococcus granulosus*); 10 had cutaneous fistulization (*E multilocularis*), 3 had cysto-cutaneo-bronchio-biliary fistulization, 2 had cysto-cutaneo-bronchial fistulization; and 1 had cutaneo-bronchial fistulization (*E multilocularis*). Twenty-nine patients were diagnosed with *E granulosus* and 11 had *E multilocularis* detected by clinical, radiological, and/or histopathological examinations.

Conclusion: Cutaneous fistulization is a rare complication of HD. Complicated HD should be considered in the differential diagnosis of cases presenting with cutaneous fistulization, particularly in regions where HD is endemic.

Abbreviations: CT = computed tomography, Endo US = endosonography, ERCP = endoscopic retrograde cholangiopancreatography, HC = hydatid cyst, HD = hydatid disease, IHA = indirect hemagglutination, MRI = magnetic resonance imaging, PAIR = puncture-aspiration-injection-reaspiration, PTC = percutaneous transhepatic cholangiography, US = ultrasonography.

Keywords: alveolar echinococcosis, complications, cystic echinococcosis, cysto-cutaneous fistulization, hydatid disease

1. Introduction

Hydatid disease (HD) is a zoonotic disease caused by *Echinococcus* sp. parasites (Family Taeniidae; Class Cestoda). Although 4 different *Echinococcus* sp. cause HD in humans, the

most common are *E granulosus* (causing cystic echinococcosis) and *E multilocularis* (causing alveolar echinococcosis).^[1–6] Cystic echinococcosis, also known as a hydatid cyst (HC), is responsible for 95% of all echinococcal diseases in humans. HCs can be located in almost every organ or tissue of the body, although the liver and lungs are the most commonly involved organs. Nevertheless, the majority of patients are asymptomatic and are incidentally diagnosed by radiology performed for other reasons. Complications, such as rupture, infection, anaphylaxis, and compression of adjacent organs, may cause various clinical symptoms.^[2] Among the most severe complications, rupture (perforation or fistulization) is the most important. Liver HCs can rupture into the bile duct, the gastrointestinal tract, bronchi, peritoneal cavity, or pleural cavity. The progression of liver HCs into subcutaneous tissue and fistulization in the skin is a rare complication.^[6–12] To date, few case reports on this complication have been published. In this article, we present a case of liver HC that spontaneously fistulized into an incision on the anterior abdominal wall. We also review the relevant literature.

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ZSB and UA performed surgical procedure and patient management, SA and FD designed the literature review and organized the report; SA, ZSB, and FD collected the data; AS provided radiological information; SA wrote the paper.

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or in different combinations (Flow Diagram). The language, journal, and country were not limited for this literature review. All case reports, letters to the editor, review articles, original articles, and other documents were reviewed. Reference lists of the reviewed papers were also reviewed to include citations that met the inclusion criteria. Articles without an accessible full text version, an abstract providing sufficient information, or sufficient data to compare with other studies were excluded. The following information was collected: publication year, country, publication language, paper type (full text, abstract, or unavailable), age and sex of patients, clinical presentation, *Echinococcus* sp. (*E. granulosus* or *E. alveolaris*), location of the fistula opening, cyst location, maximum size of cyst/lesion (mm), previous surgery, radiologic tools (ultrasonography [US], computed tomography [CT], magnetic resonance imaging [MRI], and fistulography), neoadjuvant antiparasitic chemotherapy, surgical management, postoperative antiparasitic chemotherapy, recurrence, and follow-up (months). The patient's age was given as mean \pm standard deviation (SD) and range. The secondary aim of this study was to document the clinical story and management algorithm for a 56-year-old male patient who was admitted to our clinic with cysto-cutaneous fistulization caused by complicated liver HC. Local ethics committee approval was not needed because this was a retrospective literature review.

3. Results

3.1. Literature review

The literature search retrieved 38 articles involving 40 patients with cutaneous fistulization/HD involvement.^[1–12,14–39] Of these, 6 articles were from India, 5 from France, 5 from Tunisia, 4 from Turkey, 4 from Morocco, 3 from Spain, 2 from Germany, 2 from Italy, and 1 each from Japan, Austria, Bulgaria, Greece, Lithuania, Romania, and Switzerland. Twenty-two articles were written in English, 13 in French, 1 in German, 1 in Italian, and 1 in Spanish. Full text was obtained for 32 of the 38 articles, whereas abstracts were available for 2 articles, and no text of any kind was available for 4 articles. The study details of the unavailable articles were obtained from 4 different articles.^[5,6,14,15] Therefore, the current analysis included 40 patients (21 [52.5%] males and 19 [47.5%] females; ages, 7–93 years; mean \pm SD, 54.0 \pm 21.5 years). The age range of males was 29 to 87 years (mean \pm SD, 57.5 \pm 16.9 years) and that of females was 7 to 93 years (mean \pm SD, 50.1 \pm 25.0 years). The male:female ratio was 1:1. The external opening area of the fistula was reported for all patients, and the fistula opening was localized to the right hypochondrium in 11, the right flank in 4, the right thoracic wall in 4, the left hypochondrium in 3, the sternal area in 2, the inguinal region in 2, the periumbilical region in 2, the left flank in 2, and the left thoracic wall in 1 patient. Twenty-four (60%) patients had cysto-cutaneous fistulization (*E. granulosus*); 10 (25%) had cutaneous fistulization (*E. multilocularis*), 3 had cysto-cutaneo-bronchio-biliary fistulization, 2 had cysto-cutaneo-bronchial fistulization; and 1 had cutaneo-bronchial fistulization (*E. multilocularis*). While 29 (72.5%) patients were diagnosed with *E. granulosus*, 11 (27.5%) patients had *E. multilocularis* that was detected by clinical, radiological, and/or histopathological examinations. The detailed clinical and demographic characteristics of the 40 patients with cutaneous fistulization/involvement of HD are summarized in Table 1.

3.2. Case report

A 56-year-old male patient was referred to our clinic for nausea, vomiting, and a swelling on the anterior abdominal wall. He had

undergone surgery for a perforated peptic ulcer 15 years ago. A physical examination revealed hyperemia around the abdominal wall incision and a fascial defect at the abdominal midline. Abdominal US showed that a few of the lesions were consistent with HCs in both lobes of the liver and the spleen. The patient was examined with a panendoscope to differentially diagnose the dyspeptic complaints. The endoscopic examination revealed external compression of the gastric wall. The echinococcal indirect hemagglutination test was positive at a titer of 1:256. A contrast-enhanced abdominal CT scan showed 2 calcifying lesions consistent with HCs, the largest of which had a diameter of 5 cm, in the anterior and posterior segments of the right lobe of the liver. The other HC lesion was 11 \times 10 cm and almost completely filled the left lobe of the liver. This HC was compressing the stomach, and was close to the anterior abdominal wall. Additionally, 2 more lesions resembling HCs, of which the larger had a 10-cm diameter, were detected in the spleen (Figs. 1 and 2). Surgery was scheduled, so the patient was administered two 400-mg doses of albendazole p.o. and a pneumococcal vaccine. Five days after the initial examination the patient presented to our emergency department with marked redness and discharge from the abdominal wall. A physical examination revealed a clear fluid discharge from a 1.5-cm opening. After local anesthesia, the orifice was dilated and daughter vesicles and a large volume of HC fluid were drained (Fig. 3). The patient underwent surgery the next day. The cyst that almost completely filled the left lobe of the liver and fistulized to the epigastrium was excised completely. Then, the cyst in the right lobe was drained with a partial cystectomy + omentopexy, and a splenectomy was performed. The fascia was closed, the incision was debrided, and the wound was allowed to heal. Albendazole was started 3 days after surgery; the patient received 4 cycles. No recurrences were detected at the 6-month follow-up visit.

4. Discussion

HD is a serious public health problem that is prevalent in many regions around the world where live animal husbandry is the primary source of living. Turkey is geographically part of the Middle East and Mediterranean regions and is one of several countries where HC disease is endemic. Humans have no role in the biological cycle of the causative organism and are accidentally infected after ingesting *Echinococcus* eggs in dog feces. Larvae are released into the gastrointestinal tract after the ingested eggs rupture, penetrate the intestinal wall, and pass into the portal system to reside in the hepatic sinusoids. Small-sized larvae pass through the hepatic filtration system and reach the lungs. There, the majority of larvae are cleared by a second capillary filtration system. Those that go unfiltered can easily reach many organs and systems of the body. Considering this dissemination route, it is quite easy to understand why it primarily involves the liver (50–77%) and lungs (10–40%).^[11,6]

Liver HD can remain asymptomatic for years. Most asymptomatic patients are incidentally diagnosed by radiological tests performed for other indications.^[6] Sometimes, HD can increase the size of the liver and patients are diagnosed by a physical examination in which a palpable liver is detected. Clinical symptoms can vary greatly depending on the size, number, and localization of HCs, their anatomical relationship with adjacent organs, and compression of adjacent organs.^[36]

The most common complications of HD include secondary infections (super-infected HCs), anaphylactic reactions, vascular

Table 1

Demographic and clinical characteristics of 38 articles involving 40 patients published in medical literature between 1987 and 2016.

References	Year	Country	Language	Paper	Age	Sex	Clinical presentation/ definitive diagnosis	Agent	Fistula opening/ lesion location	Cyst locations	Cyst size (mm)	Previous surgery
Virgilio	2015	Italy	English	Full text	55	F	Cysto-cutaneous-bronchial fistula	<i>E. granulosus</i>	Right hypochochondrium	Right lobe-liver	100	Hydatid surgery
Akey	2015	Turkey	English	Full text	80	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Right hypochochondrium	Right lobe-liver	60	Liver abscess?
Daidoul	2015	Tunisia	French	Full text	23	F	Cysto-cutaneous fistula	<i>E. granulosus</i>	Right flank	Right lobe-liver	50	NA
Mandolkar	2015	India	English	Full text	25	F	Cysto-cutaneous fistula	<i>E. granulosus</i>	Left hypochochondrium	Left lobe-liver	110	No
Juodelkis	2014	Lithuania	English	Full text	29	M	Cutaneous fistula	<i>E. multilocularis</i>	Right flank	Right lobe-liver	3	Abscess drainage
Sachdeva	2014	India	English	Full text	55	M	Cysto-cutaneo-bronchio-biliary fistula	<i>E. granulosus</i>	Supra umbilical	Right lobe-liver	NA	Hydatid surgery
Jayant	2014	India	English	Full text	65	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Right thoracic wall	Right lobe-liver	NA	No
Kjossev	2013	Bulgaria	English	Full text	60	F	Cysto-cutaneous fistula	<i>E. granulosus</i>	Left hypochochondrium	Left lobe-liver	50	Rib osteomyelitis
Gupta	2013	India	English	Full text	50	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Left flank	Liver- bilobar	180	No
Bouassida	2012	Tunisia	French	Full text	24	F	Cysto-cutaneous fistula	<i>E. granulosus</i>	Right hypochochondrium	Inaperitoneal	NA	Hydatid surgery
Hamid	2012	India	English	Full text	7	F	Cysto-cutaneous fistula	<i>E. granulosus</i>	8th intercostal space	Right lobe-liver	50	Hydatid surgery
Lacetera	2012	Italy	Italian	Full text	60	F	Cysto-cutaneous fistula	<i>E. granulosus</i>	Left flank	Kidney-left	105	No
De Levaissiere	2012	France	French	Full text	40	F	Cysto-cutaneous fistula	<i>E. granulosus</i>	Inguinal, left	Pelvis	70	No
Ben Ameur	2012	Tunisia	French	Full text	33	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Right flank	Right lobe-liver	50	No
Prieto-Nieto	2011	Spain	English	Full text	58	F	Cysto-Cutaneo-Bronchio-Biliary fistula	<i>E. granulosus</i>	Right hypochochondrium	Right lobe-liver	NA	Hydatid surgery
Ramia Angel	2011	Spain	Spanish	Full text	76	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Left hypochochondrium	Inaperitoneal	80	Hydatid surgery
Konwar	2011	India	English	Full text	68	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Umbilicus	Right lobe-liver	120	NA
Martin-Perez	2011	Spain	English	Full text	87	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Right hypochochondrium	Right lobe-liver	180	No
Schmoldt	2010	Germany	English	Full text	71	F	cutaneous fistula	<i>E. multilocularis</i>	Epigastrium	Left lobe-liver	75	No
Yakan	2009	Turkey	English	Full text	93	F	Cysto-cutaneous fistula	<i>E. granulosus</i>	Infra umbilical	Left lobe-liver	90	Cholecystectomy
Ali	2009	Morocco	French	Full text	45	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Right hypochochondrium	Right lobe-liver	NA	NA
Chafik	2009	Morocco	English	Full text	35	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Left thoracic wall	6th left rib	NA	Yes
Onat	2008	Turkey	English	Full text	21	F	Cysto-cutaneous fistula	<i>E. granulosus</i>	Right thoracic wall	Right lobe-liver+pulmonary	NA	Hydatid surgery
Florea	2008	Romania	English	Full text	71	F	Cysto-cutaneous fistula	<i>E. granulosus</i>	Right hypochochondrium	Right lobe-liver	60	No
El Ammari	2008	Morocco	French	Full text	64	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Inguinal, right	Kidney-right	340	No
Sakorafas	2006	Greece	English	Full text	85	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Right hypochochondrium	Right lobe-liver	NA	Cholecystectomy
Kismet	2006	Turkey	English	Full text	43	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Epigastrium	Spleen	95	No
Bastid	2005	France	French	Full text	42	M	Cysto-cutaneous fistula	<i>E. granulosus</i>	Epigastrium	Liver	140	Hydatid surgery
Grígy-Guilla	2004	France	French	Full text	39	F	Cysto-cutaneous fistula	<i>E. granulosus</i>	Epigastrium	Right lobe-liver	40	No
Reuter	2003	Germany	English	Full text	60	M	cutaneous fistula	<i>E. multilocularis</i>	Sternal area	Liver, thoracic wall	NA	Hydatid surgery
Seimi	2001	Tunisia	French	NA	67	F	Cutaneous fistula	<i>E. multilocularis</i>	Sternal area	Liver, heart, thorax	40	Hydatid surgery
Ambo	1999	Japan	English	Full text	68	M	Cutaneous fistula	<i>E. granulosus</i>	Right thoracic wall	Right lobe-liver	NA	Hydatid surgery
Harandou	1997	Morocco	French	Abstract	70	F	Cysto-cutaneo-bronchial fistula	<i>E. multilocularis</i>	Right hypochochondrium	Liver	NA	Hydatid surgery
Bresson-Hadni	1996	Austria	English	Full text	68	M	Cutaneous fistula	<i>E. granulosus</i>	Right Flank	Liver	NA	NA
Berthet	1992	France	French	NA	68	M	Cutaneous fistula	<i>E. multilocularis</i>	Epigastrium	Liver	NA	No
Golematis	1991	France	French	NA	41	M	Cutaneous fistula	<i>E. multilocularis</i>	Epigastrium	Left lobe-liver	NA	Abscess drainage
Tschudi	1988	Switzerland	German	Abstract	71	F	Cutaneous fistula	<i>E. multilocularis</i>	Right hypochochondrium	Right lobe-liver	NA	NA
Kehila	1987	Tunisia	French	NA	47	M	Cutaneous fistula	<i>E. multilocularis</i>	Right thoracic wall	Liver	NA	Hydatid surgery
					86	F	Cutaneo-bronchial fistula	<i>E. multilocularis</i>	Right flank	Right lobe-liver	NA	No

Table 1 Continued

References	Radiologic tools	Neoadjuvant treatment	Surgical management	Adjuvant treatment	Recurrence	Follow-up (mo)
Virgilio	CT + Fistulography	No	Nonoperative management	Yes-3mo	No	NA
Akay	US + CT	NA	Drainage	NA	No	6
Daidoul	US + CT	No	Excision of fistulous Tract + pericystectomy	No	No	132
Mandolkar	US + X-Ray	No	Excision of fistulous Tract + pericystectomy	ABZ -2mo	No	4
Juodeikis	US + CT	ABZ	Excision of fistulous Tract + segmentectomy (6 + 7)	ABZ -12 mo	No	12
Sachdeva	CT + Fistulography	NA	Thoracotomy + excision of cyst cavity + diaphragm repair + tube drainage	NA	NA	NA
Jayant	US + CT	ABZ	Right hepatectomy + excision of fistulous tract	ABZ	No	12
Kjossev	CT	NA	Pericystectomy + excision of fistulous tract	ABZ	No	12
Gupta	US + CT	NA	Evacuation of cyst Cavity + thoracotomy + diaphragmatic plication	NA	No	Died
Bouassida	CT	NA	Total cystectomy + excision of fistulous tract	ABZ-6 mo	No	60
Hamid	US + CT + Fistulography	NA	Excision of fistulous Tract + evacuation of cyst cavity	ABZ	No	5
Lacetera	CT	NA	Excision of fistulous tract + left nephrectomy	ABZ -6 mo	No	24
De Levaissiere	US + CT	ABZ	Nonoperative management	ABZ -3 mo	No	5
Ben Aneur	US + CT	ABZ	Excision of fistulous tract + partial cystectomy	ABZ -6 mo	No	18
Prieto-Nieto	CT + Fistulography	NA	Tissuol sealing injection in fistulous tractus	No	No	24
Ramia Angel	CT	NA	Excision of fistulous tract + total cystectomy (peritoneal and subcutaneous)	ABZ-3 mo	No	NA
Konwar	US	ABZ	Partial cystectomy + excision of fistulous tract + total cystectomy (peritoneum)	ABZ-2mo	No	60
Martin-Perez	US + CT	ABZ	Excision of fistulous tract + total cystectomy (subcutaneous cyst) + partial cystectomy (liver)	NA	No	NS
Schmoldt	CT	MBZ + ABZ	Excision of fistulous tract + left hepatectomy + partial pericardial + diaphragmatic Resection	ABZ	No	12
Yakan	CT	ABZ	Nonoperative management	ABZ	NA	NA
Ali	CT	NA	Excision of fistulous tract + partial cystectomy	ABZ	NA	NA
Chafik	CT	NA	Resection of the lateral arch of the 6th rib	ABZ-3mo	No	18
Onat	CT	NA	Cystotomy + capitonage	ABZ -6mo	No	18
Florea	US + CT	NA	Excision of fistulous tract + partial cystectomy	NA	No	3
El Ammari	CT	NA	Pericystectomy	ABZ + Praziquantel- 6 mo	No	36
Sakoratas	CT	NA	Partial cystectomy + subcutaneous cyst excision	NA	NA	NA
Kismet	US	NA	Partial cystectomy	NA	No	12
Bastid	US + CT	NA	Percutaneous drainage	ABZ	No	3
Grig-Guilla	US + CT + MR	ABZ	Excision of fistulous tract + partial cystectomy	ABZ	Regressed	3
Reuter	CT + MRI + FDG PET-CT	Amphotersin B	Nonoperative management	ABZ-1mo	No	NA
Reuter	CT + MRI + FDG PET-CT	Amphotersin B	Nonoperative management	NA	Successfully	14
Selmi	US	NA	Cystectomy	NA	Decreased	25
Ambo	CT	NA	Excision	NA	No	9
Harandou	NA	NA	Excision of fistulous tract + thoracotomy + pericystectomy	NA	No	36
Bresson-Hadhi	X-Ray	ABZ	Nonoperative management	NA	NA	NA
Bresson-Hadhi	CT + Fistulography	ABZ	Nonoperative management	ABZ	No	2
Berthet	US	NA	Partial cystectomy	ABZ	Regressed	12
Golematis	CT + Fistulography	NA	Partial cystectomy + drainage	NA	No	84
Tschudi	CT	NA	Wide lokal excision	NA	No	24
Kehilla	US + Fistulography	NA	External catheter drainage	Antibiotherapy	No	48

ABZ = albendazole, CT = computed tomography, *E. granulosus* = *Echinococcus granulosus*, *E. multilocularis* = *Echinococcus multilocularis*, F = female, M = male, MBZ = mebendazole, MRI = magnetic resonance imaging, NA = non-available, US = ultrasonography.

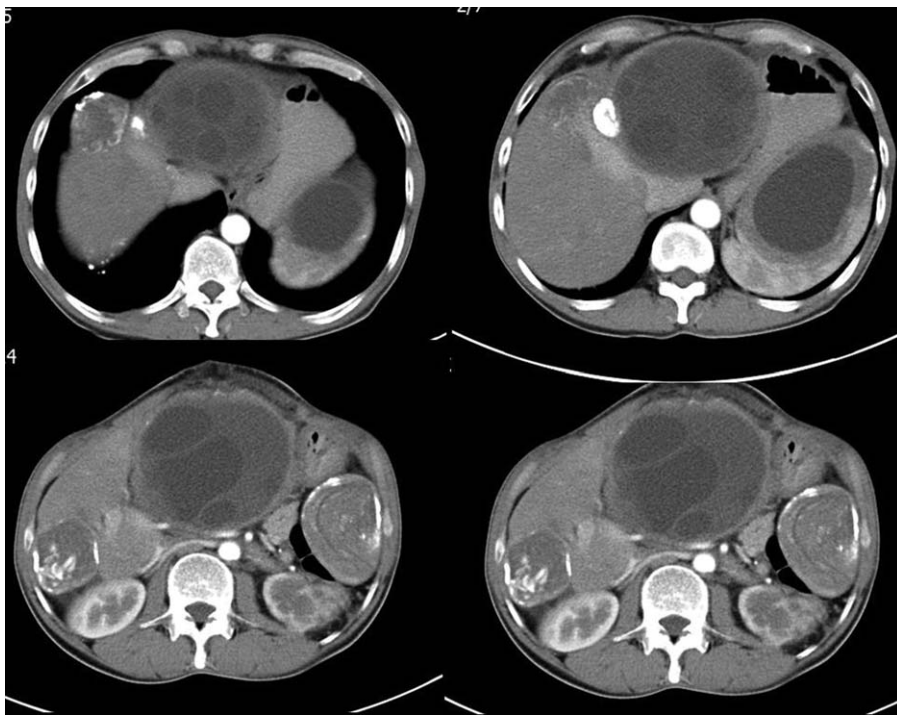


Figure 1. Axial abdominal computed tomography images obtained from different sections after injecting contrast material. A lesion consistent with a stage III hydatid cyst is seen in the posterior segment of the right liver lobe and another lesion consistent with a stage V hydatid cyst is seen in the anterior segment of the right liver lobe. Lesions consistent with stage I and stage V hydatid cysts are seen in the upper and lower poles of the spleen, respectively.

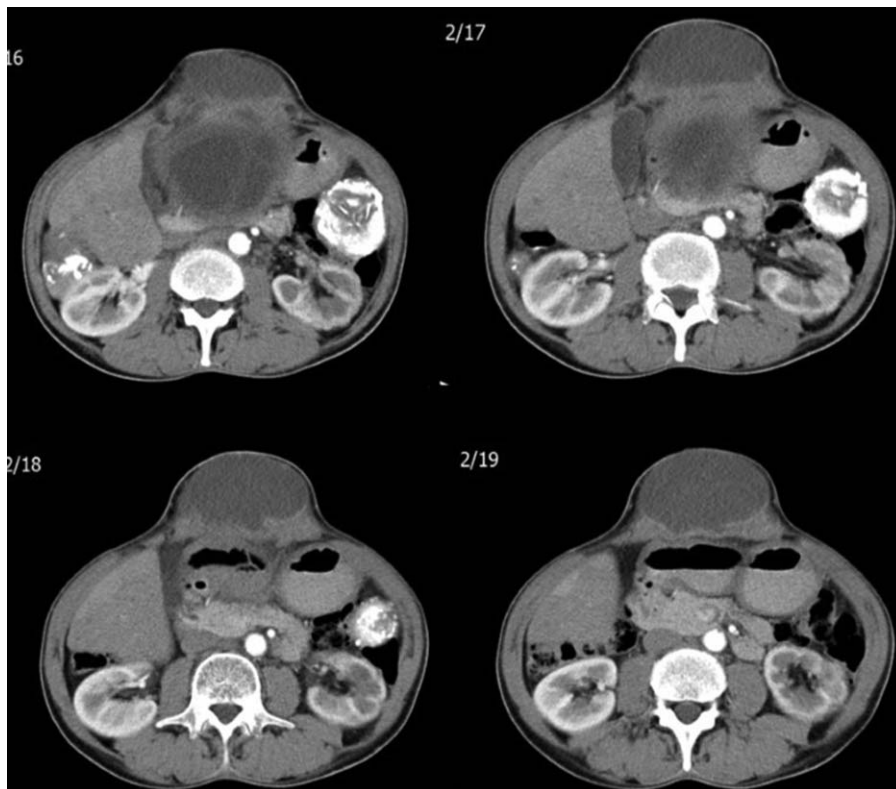


Figure 2. Sequential abdominal computed tomography images taken after injecting contrast material. A lesion consistent with a hydatid cyst is seen starting from segment 3 of the left liver lobe and protruding into the anterior abdominal wall.



Figure 3. Physical examination shows a clear fluid discharge and daughter vesicles originating from an opening in the abdominal wall.

compression (Budd–Chiari syndrome, portal hypertension, or portal vein thrombosis), biliary compression (obstructive jaundice or cholangitis), compression of adjacent structures (gastric outlet obstruction or duodenal compression), and internal or external rupture.^[6,12] Internal rupture, also known as an internal perforation, is the most serious complication of HD. Rupture of HCs into the biliary tree, gastrointestinal tract, bronchi, pleural space, or peritoneal cavity is referred to as an internal rupture.^[4] On the other hand, rupture into the external body surface is an external rupture, external perforation, or cutaneous fistulization. Cutaneous fistulization is a rare complication of HD involving many organs, including the liver.

HD follows several stages before reaching the abdominal or thoracic wall to develop into an external rupture. Stage I hydatid lesions protrude into the innermost muscular layer of the abdominal or thoracic wall. Stage II lesions pass beyond the muscular layer and protrude into subcutaneous soft tissue. Stage III is characterized by the passage of lesions into subcutaneous tissue and their fistulization in the skin, which is called an external rupture, external fistulization, or cutaneous fistula.

Clinical suspicion is the first and most important step in the diagnosis and management of complicated HD. Living in an endemic region and a history of HD are important clues for the diagnosis. Discharge of hydatid fluid or daughter vesicles from the external orifice of a fistula is a useful clinical sign in patients presenting with cutaneous fistulization. When clinical signs are used in combination with radiological tools and serological tests, the diagnosis can be easily made in almost all patients. A histopathological examination of the fluid discharged from the external orifice of a fistula can demonstrate the parasite itself and identify a superinfection, if present. US, CT, and MRI are the most commonly utilized radiological tools to diagnose cutaneous involvement and locate lesions causing complications of the HD. Endoscopic retrograde cholangiopancreatography (ERCP), percutaneous transhepatic cholangiography (PTC), and endosonography are the other invasive radiological methods used for diagnostic and therapeutic purposes. The most useful radiological method for cutaneous manifestations of the disease is contrast-enhanced fistulography. This technique helps clinicians specify the extension of the fistula, the location and size of a fistulized lesion, and its relationship with bile ducts, bronchopleural structures, and the pelviciceal system. In the

present study, 7 (17.5%) of the enrolled patients underwent fistulography in addition to other radiological examinations. Bresson-Hadni et al^[30] reported that fistulography failed to show any connection between the fistula tract and the liver lesion. Prieto-Nieto et al^[37] showed a connection between the fistula tract and bronchobiliary structures by fistulography in a patient who had undergone HC surgery years ago. Prieto-Nieto et al^[37] reported that they filled the fistula tract with Tissucol, a biological fibrin glue, and successfully treated the patient. In conclusion, the success rate of fistulography for showing organ involvement of a cystocutaneous fistula is 85.7%. In the present case, we did not use fistulography due to discharge of boluses of daughter vesicles through the incision scar. However, fistulography can be performed under appropriate conditions in all cases with a narrow fistula orifice and an unclear diagnosis. We cannot provide a clear recommendation due to a lack of sufficient data on this subject.

The most commonly used serological tests for diagnosing complicated and noncomplicated HD and monitoring posttreatment recurrences are enzyme-linked immunosorbent assay, indirect hemagglutination, serum immunoelectrophoresis, the complement fixation test, and immunofluorescence assay. Nineteen of 40 enrolled patients underwent various serological tests; 94.7% had a positive test result. Only 1 patient returned negative serology, although costal HD was shown histopathologically.

The most appropriate strategy to minimize recurrence in patients with complicated HD and cutaneous fistulization is to combine surgical and medical treatment modalities. A 2 to 4-week neoadjuvant medical treatment followed by elective surgery is the most appropriate approach for patients who are not in need of urgent surgical intervention. Surgical treatment includes en block resection of the primary hydatid lesions causing the complication, diseased skin region, and fistula tract. Radical options, such as pericystectomy, segmentectomy, and lobectomy, can be used to minimize recurrence. Adjuvant medical treatment is not necessary in patients treated with radical surgery. On the other hand, all daughter vesicles should be removed with the germinative membrane in patients treated conservatively, such as partial cystectomy or cystotomy, and medical treatment should be administered for 4 to 12 weeks after the surgery. Fascial defects formed on the abdominal or thoracic wall after resecting the skin, and the fistula tract should be closed primarily or using artificial graft material. Patients diagnosed with a superinfection or abscess should be treated with drainage and antibiotic therapy specific to the bacteria that grow in culture. The situation is somewhat more complicated in skin fistulae associated with an *E. alveolaris* infection in which multiple cutaneous fistula orifices and multiple hepatic/thoracic lesions are present. These lesions should be resected with a 1-cm clean surgical border as if a tumor were present.^[3,17] Regardless of the surgical method used, long-term medical treatment should be administered for *E. alveolaris* after surgical resection.^[3] In the present short literature review, adjuvant medical treatment was administered for 1 to 12 months in 60% of cases with cutaneous involvement. Four of the treated patients were diagnosed with an extensive *E. alveolaris* infection and received long-term medical treatment without any surgical treatment. Reuter et al^[27] reported favorable results administering amphotericin B in 2 cases with hepatic toxicity to benzimidazole. Four studies have reported using amphotericin B to treat *E. alveolaris* and 3 of these were reported by Reuter et al.^[27] The authors suggested using amphotericin B in cases intolerant or resistant to benzimidazole. In conclusion, we cannot make any solid recommendations for managing HD with

cutaneous fistulization, as a limited number of such cases have been reported so far. This topic should be explored further in future studies.

4.1. Topic highlights

- Cutaneous fistulization is a rare but serious complication of hydatid disease.
- A preoperative diagnosis is quite challenging when no cyst material has been drained from the external os of a fistula tract.
- Awareness is an important approach to hydatid disease and its complications.
- Complicated hydatid disease should be considered in the differential diagnoses of patients presenting with cutaneous fistulization, particularly in regions where hydatid disease is endemic.

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