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Case Report

Hydatid Disease Presenting as Cutaneous Fistula: Review of a Rare Clinical Presentation

Vijay Korwar, Gokulakkrishna Subhas, Prabhuraj Gaddikeri, Shivaswamy B. S.

Department of General Surgery, Victoria Hospital, Bangalore, Karnataka, India

Hydatid disease has a worldwide distribution as a result of more global travel. Liver and lungs are the most common sites for the primary hydatid cysts in the human body. We managed a 68-year-old man who presented with abdominal distension and umbilical fistula, discharging daughter cysts. Ultrasound imaging of the abdomen showed hepatic hydatid cyst forming a fistula at the umbilicus. The patient underwent a laparotomy with partial cystectomy and excision of the fistula tract. The umbilicus healed after the procedure. The patient did not have any recurrences in 5 years of follow-up. Spontaneous cutaneous fistulization of liver hydatid cyst is a rare presentation. A detailed literature search revealed 15 cases published in all languages. Hydatid disease presenting as an umbilical fistula has not been reported yet. We summarize all these cases including the presentation, findings, management, and outcome. Combined surgical and medical treatment is successful in healing of hepatic hydatid cutaneous fistula.

Key words: Hydatid disease – Cutaneous fistula – Complications – Umbilical fistula – Cystectomy – Albendazole

Hydatid disease is a parasitic infestation caused by the larval stage of genus *Echinococcus*. Liver is the most frequently involved organ, followed by the lungs.[1] Cutaneous involvement in hydatid disease is very rare. A Medline search in all languages using the keywords hydatid, cutaneous, and fistula revealed only 15 relevant cases.[2–16] We managed a case of spontaneous fistulization of a hydatid cyst through the umbilicus.

Case Report

A 68-year-old male agricultural worker presented with abdominal distension and pain for 4 months and discharge of white grape skin-like membranes from the umbilicus for 1 week (Fig. 1). The abdomen was distended with hepatomegaly, and a few masses could be felt in the upper abdomen. There was a fistula opening at the bottom of the umbilicus

Reprint requests: Vijay A. Korwar, MS, MRCS, Department of General Surgery, Victoria Hospital, Fort Road, Bangalore 560002, India. Tel. and Fax: +91 80 26703294; E-mail: vkorwar@gmail.com

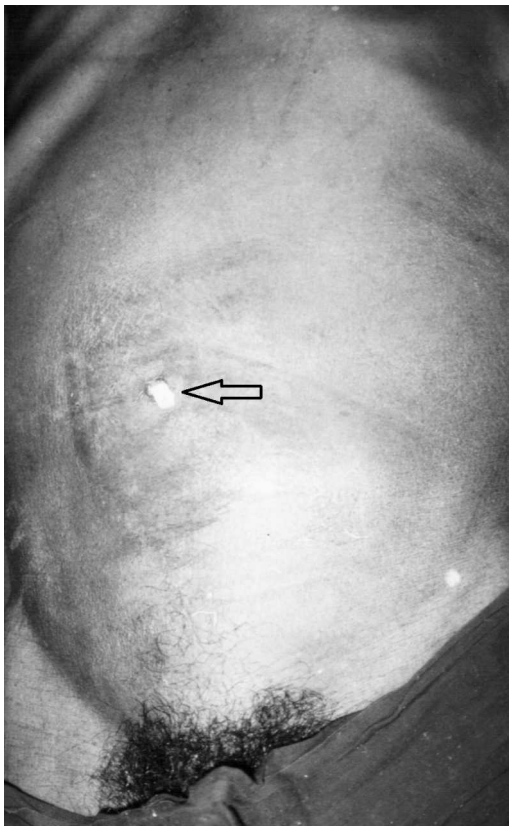

Fig. 1 Arrow showing daughter cyst extruding from umbilicus.

discharging clear fluid and white grape skin-like membranes (daughter cysts). Ultrasound examination showed hydatid cysts in segments IV, V, and VIII of the liver and peritoneum. The cyst occupying segments V and VIII was forming a fistula opening at the umbilicus.

A preoperative course of albendazole, 10 mg/kg/d for 7 days, was given before the procedure. A midline laparotomy was done under general anesthesia. After opening the abdomen, two large hydatid cysts were found in the liver and three smaller hydatid cysts were seen in the peritoneum. The largest cyst measured 12 cm in segments V and VIII. This cyst in the right lobe of the liver was forming a cutaneous fistula at the umbilicus. Liver hydatid cysts were aspirated using a 20-G needle to reduce the intracystic pressure and the cysts were injected with hypertonic (20%) saline (scolicidal). The cyst was opened with a small incision and a suction tip was introduced to evacuate the remaining fluid. Germinative membrane was easily removed after widening the incision and visible biliary openings were sutured. Peritoneal cysts were carefully removed intact and the omentum was placed in the liver in the residual cavity. The fistula tract was excised and the umbilicus was preserved. A drain was inserted in the peritoneum near the residual cavity in the liver.

Postoperatively, the patient was given 2 additional cycles of albendazole, 10 mg/kg/d for 28 days, with an interval of 2 weeks. The laparotomy wound and the umbilical fistula site healed well after the surgery. Follow-up ultrasound examination at 5 years did not reveal recurrence of the disease.

We performed a literature search in Medline using the terms hydatid disease and cutaneous fistula, which revealed 15 cases. A summary of these cases is presented in Table 1.

Literature Review

On review of the literature for cutaneous involvement in hydatid disease (Table 1), we found 7 publications in English, 6 in French, 1 in German, and 1 in Russian. The age of the patients with hydatid cyst–cutaneous fistula ranged from 11 to 86 years (mean age, 58 years) with a male-to-female ratio of 1.7:2. The primary location of the hydatid cyst was right lobe of the liver, except in 3 patients where the primary location was the spleen,[4] left lobe of the liver,[11] and skin.[1] Two of the patients had cutaneous involvement 8 years[11] and 13 years[10] after having undergone surgical treatment for hydatid disease of the liver. The majority of the patients presented with mass and draining fistulas. Skin on the right side of the torso was the most common site involved. The size of the cyst at the primary location varied from 3 to 14 cm. The diagnosis was confirmed on imaging using ultrasound and computed tomography scans in all the patients expect in the patient with primary skin involvement,[14] where the diagnosis was confirmed by microbiological examination. All 16 patients were treated by surgery and partial cystectomy was the most commonly performed procedure. Follow-up ranged from 3 months to 7 years. One patient died 2 months after the excision of skin lesion[12] and there was no recurrence reported in any of the remaining patients.

Discussion

Hydatid disease is seen worldwide due to increasing migration and growing global travel. Hydatid disease is a parasitic infestation caused by the larval stage of the genus *Echinococcus*. The most common form, cystic hydatid disease, is caused by *Echinococcus granulosus*, whereas the alveolar type is caused

Table 1 Details of cases from review of literature for cutaneous involvement in hydatid disease

Ref. no	Author	Year	Age	Sex	Country (Language)	Fistula opening	Site (hydatid cyst)	Investigations	Treatment	Follow-up, outcome
1	Current case	2009	68	M	India (English)	Umbilicus	Liver Rt. lobe (V, VIII)	USG	Cystectomy	5 y, NR
2	Florea *et al*	2008	71	F	Romania (English)	Rt. hypochondrium	Rt. lobe	USG, CT	Partial cystectomy	3 y, NR
3	Sakorafas *et al*	2006	85	M	Greece (English)	Rt. hypochondrium	Liver	CT	Partial cystectomy	1 y, NR
4	Kismet *et al*	2006	43	M	Turkey (English)	Periumbilical	Spleen	USG	Partial cystectomy	3 mo, NR
5	Bedioui *et al*	2006	41	F	Tunisia (French)	Epigastric	Liver Rt. lobe	CT , USG	Partial cystectomy	4 mo, NR
6	Salerno *et al*	2006	57	F	Italy (English)	Rt. flank (9th rib)	Liver Rt. lobe (V–VII)	CT	Marsupilization	3 mo, NR
7	Grigy-Guillaumot *et al*	2004	39	F	France (French)	Epigastric	Liver Rt. lobe (IV)	USG	Partial cystectomy	1 mo, NR
8	Selmi *et al*	2001	11	F	Tunisia (French)	Rt. thoracic posterolateral	Liver Rt. lobe (VII)	USG	Cystectomy	9 mo, NR
9	Parmar *et al*	2001	67	F	India (English)	Rt. hypochondrium	Liver Rt. lobe	CT	Cystectomy	Not specified
10	Ambo *et al*	1999	58	M	Japan (English)	Rt. hypochondrium	Liver Rt. lobe	CT	Excision of subcut. lump	3 y, NR
11	Bresson-Hadni *et al*	1996	68	M	Austria (English)	Epigastric	Liver	USG	Excision of skin lesion	Died in 2 mo
12	Berthet *et al*	1992	41	M	France (French)	Rt. anterolateral abdomen	Liver (VII–VIII)	USG	Partial cystectomy	7 y, NR
13	Golematis *et al*	1991	71	F	France (French)	Rt. hypochondrium	Liver	Fistulography, CT	Partial cystectomy & drainage	2 y, NR
14	Tschudi *et a*l	1988	47	M	Switzerland (German)	Rt. thoracic wall	Liver	CT	Wide local excision	4 ys, NR
15	Kehila *et al*	1987	86	F	Tunisia (French)	Rt. flank	Liver (VI–VII)	Fistulography, USG	Antibiotics, ext. catheter drainage	Not specified
16	Bazarov *et al*	1984	82	F	Russia (Russian)	Rt. abdomen	Liver	USG	Excision	2 mo, NR

USG, ultrasonography; CT, computed tomography; NR, no recurrence.

by *E. multilocularis*. *E. granulosus* is endemic in Mediterranean countries, the middle and far East, South America, Australia, New Zealand, and East Africa.[1] *E. multilocularis* has been frequently reported in the northern hemisphere, particularly Russia and Europe. The life cycle of this parasite exists in carnivores and in herbivores such as dogs and sheep. Humans are an accidental intermediate host and an end point in the parasite's life cycle. Although hydatid disease can develop anywhere in the human body, liver (52%–77%) is the most frequently involved organ, followed by the lungs (10%–40%).[17]

Echinococcosis may be asymptomatic for many years, its presence becoming evident when the liver is found to be enlarged or a cystic lesion is noted on imaging.[18,19] The clinical symptoms are related to pressure on adjacent organs or the development of complications. The cyst consists of an inner single-cell-thick germinal membrane, the endocyst, an acellular outer laminated ectocyst, and the external host-derived pericyst formed by hepatic parenchyma infiltrated by fibroblastin, endotheloid, and giant cells. A viable hydatid cyst is a space-occupying lesion with a tendency to grow. The cyst grows in the direction of the least resistance, and as a consequence of cyst enlargement, it can rupture. The known complications of hydatid cyst are, rupture into the peritoneal cavity, infection, compression of the biliary tree, intrabiliary rupture, anaphylaxis, and secondary hydatosis.[17] Rare modes of rupture are fistulization into the duodenum, perforation of the diaphragm, infiltration of ribs, and rupture into the vascular and urinary systems.[20–22]

Lewall and McCorkell[23] have classified the rupture of hepatic hydatid cysts into 3 categories: contained, communicating, and direct. Contained rupture occurs when only the endocyst ruptures and the cyst contents remain confined to the intact pericyst. Communicating rupture consists of a rupture of the endocyst with the escape of cyst contents into bronchioles or biliary radicals that are incorporated into the pericyst. Direct rupture is when both the endocyst and pericyst tear, causing a leakage of contents into the pleural or peritoneal cavities or other adjacent tissues.

Cutaneous involvement and spontaneous cutaneous fistulization is a very rare complication of liver hydatid cysts. Reasons for cutaneous fistulization include infection and continued expansion of the cyst causing pressure erosion and adhesion to the adjacent structures. In time, with increasing intracystic pressure, the cyst ruptures and inflammation leads to necrosis causing fistulization. Occult trauma could be one of the causes for rupture of hydatid cyst leading to fistula formation.[4] Erosion of rib and cyst–cutaneous fistula was reported in 1 patient with hydatid disease.[6] Usually cysts originating from the right hepatic lobe invade the right lateral abdominal wall and cysts from the left lobe invade the anterior abdominal wall.[6] Any subcutaneous hydatid cyst is more likely to be a primary invasion of hepatic echinococcosis or contamination during previous surgery rather than hematogenous metastasis.[24]

Surgical treatment combined with medical treatment to remove the cysts has good results in patients who are fit for the surgery. Surgical treatment includes resection of the primary cyst with excision of the fistula tract and diseased skin. It is important to carefully remove all the cysts from the peritoneal cavity to prevent recurrence. Also the patients should be treated postoperatively with albendazole for 4 to 6 weeks. Medical treatment alone may be useful in patients too frail to undergo a surgical procedure.

In summary, cutaneous involvement or cutaneous fistulization of hydatid cyst is a rare complication. Umbilical fistula caused by a liver hydatid cyst is an extremely rare presentation, which has not been reported in the past. Surgical treatment to remove the cyst and fistula tract combined with medical treatment is a successful approach in the treatment of hydatid cyst causing cutaneous fistula.

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