CASE REPORT

Spontaneous external fistula: the rarest presentation of hydatid cyst

Kumar Jayant, Swati Agrawal, Rajendra Agarwal, Krishna Gopal Dayama

Department of General Surgery, Sudha Hospital & Medical Research Centre, Kota, Rajasthan, India

Correspondence to

Dr Kumar Jayant,
jayantsun@yahoo.co.in

Accepted 16 April 2014

SUMMARY

A 65-year-old man with hydatid disease of the liver presented with a spontaneous cutaneous rupture draining about 10 L of fluid with daughter cysts. Contrast-enhanced CT showed multiple hypodense areas in the residual right lobe of the liver and abdominal wall with compensatory hypertrophy of the left lobe, confirming the diagnosis of a hydatid cyst presenting as external fistulisation which represents a very advanced stage of hydatid disease. Once the patient's nutritional status improved he was taken for surgery. On laparotomy there were multiple cysts seen in the residual right lobe of the liver and inflammatory changes in the abdominal wall with an external fistulous opening. Right-lobe hepatectomy was carried out along with excision of the fistulous tract. The specimen showed multiple degenerated contents of the cyst with numerous daughter cysts along the fistulous tract and examination of cyst fluid demonstrated protoscolices. The postoperative period was uneventful. He was regularly followed and was doing well.

BACKGROUND

Human echinococcosis is caused by the larval stages of cestodes of genus *Echinococcus*. Four species of *Echinococcus* are of medical importance. *Echinococcus granulosus* causes the most common form, that is, cystic echinococcosis (CE); *E. multilocularis* causes alveolar echinococcosis; *E. vogeli* causes polycystic echinococcosis. CE is seen all over the world and is endemic to South and Central America, Western Europe and the Middle East. Humans are accidental hosts and dogs the definitive hosts.¹ Most infections in humans are asymptomatic, but CE, also known as hydatid disease, causes slowly enlarging masses which can affect any part of the human body, but which most commonly affect the liver (55–68%) and lungs (18–28%). The cyst can rupture in the peritoneal cavity, pleural cavity, lung and into the biliary tree causing various complications, but cutaneous rupture leading to formation of spontaneous external fistula is terminal and extremely rare.^{2 3} After an extensive literature search we found very few reported cases of cutaneous rupture of liver hydatid and this is probably the first case of the disease ever reported with massive discharge of daughter cysts of approximately 10 L, leading to external fistula formation.

CASE PRESENTATION

A frail 65-year-old man who was a farmer by occupation presented to our emergency department with fever, abdominal pain and distension with

spontaneous occurrence of abdominal fistula draining about 10 L of fluid with jelly-like matter. There was no history of jaundice, breathlessness or trauma. The patient gave a history of having kept a pet dog for 10 years in his adulthood. On examination, there was an external fistula in the mid axillary line on the right at the sixth intercostal space, draining fluid with daughter cysts (figure 1). After initial resuscitation and stabilisation oral albendazole 400 mg twice daily was started.

INVESTIGATIONS

On an earlier hospital visit, ultrasonography of the abdomen was carried out for abdominal pain; the image showed numerous cysts in the right lobe of the liver and abdominal wall (figure 2). Blood investigations revealed haemoglobin (Hb) of 10 g/dL, total leucocyte count 20 000/mm³ with eosinophilia, erythrocyte sedimentation rate–100 mm/h, total bilirubin–7.1 mg/dL, direct bilirubin–6.1 mg/dL, normal levels of liver enzymes and normal renal function test. Tubercular serology was negative and serology for HIV, HBsAg and HCV was also negative while a serological ELISA test for hydatid disease was positive. Chest X-ray was normal. Later CT scan was performed which showed multiple hypodense areas in the residual right lobe of the liver and abdominal wall with non-calcified cyst wall. There were diffuse inflammatory changes in the abdominal wall which corresponded with the site of the external fistula. There was compensatory hypertrophy of the left lobe forming future liver remnant of 1000 mm³ (figure 3).

TREATMENT

After proper evaluation of the extent of the disease and improvement in the patient's nutritional and functional status (Hb–11 g/dL, serum albumin–3.6 g/dL), he was planned for surgery. On laparotomy there were multiple cysts present in the residual right lobe of the liver and inflammatory changes in the abdominal wall with an external fistulous opening. Before the dissection of cystic lesions, cyst fluid was aspirated and 10% povidone iodine was injected into the cyst, the fistulous tract was irrigated as well. Excision of the residual right lobe of the liver and abdominal wall along with the fistulous tract was carried out. An intraoperative specimen showed multiple heterogeneous degenerative contents of the cyst with some calcification in its wall; numerous small daughter cysts were also seen along the fistulous tract and cystic fluid examination showed protoscolices.

CrossMark

To cite: Jayant K, Agrawal S, Agarwal R, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-203784

Figure 1 External fistula in the right mid axillary line at sixth intercostal space draining fluid with daughter cysts.

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful and the patient was prescribed albendazole tablets and discharged. He was regularly followed and after 1 year he was symptom free.

DISCUSSION

Hydatid disease is caused by the *Echinococcus* spp, namely *E. granulosus*, *E. multilocularis*, *E. oligartus* and *E. vogeli*. The dog is the definitive host and sheep the intermediate host. Human is an accidental intermediate host and is the dead end of the parasite's life cycle.

The liver (55–70%) is the obvious first site after entry through the gut and passage in the portal circulation. Most cysts tend to be located in the right lobe (60–70%). Most commonly affected extrahepatic organs are the spleen, lung, kidney and brain.⁴ Uniocular lesions are typical of *E. granulosus*, and multilocular lesions are typical of *E. multilocularis* infection. When infection first occurs, the echinococcal larvae implant within the capillaries, triggering an inflammatory reaction composed of monocytes and eosinophils. Some larvae are destroyed by the host, but others encyst. Microscopic examination of hydatid cyst tissue demonstrates a characteristic encapsulated and calcified

cyst (eggshell calcification) that contains fluid and budding cells that will eventually become daughter cysts. The outer cyst wall is composed of many layers of delicate gelatinous sheets surrounded by a thick fibrous capsule. Hydatid disease shows a spectrum of presentation ranging from asymptomatic to rupture of the cyst, infection, fever, jaundice due to compression of the biliary radical or cystobiliary fistula, cholangitis and anaphylaxis, etc.⁵

Echinococcal cysts of the liver can cause complications in about 40% of cases. The most common complications in order of frequency are infection, rupture to the biliary tree; rupture to the peritoneal cavity; rupture to the pleural cavity. However, rupture in the gastrointestinal tract, bladder and vessels are rare.

We have discussed an extremely rare kind of complication not reported in the literature to date, that is, sudden spontaneous rupture of a hydatid cyst through the skin with voluminous discharge of fluid containing daughter cysts leading to external fistula formation. Although the exact mechanism of abdominal wall involvement is not known, it is presumed that growing cysts tend to slowly erode nearby structures. External fistulisation represents the terminal event of abdominal wall involvement in hydatid disease. Usually, a right-hepatic lobe cyst infiltrates into the right-lateral abdominal wall and a left-lobe cyst involves the anterior abdominal wall. Most commonly it is due to acute suppurative infection with increasing intracystic pressure leading to destruction of all layers of the abdominal wall with fistula formation.^{6,7} In our case the affected lobe of

Figure 2 Ultrasonography of the abdomen showing numerous cysts in the right lobe of the liver and abdominal wall.

Figure 3 Multiple hypodense areas in the right lobe of the liver and abdominal wall with non-calcified cyst wall with compensatory hypertrophy of the left liver.

Table 1 WHO Informal Working Groups on Echinococcosis (IWGE)

WHO-IWGE	Description	Stage
CE1	Unilocular anechoic cystic lesion with double-line sign	Active
CE2	Multiseptated, 'rosette like' 'honeycomb' cyst	Active
CE3 A	Cyst with detached membranes (Water lily sign)	Transitional
CE3 B	Cyst with daughter cysts in solid matrix	Transitional
CE4	Cyst with heterogeneous hypoechoic/hyperechoic content. No daughter cysts	Inactive
CE5	Solid cyst with calcified wall	Inactive

liver was also on the right and had involved the right-lateral wall of the abdomen leading to external fistula formation.

The diagnosis of hydatid cyst is usually made by ultrasound. Based on ultrasound findings, WHO Informal Working Groups on Echinococcosis (IWGE) has described five categories. This classification system not only tells the functional status of the cyst but also helps in the selection of standardised treatment and follow-up. Types CL, CE1 and CE2 are active cysts while CE4 and CE5 are inactive^{8 9} (table 1). Unfortunately, in our case complication of spontaneous external fistulisation had already occurred, so assessment of morphology and structure of the cyst by ultrasound had no role. Contrast-enhanced CT has evolved as the most sensitive modality to diagnose complicated hydatid disease. It has an important role in localising the site of origin,

extent of organ and surrounding structure involvement along with assessment of the fistulous tract.¹⁰ CT of the index case showed massive destruction of the right lobe of the liver with compensatory hypertrophy of the residual left lobe, destruction of the anterolateral abdominal wall and residual cysts. The severity of the disease and poor nutritional status of the patient made this case very challenging.

There are multiple treatment options for hydatid disease which include medical management, minimally invasive methods such as Percutaneous Aspiration, Instillation and Reaspiration (PAIR) and surgery such as deroofing, pericystectomy and liver resection.¹¹ However, our case was of complicated right-lobe liver hydatid with compensatory left-lobe hypertrophy and abdominal wall destruction with cutaneous fistula formation. Right-lobe hepatectomy and fistulous tract excision with abdominal wall reconstruction was carried out. The patient was also given preoperative and postoperative albendazole therapy for 12 weeks. Medical treatment alone may be used in patients who are not able to tolerate surgery.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Sakorafas GH, Stafyla V, Kassaras G. Spontaneous cyst cutaneous fistula: an extremely rare presentation of hydatid liver cyst. *Am J Surg* 2006;192:205–6.
- 2 Grigylis G, Yzet T, Flamant M, et al. Cutaneous fistulization of a liver hydatid cyst. *Gastroenterol Clin Biol* 2004;28:819–20.
- 3 Korwar V, Subhas G, Gaddikeri P, et al. Hydatid disease presenting as cutaneous fistula: review of a rare clinical presentation. *Int Surg* 2011;96:697–103.
- 4 Kehila M, Allègue M, Abdesslem M, et al. Spontaneous cutaneous-cystic-hepatic-bronchial fistula due to a hydatid cyst. *Tunis Med* 1987;65:267–70.
- 5 Borras JA. Subcutaneous rupture of a hydatid cyst of the liver. *Med Esp* 1958;40:344–51.
- 6 Di Cataldo A, Latino R, Cocuzza A, et al. Unexplainable development of a hydatid cyst. *World J Gastroenterol* 2009;15:3309–11.
- 7 Yakan S, Yildirim M, Coker A. Spontaneous cutaneous fistula of an infected liver hydatid cyst. *Turk J Gastroenterol* 2009;20:2993.
- 8 Bastid C, Pirro N, Sahel J. Cutaneous fistulation of a liver hydatid cyst. *Gastroenterol Clin Biol* 2005;29:748–9.
- 9 Brunetti E, Kern P, Vuitton DA, Writing Panel for the WHO-IWGE. Expert consensus for the diagnosis and treatment of cystic and alveolar echinococcosis in humans. *Acta Trop* 2010;114:1–16.
- 10 Salerno C E, Lo Casto A. Subcutaneous rupture of a hepatic hydatid cyst: CT findings. *Dig Liver Dis* 2006;38:619–20.
- 11 Kjossev K, Belokonski E, Vladov N. TN (R) C points system for cystic echinococcosis: clinical evaluating and monitoring of the disease. *Proceedings of 44th Congress of the European Society for Surgical Research Nimes, France/May 20–23*. Monduzzi Editore; 2009. p. 61–14.

Learning points

- External fistula formation following spontaneous cutaneous rupture of a hydatid cyst of the liver with massive discharge of daughter cysts is the rarest presentation of this disease so far.
- External fistulisation represents a very advanced stage of hydatid disease.
- A hydatid cyst is usually diagnosed on ultrasound but contrast-enhanced CT is diagnostic for complicated hydatid disease.
- Treatment consists of surgery such as cystectomy or hepatectomy with removal of the fistulous tract combined with medical treatment.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow