Rare disease

Primary disseminated extrahepatic abdominal hydatid cyst: a rare disease

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Summary

Hydatid disease (HD) is a parasitic disease, most frequently caused by *Echinococcus granulosus* or *Echinococcus multilocularis* and rarely by *Echinococcus vogeli* or *Echinococcus oligarthus*. Mostly, hydatid cyst is primarily in liver (75%) and lung (15%). Peritoneal HD (13%) is usually secondary. Dissemination of HD may occur following rupture of the hydatid cyst into the peritoneal cavity. Primary peritoneal hydatid cyst disease is a rare phenomenon. We present a case of primary disseminated abdominal HD presenting with the complaint of a pain in the epigastric region along with intermittent fever. Radiological evaluation and serological examination confirmed it as primary disseminated HD involving pancreas, a cyst anterior to the left lobe of the liver just below the peritoneum and the left kidney.

BACKGROUND

Hydatid disease (HD), mostly caused by cestode *Echinococcus granulosus*, is a significant health problem where animal husbandry is common. Dogs or other carnivores are definitive hosts, whereas sheep or other ruminants are intermediate hosts. Man becomes an accidental intermediate host by the ingestion of eggs that develop into cysts, causing complication and even mortality (4%). Common sites include the liver (75%) and lungs (15%). Peritoneal echinococcosis (13%) is usually secondary. Primary extrahepatic peritoneal echinococcosis is rare.¹

CASE PRESENTATION

A 40-year-old male patient presented with a pain in the epigastric region for 3-year duration with abdominal distension in the epigastric region with intermittent fever. Bowel and bladder habits were normal. Abdominal

examination revealed firm mass in the epigastric region with mild tenderness over the lump.

INVESTIGATIONS

Haematological and biochemical profiles were within normal limits. Ultrasonography of the abdomen revealed well-defined multiple calcified cystic lesions with a well-defined wall in the pancreas, suggestive of a hydatid cyst. Contrast-enhanced CT (CECT) of the abdomen revealed multiple well-defined peripherally enhancing cystic lesions in the body and tail of pancreas, a cyst just anterior to the left lobe of the liver abutting the peritoneum and a cyst in the left kidney (figures 1 and 2). ELISA for hydatid was positive. Chest x-ray was normal.

DIFFERENTIAL DIAGNOSIS

Abdominal tuberculosis and multiple pseudopancreatic cysts.

Figure 1 Contrast-enhanced CT of the abdomen, showing a cyst below the peritoneum, multiple cysts in the pancreas and a cyst in the left kidney.

Figure 2 Contrast-enhanced CT of the abdomen, showing multiple hydatid cysts in the pancreas.

TREATMENT

Exploratory laparotomy was done after giving albendazole (400 mg twice daily) for 1 month. There was a cyst in the epigastric region just below the peritoneum, multiple cysts in the pancreas and a cyst in the left kidney was confirmed on needle aspiration. The epigastric cyst was completely excised and sent for histopathological examination. Other cysts were treated by the Percutaneous Aspiration and Instillation of a scolical agent (10% povidine iodine) and Reaspiration (PAIR) therapy. Postoperative period was uneventful. Histopathological examination report was suggestive of a hydatid cyst.

OUTCOME AND FOLLOW-UP

The patient is in follow-up and doing well.

DISCUSSION

Extrahepatic intraperitoneal hydatid cyst disease is a rare presentation. HD or echinococcosis is a parasitic disease caused by infection with the larva (metacestode) of the cestode echinococcus. Four species of the genus *Echinococcus* are known to cause infection in humans: *E. granulosus* (cystic HD), *Echinococcus multilocularis* (alveolar HD) *Echinococcus vogeli* and *Echinococcus oligarthus* (both causing polycystic HD).² The life cycle of *E. granulosus* involves two hosts. The definitive host is usually a dog but may also be some other carnivore. The adult worm of the parasite lives in the proximal small bowel of the definitive host, attached by hooklets to the mucosa. Eggs are released into the host's intestine and excreted in the faeces. Sheep are the most common intermediate hosts. They ingest the ovum while grazing on contaminated ground. The ovum loses its protective chitinous layer as it is digested in the duodenum. The released hexacanth embryo or oncosphere passes through the intestinal wall into the portal circulation and develops into a cyst within the liver (figure 3). When the definitive host eats the viscera of the intermediate host, the cycle is completed. Humans may become intermediate hosts through contact with a definitive host (usually a domesticated dog) or

ingestion of contaminated water or vegetables.³ Therefore, humans become accidental intermediate hosts. The most common site involved is the liver (59–75%), followed in frequency by lungs (27%), kidney (3%), bone (1–4%) and brain (1–2%). Other sites such as the heart, spleen, pancreas, omentum, ovaries, parametrium, pelvis, thyroid, orbit or retroperitoneum, and muscles are very rarely affected.^{4 5}

Clinical manifestations are caused by the mass effect of an enlarging abdominal cyst. Pain may be of acute onset if the cyst ruptures but later on becomes of continuous dull aching in nature. Fever supervenes if there is secondary infection. Jaundice develops if there is intrabiliary rupture of the cyst.⁶ Primary peritoneal echinococcosis accounts for 2% of all abdominal hydatidosis⁴ without evidence of cysts in other intra-abdominal organs. Dissemination occurs by either lymphatic⁷ or systemic⁸ circulation. Intraperitoneal hydatid cysts usually develop secondary to spontaneous, traumatic or iatrogenic rupture of hepatic, splenic or mesenteric cysts.³ Balik *et al* reviewed 27 patients retrospectively who were treated surgically between 1981 and 1999 because of extrahepatic HD. In the study, 19 patients had a coexistent hepatic cyst (70.4%), while 8 patients (20.6%) had only extrahepatic cysts located in the spleen (3 patients), pancreas (2 patients), adrenal glands (4 patients), mesocolon (5 patients), mesentery of the small intestine (1 patient), ovaries (1 patient), retroperitoneum (4 patients) and omentum (2 patients).⁹ Another study by Wani *et al* reviewed retrospectively 183 patients of abdominal hydatid cysts managed surgically from January 1998 to December 2000. They found that 12% of the patients had only extrahepatic abdominal cyst.¹⁰

In our case, multiple cysts involving pancreatic parenchyma, a cyst anterior to the left lobe of the liver and a cyst in the left kidney were found. Serology and imaging are the main tools for establishing diagnosis. Ultrasound is the preferred first-line imaging, but CECT gives more precise information regarding the morphology (the size, location, neighbourhood and number) of the cyst.¹ The

Figure 3 Life cycle of *Echinococcus granulosus*.

most common form of treatment is surgical removal of the cysts combined with chemotherapy using albendazole and/or mebendazole before and after surgery.¹

Learning points

- Extra-hepatic primary abdominal disseminated disease is a rare presentation and should be kept in mind during the diagnosis of chronic pain in the abdomen.

Competing interests None.

Patient consent Obtained.

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