Rare disease

Primary retroperitoneal echinococcal cyst

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Summary

A 74-year-old man was admitted with right flank pain and discomfort lasting for 2 months. CT scanning revealed a large retroperitoneal cystic mass. There were no cysts elsewhere. Serological testing revealed hydatid disease. Preoperatively he was treated by Albendazole 400 mg for 1 month and then underwent laparotomy. The entire mass was excised en bloc and intact and right hemicolectomy was simultaneously performed for excision of the adhered ascending colon. He recovered uneventfully and was discharged on the eighth day, Albendazole was given and follow-up visits were arranged for every 6 months. Total cystectomy in case of active echinococcal cysts remains the treatment of choice.

BACKGROUND

Hydatid disease is a zoonotic infection prevalent in sheep and cattle farming areas of Australasia, North and East Africa, South America and the Southeast Mediterranean basin.^{1–3} In 85–95% of the cases, the liver and/or the lungs are involved.⁴ In humans, echinococcosis can be a serious disease, with death rate of 4–5%.⁴ Primary retroperitoneal echinococcal cyst is one of the rarest forms of presentation of echinococcosis.^{2,5}

CASE PRESENTATION

A 74 year-old man, who did not report medical problems and was never operated for any reason in the past addressed his family physician complaining of right flank pain and abdominal discomfort lasting for 2 months. During this period he was afebrile without any signs of acute infection. Bowel and bladder habits were normal. Abdominal examination revealed a firm mass in the right flank with mild tenderness over the lump. First, he was investigated in the local district hospital and the CT abdominal scan revealed retroperitoneal multiseptated hydatid tumour with daughter cysts inside and without calcification. He was referred to our tertiary anticancer hospital for further treatment.

INVESTIGATIONS

CT abdominal scan confirmed the diagnosis of retroperitoneal isolated multiseptated hydatid cyst containing daughter cysts without calcifications (cystic echinococcosis 2 according to WHO informal working group on echinococcosis; figures 1 and 2). ELISA for hydatid was positive. CT scan of the head and chest were negative. Haematological and biochemical profiles were within the normal limits. Colonoscopy revealed extrinsic pressure on the region of the ascending colon.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of retroperitoneal cystic mass includes retrorectal cyst, teratoma, cystic lymphangioma, abscesses and necrotic malignant soft tissue tumours.⁵ It

Figure 1 CT scan of the abdomen showing a multiseptated, fluid-filled structure, without calcifications.

would be prudent to include hydatid disease, especially if the patient comes from the endemic area.

TREATMENT

Preoperatively, the patient was treated by albendazole 400 mg twice daily for 1 month. A retroperitoneal mass

Figure 2 CT scan of the abdomen showing a multiseptated, fluid-filled structure, without calcifications.

adhered to the ascending colon was identified on laparotomy. After careful examination of the rest of the abdomen, no pathology was detected elsewhere. The whole mass was excised intact by performing simultaneously total cystectomy and right hemicolectomy; consequently ileo-transverse anastomosis performed. The histopathological examination report confirmed the diagnosis of the hydatid cyst.

OUTCOME AND FOLLOW-UP

The patient is in follow-up and doing well.

DISCUSSION

Hydatid disease or echinococcosis is a parasitic disease caused by infection with the larva of the cestode echinococcus.^{6 7} Accidental ingestion of garden vegetables, water or soil contaminated with tapeworm eggs from dog faeces initiates hydatid disease^{4 6 7} so it is important to educate populations in endemic areas about washing of food and hands. The feeding of livestock entrails to dogs is a common starting point for the tapeworm life cycle and should be avoided.⁶ Educational programmes should be started early because presumably most infections are acquired in childhood.⁶⁻⁸

In humans, echinococcosis can be a serious disease, with death rate of 4–5%.⁴ Serological testing is more specific but less sensitive than most imaging modalities.^{9 10} Cysts may be asymptomatic for 10–20 years until they become large enough to produce symptoms.⁸ Cysts grow from 1 to 5 cm /year.^{6 8} Echinococcal cysts are classified into six types according to WHO-IWGE classification of ultrasound images of echinococcal cysts.¹¹ CL is defined cystic lesion unilocular with anechoic content. They need differential diagnosis with further imaging techniques. CE1 is an active status and are characterised with visible cyst wall and snow flake sign. CE2 are multivesicular that produce ‘wheel-like’ structures and the presence of daughter cysts is indicated by ‘rosette-like’ or ‘honey-comb-like’ structures. Their status is active and fertile. CE3 are characterised by anechoic content with detachment of laminated membrane visible as ‘water-lily’ sign. CE4 are characterised by heterogeneous hypoechoic or hyperechoic content. There are no daughter cysts and their status is inactive. Their features are not pathognomonic and further diagnostic tests are required to ascertain the diagnosis. CE5 are characterised by thick calcified wall that is arch shaped producing a cone-shaped shadow and in most cases are not fertile. So far, retroperitoneal lesions of newly diagnosed hydatid cysts consist only 1.1%⁵ of all the reported cases.⁵ They are usually discovered as a palpable mass or as a cause of flank pain. Retroperitoneal involvement was always thought to be secondary to rupture or spillage during surgery for liver echinococcosis. Primary retroperitoneal hydatid cyst without other organ involvement was first reported by Lockhart J and Sapinza VC in 1958.¹² In 1973 Mukherjee *et al*¹³ reported nine cases, two had died due to anaphylactic reaction resulting from spillage during excision or biopsy done with the

misdiagnosis of a retroperitoneal tumour. Preoperative diagnosis of retroperitoneal hydatid disease is difficult. The differential diagnosis of retroperitoneal cystic mass includes retrorectal cyst, teratoma, cystic lymphangioma, abscesses and necrotic malignant soft tissue tumours.⁵ It would be prudent to include hydatid disease, especially if the patient comes from the endemic area. Therapeutic strategy of extrahepatic hydatid disease is based on considerations regarding size, location and manifestations of the cyst. Asymptomatic small cysts can be treated with antihelminthic drugs administered for 28 days in one-to-eight repeating cycles separated by drug-free intervals of 2 weeks.¹⁴ For symptomatic large peritoneal cysts, when feasible, total cystectomy is the gold standard. In case where cysts are firmly attached to vessels or multiple viscera, deroofting and drainage has been proved to be a safe method.¹⁴

Learning points

Extrahepatic primary echinococcosis is a rare presentation and should be kept in mind during the differential diagnosis of flank pain.

Competing interests None

Patient consent Obtained

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