CLINICAL IMAGES

Obstructive jaundice and hydatid cysts mimicking choledochal cyst

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Hydatid disease remains a common problem in endemic regions throughout the world. Endemic areas include Australasia, Africa, the Americas, the Middle East, Greece, Spain and Eastern Europe.[1-4]

Hydatid cysts (HCs) typically occur in the liver and lungs, but can occur at multiple other sites throughout the body.[4,5] Sites of 1 802 cysts recorded in adult patients in the Australian Hydatid Register included: liver 63%; lung 25%; muscles 5%; bones 3%; kidney 2%; spleen and brain 1%; and heart, breast, parotid glands and pancreas < 1%.[6] HCs usually occur at typical sites in adults, however there is a higher percentage of atypical sites and unusual presenting symptoms in children.[5]

Obstructive jaundice is an infrequent association with HCs. It usually results from the rupture of a cyst into a bile duct,[7] from a choledochal cyst,[5,8] or from common bile duct (CBD) compression.[6,9] More recently, a HC within the CBD wall was reported for the first time.[8] Case 1 of this study constitutes the second such report.

We present 2 cases where severe obstructive jaundice resulted from occlusion of the CBD lumen. In case 1 this was caused by a HC localised within the CBD wall itself, and in case 2 by external compression from a HC localised in the head of the pancreas. Both were difficult to differentiate from a type 1 choledochal cyst on clinical grounds and presented difficult surgical decision making.

Case 1

An 8-year-old girl was referred to Tygerberg Hospital with a 3-week history of abdominal pain and progressive jaundice. She had previously lived in a high-prevalence area for echinococcosis and was otherwise stable without any signs of systemic infection.

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Her preoperative laboratory results confirmed obstructive jaundice on liver function tests. The hepatitis screen was negative, and serology for *Entamoeba histolytica* and *Echinococcus granulosus* was also negative. Clinically, a central, painless epigastric mass was palpable on abdominal palpation.

Preoperative chest and abdominal radiographs were within normal limits but an abdominal ultrasound scan revealed a cystic 5. 7 x 5. 1 x 5. 2 cm subhepatic mass, with a wall thickness of 2.2 mm. Dilated intra- and extra-hepatic ducts were identified (Fig. 1). A subsequent hepatic iminodiacetic acid (HIDA) scan demonstrated impaired hepatocellular function, delayed passage of contrast to the bile duct, and a photopenic

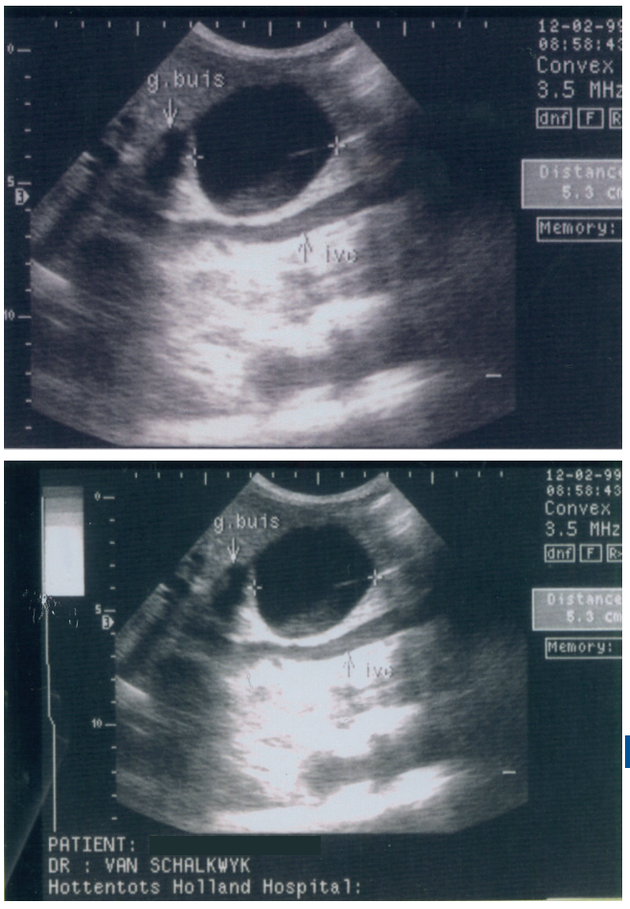

Fig. 1. Abdominal ultrasound scan, patient 1.

area below the liver. CT scan showed the cystic mass adjacent to the distended CBD and gallbladder (GB), suggestive of a type 1 choledochal cyst (Fig. 2).

At laparotomy a large, distended GB with associated cystic dilatation of the CBD was identified, extending towards the head of the pancreas. The cyst also tapered clearly towards the head of the pancreas as one would expect with a type 1 choledochal cyst. An intraoperative cholangiogram, performed via the GB, demonstrated dilated intrahepatic ducts, but no passage of contrast downwards to the CBD (Fig. 3). The cyst did not fill with contrast.

A cholecystectomy and excision of the cyst was performed with difficulty because of the inflammatory reaction around the cyst. The cyst contained clear fluid without bile content, and on opening the cyst the inner layer stripped very easily although it was abnormally thick. The cyst was excised and ligated where it tapered clearly towards the head of the pancreas (as in a type 1 choledochal cyst). A Roux-en-Y hepaticoenterostomy was performed to re-establish the bile flow.

Histological examination of the cyst wall reported typical features of an echinococcal cyst. As the diagnosis was not anticipated, the cyst was not injected or packed with impregnated swabs before opening.

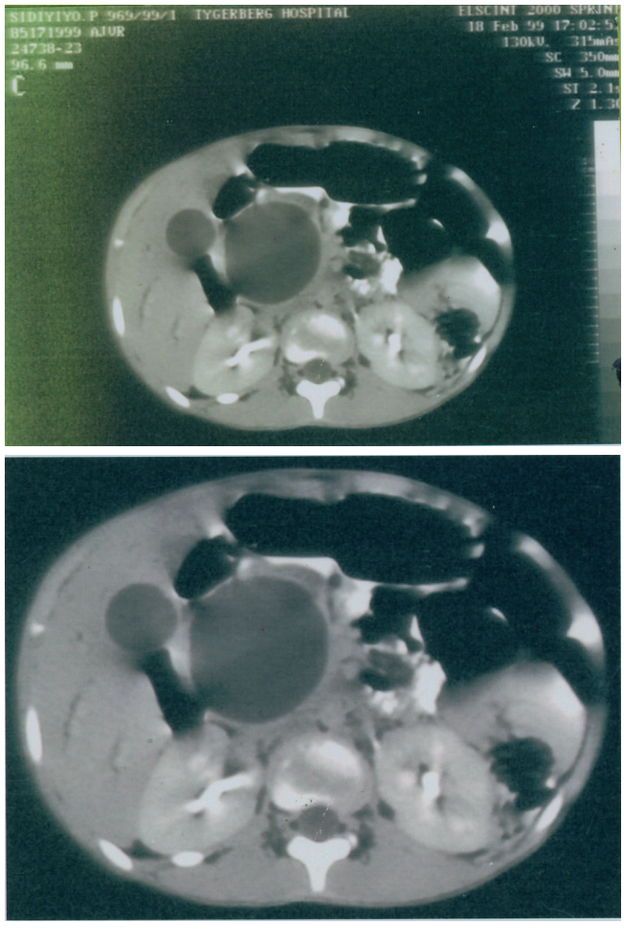

Fig. 2. CT scan, patient 1.

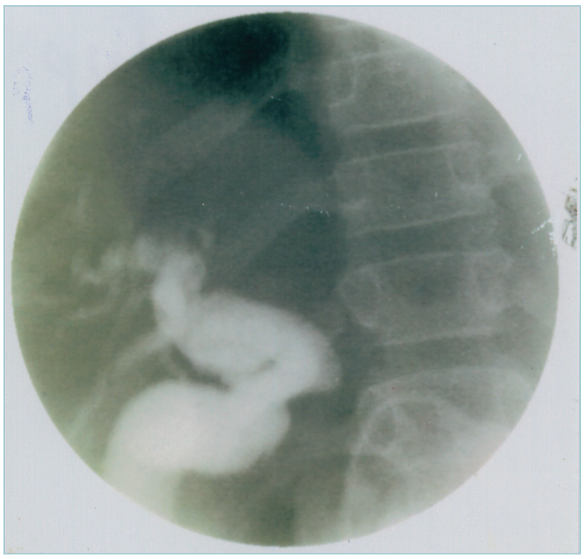

Fig. 3. Intraoperative cholangiogram of patient 1.

The postoperative course was complicated by a secondary haemorrhage, with blood oozing from the raw edges of the cyst, requiring re-laparotomy on day 2. The jaundice settled and the initial postoperative course was uneventful. On day 10 after surgery the patient developed acute abdominal pain associated with a left-side pleural effusion, necessitating an intercostal drain. Serum amylase levels and amylase levels from the pleural fluid were significantly raised leading to a diagnosis of postoperative pancreatitis. The patient recovered well with parenteral nutritional support and supportive care and was discharged after 14 days. Treatment with albendazole was commenced as soon as the ileus had resolved. A localised intra-abdominal abscess required surgical drainage 1 month later. Long-term outcome was good, with no postoperative cholangitis and no recurrence of hydatid disease.

Case 2

A 14-year-old boy from a country town was admitted with a 1-month history of obstructive jaundice associated with a mild pruritus. Liver function tests confirmed an obstructive jaundice (total bilirubin 122 µmol/l, conjugated bilirubin 76 µmol/l, gamma-glutamyl tranferase 394 U/l, and alkaline phosphatase 670 U/l). He was otherwise clinically well with no palpable abdominal tumour on examination.

Abdominal computed tomography (CT) scan revealed the presence of a large cystic mass in the area of the head of the pancreas causing intrahepatic bile duct dilatation suggestive of a type 1 choledochal cyst. The patient was prepared for surgery and at laparotomy a cystic mass in the region of the pancreas head was identified surrounded by numerous large vessels.

Clear fluid and obvious hydatid daughter cysts were aspirated from the cyst. Cyst contents were then aspirated, and surrounding tissues rinsed with 5% saline. The cyst was then excised and a Roux-en-Y hepaticoenterostomy performed.

The postoperative course was uneventful. The patient's jaundice resolved clinically and he was discharged home on albendazole therapy. Long-term outcome was good with no recurrence of hydatid disease.

Discussion

HCs are formed as a result of infestation with the cestode *E. granulosus* (occasionally *E. multilocularis*). The adult form of the worm lives in the jejunum of the definitive host (usually dogs). Eggs passed in the stools are ingested by the intermediate host (humans, sheep, pigs and cows). Embryos pass through the intestinal mucosa and via the portal system reach the liver or bypass it and then reach the lungs or other organs. There are reports of HCs occurring in the liver (63%), lung (25%), muscles (5%), bones (3%), kidney (2%), spleen and brain (1%), and heart, breast, parotid glands and pancreas (< 1%).[6]

Subsequently a cyst develops, comprising two layers. The outer pericyst is fibrous and the inner layer consists of germinative tissue. The scolices are contained within the inner layer and daughter cysts may be seen floating in clear fluid within the cyst.[6] Although the daughter cysts may be recognised on CT scan, they were absent on imaging in both of our patients. It is worthwhile mentioning that the CT scans of the two patients looked almost identical; we therefore present a scan for the first case only (Fig. 2).

The accuracy of serodiagnosis has been estimated at 75 - 95%[10] but may be complicated by cross-reactivity with other parasites. It is interesting to note that both these particular patients were seronegative on testing.

Reported complications of hydatid disease include infection, rupture, anaphylaxis, biliary obstruction, and parenchymal damage by disease. Although obstructive jaundice has been reported it is a rare association and may occur as a result of cyst rupture into a bile duct, or compression of the CBD by a cyst localised either in the liver or in the head of the pancreas.[9,11] Case 1 in this report illustrates that a cyst primarily localised in the CBD wall results in obstruction and may as such be confused with a choledochal cyst because of its fusiform dilatation.

Atypical presentation and sites are not uncommon in children[5] and should be considered in children where hydatid disease enters the differential diagnosis. Both of these cases illustrate how atypical features may lead to diagnostic difficulties and difficult management decisions. In both cases the preoperative tests failed to suggest hydatid disease (serology was negative and imaging did not disclose daughter cysts). The only report of HCs of the CBD causing obstructive jaundice[8] was published after our first case. HC should therefore be added to the differential diagnosis of a CBD cystic dilatation in endemic areas for hydatid disease, bearing in mind its tendency to unusual presentation. Magnetic resonance cholangio-pancreatography (MRCP) may be of value in delineating anatomy in future cases.

The treatment of hydatid disease remains surgical, with inactivation of the cysts with hypertonic saline, peri-cystectomy and biliary drainage being the mainstays of treatment. The use of albendazole for multiple cysts, intraoperative spill, non-surgical removal or preoperative sterilisation of the cysts remains an increasingly popular means of management. Surgical treatment was successful in both cases. Although the first patient had a complicated postoperative course (bleeding, pancreatitis, abscess) she eventually made an excellent recovery and was well on follow-up, receiving treatment with albendazole. Our second patient had a totally uncomplicated postoperative course and follow-up.

In conclusion, a HC should be considered in the differential diagnosis of an older child from an endemic area presenting with obstructive jaundice and a cystic mass in the region of the CBD.

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