Primary hydatid cysts of the pancreas

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Summary

Pancreatic involvement by hydatid disease is uncommon. Establishing a precise diagnosis may be difficult because the presenting symptoms and findings of investigations may be similar to other more commonly encountered cystic lesions of the pancreas. We report 4 patients with primary hydatid cysts in the head of the pancreas. The records of all patients treated for hydatid disease from 1980 to 2000 were reviewed. During the study period a total of 280 patients were treated, 4 of whom had hydatid disease involving only the pancreas. The 4 patients (3 women, 1 man) ranged in age from 17 to 60 years. Three patients presented with jaundice, abdominal pain and weight loss, 2 with hepatomegaly and 1 with an epigastric mass. All 4 lesions involved the head of the pancreas and ranged in size from 3 to 10 cm in diameter. In 2 patients the investigations incorrectly suggested a cystic tumour and both underwent pancreaticoduodenectomy. In 2 patients the correct diagnosis allowed local excision to be performed. Hydatid cyst is a rare cause of a cystic mass in the head of the pancreas, but should be included in the differential diagnosis of cystic lesions of the pancreas, especially in endemic areas.

Case reports

Case 1

A 60-year-old woman presented with a 2-month history of jaundice. An 8 cm hepatomegaly and an enlarged gallbladder were palpable. A separate firm, non-tender epigastric mass was present. Biochemistry was as shown in Table I. Ultrasound demonstrated dilatation of both intra- and extrahepatic bile ducts with a large gallbladder containing sludge. The common bile duct was obstructed by a 10 cm diameter cyst in the head of the pancreas. Endoscopic retrograde cholangiopancreatography (ERCP) showed tapering of the main pancreatic duct in the pancreatic head with an obstructed duct at the junction of the pancreatic head and body.

At laparotomy a 10 cm cystic mass suggestive of a hydatid cyst was present in the head of the pancreas. The distal pancreas was atrophic with dilatation of the obstructed main pancreatic duct. The cyst was isolated with swabs soaked in fresh 0.5% Eusol solution, aspirated with a wide-bore needle, and filled with Eusol. The cyst was carefully opened and the intact endocyst removed. No communication with the pancreatic duct was present. The ectocyst was excised. Histopathological examination confirmed a hydatid cyst and demonstrated a periodic acid Schiff (PAS)-positive chitinous layer and a germinal layer containing scolices. Postoperative recovery was complicated by transient right basal pneumonia.

Case 2

A 17-year-old woman presented with jaundice and a 5 cm, firm hepatomegaly. There was no history of pancreatitis or identifiable antecedent factors, including alcohol abuse, gallstones, or trauma. Biochemistry was as shown in Table I. A computed tomography (CT) scan demonstrated a 3 cm cyst in the head of the pancreas obstructing both pancreatic and bile ducts. ERCP demonstrated a grossly dilated common bile duct with a tapered stenosis at the lower end. The proximal pancreatic duct was displaced medially with stenosis in the neck region and marked upstream duct dilatation. No communication between the cyst and the biliary or pancreatic duct was demonstrated. At duodenoscopy a distinct intraluminal bulge was noted in the peripancreatic region. Ultrasound-guided aspiration of the cyst produced clear fluid with a low amylase content, no malignant cells, and no

Methodology

The records of all patients treated for hydatid disease from 1980 to 2000 in the Department of Surgery at the University of Cape Town and Groote Schuur Hospital, Cape Town, were reviewed. During this 21-year period a total of 280 patients were treated, 4 of whom had hydatid disease involving the pancreas.
hydatid hooklets or protoscolices. Aspiration did not relieve the obstruction. Laparotomy revealed a cystic mass in the head of the pancreas with gross dilatation of the main bile duct and gallbladder. The body and tail of the pancreas appeared normal. A pylorus-preserving pancreatoduodenectomy was performed with an end-to-side pancreatojejunostomy. Histology of the resected specimen showed necrotic hydatid laminated membrane but no scolices or hooklets. Postoperative recovery was uneventful.

Case 3

A 52-year-old woman presented with a 5-month history of epigastric pain and jaundice. An abdominal CT scan showed intra- and extra-hepatic biliary dilatation with common bile duct diameter of 9.4 mm and a 4 cm cystic mass in the head of the pancreas (Fig. 1). ERCP was unsuccessful. A preoperative diagnosis of a cystic neoplasm of the pancreas was made. Biochemistry was as shown in Table I. Laparotomy revealed a cystic mass in the head of the pancreas. Intraoperative fine needle aspiration cytology showed no scolices or malignant cells. A pylorus-preserving pancreatoduodenectomy was performed with an end-to-side pancreatojejunostomy. The histology of resected specimen showed the features of a hydatid cyst.

Case 4

A 17-year-old male presented with a 3-week history of epigastric and left upper quadrant pain associated with nausea and vomiting. There was no history of trauma, alcohol abuse or gallstones. A mass was palpable in the left upper quadrant. An abdominal CT scan showed a large cyst in the region of the pancreatic tail. The serum amylase was over 10 times the normal limit. A second multiloculated cystic mass measuring 9 x 8 cm was noted in the head of the pancreas (Fig. 2). At laparotomy a cystic mass was found in the head of the pancreas and a second cystic mass was present in the tail of the pancreas. Both cysts were aspirated and cyst fluid was sent for measurement of the amylase level. The cyst fluid amylase levels were 120 and 134 000 IU/l respectively. The pancreatic tail cyst was excised and histological examination of this confirmed a pseudocyst. The cyst in the head of the pancreas (amylase level 120 IU/l) was opened and found to contain hydatid sand with a laminated mem-

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**TABLE I. CLINICAL DATA: PATIENTS WITH HYDATID CYST IN PANCREAS**

<table>
<thead>
<tr>
<th>Patient 1</th>
<th>Patient 2</th>
<th>Patient 3</th>
<th>Patient 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (yrs)</td>
<td>60</td>
<td>17</td>
<td>52</td>
</tr>
<tr>
<td>Sex</td>
<td>Female</td>
<td>Female</td>
<td>Female</td>
</tr>
<tr>
<td>Jaundice</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Weight loss</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Hepatomegaly</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Epigastric mass</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Haemoglobin (g/dl)</td>
<td>12.3</td>
<td>10.9</td>
<td>14.4</td>
</tr>
<tr>
<td>White cell count (×10^9/l)</td>
<td>6.0</td>
<td>5.8</td>
<td>7.5</td>
</tr>
<tr>
<td>Total bilirubin (µmol/l)</td>
<td>74</td>
<td>107</td>
<td>16</td>
</tr>
<tr>
<td>Alkaline phosphatase (IU/l)</td>
<td>400</td>
<td>243</td>
<td>241</td>
</tr>
<tr>
<td>Dilated bile duct on US</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Maximum diameter of hydatid in HOP</td>
<td>10 cm</td>
<td>3 cm</td>
<td>4 cm</td>
</tr>
<tr>
<td>Surgical management</td>
<td>Cystectomy</td>
<td>PPPD with end-to-side PJ and CJ</td>
<td>PPPD with end-to-side PG and CJ</td>
</tr>
</tbody>
</table>

HOP = head of pancreas; US = ultrasound; PPPD = pylorus-preserving pancreatoduodenectomy; PJ = pancreaticojejunostomy; PG = pancreaticogastrostomy; CJ = choledochojjunostomy.
brane. The cyst was excised, and histology of the excised specimen confirmed a hydatid cyst. The hydatid cyst in the head of the pancreas had presumably obstructed the pancreatic duct and caused the pseudocyst in the tail of the pancreas. The patient made an uneventful recovery and was well 8 months later.

Discussion

The most common form of hydatid disease is caused by *Echinococcus granulosus*, a dimorphic parasite that exists exclusively as a short flat worm (cestode) in the primary host (usually a dog) or as a cystic form in the intermediate host (usually sheep, cows or pigs). Man is an accidental intermediate host.1 The eggs of the worm are excreted in the infected dog’s faeces. A grazing animal swallows the ova, which hatch in the jejunum and the embryos cross the intestinal mucosa into the portal system. In most cases the liver is the final destination of the intermediate host or, if the embryo bypasses the liver, the lung is the next most frequently involved organ.8 If the embryo continues through the pulmonary capillary bed, the hydatid cyst may develop at any site in the body. Other less commonly involved organs are bone, brain, kidney, spleen and orbit.8 Pancreatic involvement is rare.1011 The most probable route of pancreatic infestation is haematogenous dissemination after passage of the embryo through both liver and lung.8 Other possible routes, such as migration of embryos down the bile duct into the pancreatic duct, via the lymphatic circulation from the intestinal mucosa to the pancreas, or by portal blood into pancreatic veins, have been suggested but are less likely.1011

The clinical presentation of hydatid disease of the pancreas is the result of pressure by the cyst on adjacent structures and depends on the size and anatomical location of the cyst.12-14 An abdominal mass, pain and weight loss are the most common presenting symptoms. Obstructive jaundice,10 cholangitis,12 duodenal stenosis or fistula,1 acute and chronic pancreatitis,12-14 pancreatic abscess1 and pancreatic fistula due to compression and erosion into the pancreatic ducts are unusual complications of hydatid cysts involving the head of the pancreas.7 Cysts in the body and tail of the pancreas may be asymptomatic or may present as a palpable mass,1013 while mesenteric vein thrombosis or segmental portal hypertension due to splenic vein thrombosis are uncommon presentations of cysts in the body and tail.8

Spontaneous rupture into the peritoneal cavity or gastrointestinal tract and abscess formation are rare complications.12

The differential diagnosis of cystic lesions of the pancreas is extensive and includes pseudocysts, cystic neoplasms such as serous and mucinous cystadenomas,20 cystadenocarcinomas,19 cystic islet cell and papillary cystic and solid tumours,10-13 cysts which occur in association with polycystic disease or von Hippel-Lindau disease, vascular tumours (haemangioma or lymphangiomia), cystic metastases from cancer of the lung, ovary, or melanoma, rare enterogenous or lymphoepithelial cysts22 and primary hydatid disease of the pancreas. Differentiation of the various types of pancreatic cysts is important as 80% are pseudocysts but 10% are cystic neoplasms which require excision and are potentially curable after complete resection.26 Clinical presentation, absence of alcohol, gallstones, hyperlipidaemia or trauma and normal serum lipase and amylase levels should suggest a cause other than a pseudocyst.

Hydatid cysts of the pancreas are generally difficult to diagnose preoperatively and because of their rarity may be mistaken for more commonly encountered cystic pancreatic lesions. Although the presence of cystic lesions of the pancreas are easily identified by ultrasound, CT scan and magnetic resonance imaging (MRI), these methods have limited sensitivity in making a specific diagnosis because of the considerable overlap of imaging features.26 A specific diagnosis is seldom made preoperatively unless hydatid disease is suspected. Features suggestive of pancreatic hydatidosis include the epidemiological environment, the characteristic egg-shell cyst wall calcification on abdominal radiograph, the presence of daughter cysts or an undulating double lining membrane demonstrated by ultrasound, CT scan or MRI, peripheral eosinophilia and positive hydatid serology.30 The diagnosis is confirmed by demonstrating scolecis, hooklets or a parasitic membrane on cytology.

Aspiration of cyst fluid for analysis or biopsy of the cyst wall has been recommended as methods of distinguishing hydatid cysts of the pancreas from more commonly occurring pseudocysts or cystic tumours.32 However, if the lesion is malignant, percutaneous CT or ultrasound-guided needle aspiration carries the potential risk of needle tract or peritoneal dissemination of viable parasitic or neoplastic cells as well as spillage of malignant cyst contents. Intraoperative differentiation of pancreatic cysts may also be difficult. While cystic tumours are often discrete with a well-defined margin and a thin opalescent wall with a normal surrounding pancreas, pseudocysts have a thicker wall with marked inflammatory changes and may be adherent to omentum or stomach. Hydatid cysts may closely mimic either of these. Intraoperative biopsy and histological examination of biopsy material may fail to differentiate cystic lesions reliably in 20% of cases. Biopsy of the wall of a cystic neoplasm of the pancreas is unreliable because serous cystadenoma and mucinous cystic neoplasms have incomplete cyst epithelium in 40% and 72% of cases, respectively.6 Up to 90% of the cyst lining may be epithelium-free. In addition, some tumours have a mixture of cuboidal, benign columnar and malignant cells within the same lesion. Aspiration of cyst fluid for scolices was negative in 2 of our patients. These data provide a cogent argument in favour of complete excision of an unidentified cystic lesion of the pancreas. Since radiological features, cyst fluid analysis and even operative differentiation with frozen section cannot always accurately discriminate between the various cystic lesions, the only definitive way of excluding a cystic tumour is complete excision.

The current definitive treatment of hydatid disease is surgical excision.11 Provided there is no communication between the cyst cavity and the bile or pancreatic ducts, sclerosidal agents (Eusol, povidone iodine, silver nitrate, or cetrimide) are injected to sterilise the cyst contents before aspiration. However, fluid for scolices was negative in 2 of our patients. These data provide a cogent argument in favour of complete excision of an unidentified cystic lesion of the pancreas. Since radiological features, cyst fluid analysis and even operative differentiation with frozen section cannot always accurately discriminate between the various cystic lesions, the only definitive way of excluding a cystic tumour is complete excision.

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REFERENCES