Case report

Periampullary cyst: a surgically remediable cause of pancreatitis

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SUMMARY We report two patients with periampullary cysts associated with recurrent attacks of acute pancreatitis. In both patients the diagnosis was made preoperatively by upper gastrointestinal endoscopy and ERCP, which was also useful in determining the relationship of the cysts to the biliary and pancreatic ductal systems. Simple marsupialisation of the cysts resulted in long term relief of symptoms. Congenital cystic anomalies in the second part of the of the duodenum should be diligently sought in patients with pancreatitis of unexplained cause, as surgical therapy is safe and effective.

With the advent of endoscopic retrograde cholangio-pancreatography (ERCP), surgically remediable conditions such as biliary calculi, ampullary stenosis, and local pancreatic duct obstruction can be identified in up to two-thirds of patients with recurrent non-alcoholic pancreatitis.^{1,2} Endoscopic retrograde cholangiopancreatography has also contributed to the recognition of an association between relapsing pancreatitis and duodenal duplication,³⁻⁵ choledochocoele,^{6,7} and other rare structural anomalies.⁸ We report two patients who presented with recurrent attacks of acute pancreatitis caused by duodenal cysts in relation to the accessory and main papilla respectively, highlighting the delay in diagnosis of these cysts, the importance of ERCP in their assessment and the long term response to appropriate surgical management.

Case reports

PATIENT 1

A 21 year old woman was investigated for recurrent pancreatitis. She had been admitted to hospital on four occasions in the preceding two years for episodes of right upper quadrant and epigastric pain radiating to the back, relieved by adopting the 'jack-knife' posture. Serum amylase was measured on two admis-

sions and raised on both occasions (6100 and 4780 U/l respectively) (normal <300 U/l). Two ultrasonographic examinations of the upper abdomen and one barium meal had been interpreted as normal. There was no other relevant past medical history, family history or drug history and she denied alcohol intake. On examination, two weeks after her last episode of abdominal pain, the only abnormal finding was a small, non-tender, firm epigastric mass.

The serum amylase was 305 U/l and the total serum bilirubin 20 µmol/l. The transaminase and alkaline phosphatase concentrations, the serum calcium and the plasma lipids were normal. Review of the barium meal done two years previously revealed an intraluminal filling defect in the second part of the duodenum. The lesion was cystic on computed tomographic scanning (Fig. 1). At ERCP a 4×5 cm smooth cystic mass projected into the duodenal lumen immediately proximal to the major papilla (Fig. 2). Cholangiography was normal and pancreas divisum was demonstrated on pancreatography.

Laparotomy was carried out in view of the suspected association between the cyst adjacent to the ampulla of Vater and the history of relapsing pancreatitis. The pancreas and gall bladder were macroscopically normal but a longitudinal duodenotomy revealed a 3-4 cm tense cyst attached to the medial border of the second part of the duodenum just proximal to the major papilla (Fig. 3). The cyst overlay the expected site of the accessory papilla which could not be clearly identified. Aspira-

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Fig. 1 Computed tomography scan (patient 1) showing a cyst in the duodenum (marker).

tion of the cyst produced clear fluid with low amylase concentration, and contrast injection revealed no communication with the common bile duct or pancreatic duct. The cyst wall was excised except for the portion attached to the duodenal wall which was marsupialised. Postoperative recovery was uneventful. Histology of the excised wall of the cyst revealed normal duodenal mucosa on both surfaces, separated by a thin muscularis mucosa but no muscularis propria. Six months later the accessory papilla was identified at ERCP and a dorsal pancreatogram was normal. Two years after surgery the patient has experienced no further episodes of pancreatitis.

Fig. 2 Endoscopic view of second part of the duodenum at ERCP (patient 1) showing a large mass protruding into the lumen.

Fig. 3 Operative appearance (patient 1) of cyst projecting from the medial wall of the duodenum just proximal to the cannulated major papilla (arrow).

PATIENT 2

A 60 year old man was admitted for investigation of recurrent abdominal pain. For 13 years he had experienced two to three episodes per year of severe epigastric pain radiating to the back, associated with jaundice on one occasion. Abdominal ultrasonography, oral cholecystography, and upper gastrointestinal barium radiology were previously normal, but endoscopy done two weeks before his admission showed a cystic swelling in the region of the ampulla of Vater. There was no relevant family history and no drug or alcohol intake. On admission he was asymptomatic and physical examination was unremarkable. The serum amylase was 821 U/l, the total serum bilirubin 21 $\mu\text{mol/l}$ and the alkaline phosphatase, serum calcium and blood lipids were normal. At ERCP a swelling was noted bulging into the lumen of the second part of the duodenum on its medial aspect. A tiny opening on the mass was cannulated and contrast injection filled a large cystic lesion which did not appear to communicate with either the biliary or pancreatic ductal systems. It was suspected, however, that the cyst occupied the ampulla of Vater, as no other structure resembling a major papilla, could be found.

At laparotomy and duodenotomy a cyst of the

ampulla of Vater was identified (Fig. 4) and laid open. Both the common bile duct and pancreatic ducts opened into the floor of the cyst and were cannulated separately. The cyst was marsupialised and the patient made a good recovery. He has remained asymptomatic three years after surgery. Histology of the cyst wall was inadequate to determine its mucosal origin.

Discussion

Cystic lesions in the periampullary region may be due to a number of different causes, some of which carry a bewildering array of synonyms. These include enterogenous cyst of the duodenum,^{9,10} duodenal duplication,^{5,11,12} juxtampullary bile filled duodenal duplication cyst,¹³ intraluminal duodenal diverticulum,^{14,15} type 111 choledochal cyst (choledochocoele),^{6,16} diverticulum of the common bile duct¹⁷ and cyst of the ampulla of Vater.¹⁸ The terminology is further complicated by the suggestion that the intraluminal duodenal diverticulum and the enterogenous or duplication cyst are varieties of the same entity.¹⁹ Similarly it has been considered that the choledochocoele represents another form of duodenal duplication, occurring at the ampulla.²⁰

Fig. 4 Operative view (patient 2) of duodenotomy showing an ampullary cyst.

This contention is supported by the observation that duodenal rather than biliary tract mucosa lines the majority of choledochocoeles.²¹

Our patients amply illustrate the difficulties encountered in placing duodenal wall cysts into precise diagnostic categories. Patient 1 does not fulfil the criteria for a duplication cyst²² in view of the absence of a circular muscle layer within the cyst wall. Furthermore the cyst did not fill with barium and cannot therefore be classified as an intraluminal duodenal diverticulum.²³ A precise diagnosis is even more difficult in patient 2 whose lesion could represent a duplication cyst or diverticulum occurring at the ampulla of Vater, or even a choledochocoele. Therefore, in the absence of specific diagnostic features, we have simply termed the lesions 'periampullary cysts'.

The clinical presentation, investigation, differential diagnosis and management of duodenal wall cysts have recently been reviewed.³ Relapsing pancreatitis as exemplified by the patients in the present report, is uncommon but well described in association with duodenal duplication cyst^{3,4} intraluminal duodenal diverticulum^{14,15} and choledochocoele.^{6,16} A recent review⁴ of eight previously reported cases of acute pancreatitis associated with duodenal duplication cysts suggests that pancreatitis results from obstruction of the ampulla of Vater caused by intermittent distension of the duplication, or to duodenal obstruction. Pancreatitis and jaundice in patient 2 of the present report may have been the consequence of an intermittent increase in pressure within the ampullary cyst resulting in ductal stasis because of obstruction or distortion.

The association of periampullary cyst with pancreas divisum (patient 1) has not previously been documented. The absence of further attacks of pancreatitis after surgical treatment of the cyst related to the accessory duct opening suggests that the latter, rather than the pancreas divisum²⁴ was responsible for recurrent pancreatitis. Periampullary cysts are treated by surgery. In all cases a clear preoperative definition of the relationship between the cyst and the biliary and pancreatic ducts must be obtained, and is best achieved by ERCP. Although various surgical approaches have been used³ the procedure currently recommended is internal drainage of the cyst into the duodenum^{3,5} a simple, safe, and effective operation.

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