Occult carcinoma in an adult choledochal cyst

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Summary: The complications of choledochal cyst are avoidable if diagnosed early, and adequate resection undertaken. This case report describes the long history of right subcostal pain in a young man of 26 who had a squamous carcinoma in a choledochal cyst diagnosed after serial section of the excised cyst. Subsequent resection of the head of the pancreas showed histological residual tumour from which he died 4 months later. This case illustrates the need for complete early excision of a choledochal cyst to prevent this complication.

Introduction

Focal dilatation of the biliary tree consistent with the modern concept of choledochal cyst was first described by Douglas in 1852,¹ although Vater may have described a case in the previous century.² The disease remained difficult to define and categorize until the classification of Todani *et al.*³ based on the earlier work of Alons-Lej *et al.*⁴ The importance of the condition lies in the complications,^{5,6} of which carcinoma is well described. The incidence of carcinoma is 2.4–14%,^{5,7–11} but varies considerably with age, being 0.7% in those less than 10 years of age, 6.8% between the age of 10 and 20 years and 14.3% over the age of 20.¹²

This case report is presented in order to draw attention to the diagnostic difficulties of the condition, and the rapid fatality which can occur even after radical treatment.

Case report

A 26 year old white male presented in July 1988 with a one week history of acute cholangitis, with severe right upper quadrant and posterior scapular pain, jaundice, fever and rigors. The pain was partially relieved by vomiting and he had lost 1–2 kg in weight. Similar but shorter attacks (1–2 days) of identical but less severe pain had occurred every 2–4 months since the age of 14 but had never previously been associated with fever or jaundice. There was no other personal or family history of note. Examination revealed a temperature of 38.5°C, marked right upper quadrant tenderness

with voluntary guarding and mild scleral icterus. The haemoglobin was 14.5 g/dl, white cell count $7.0 \times 10^9/l$, bilirubin 252 $\mu\text{mol/l}$ (NR: 3–17), alkaline phosphatase 828 IU/l (NR: 100–280), albumin 42 g/l (NR: 35–53), aspartate transaminase 559 IU/l (NR: 5–40), calcium 2.42 mmol/l (NR: 2.2–2.6) and urea 4.8 mmol/l (NR: 3.0–8.0). Serum amylase was normal. Ultrasound and computed tomographic (CT) scan (Figure 1a) showed cystic dilatation of the common bile and hepatic ducts and the distal left and right hepatic ducts with mildly thickened walls; there was no dilatation of the proximal intrahepatic ducts and the liver echogenicity was normal. The gallbladder was not seen. The cyst appeared to extend deep into the posterior pancreatic substance down to the papilla. There were no filling defects seen within the cyst. A Todani Type 1a choledochal cyst was diagnosed. The cholangitis settled progressively with intravenous antibiotics but the pain continued constantly with added intermittent severe attacks requiring parenteral narcotics. Preoperative endoscopic retrograde cholangiopancreatography (ERCP) under antibiotic cover confirmed the presence of a choledochal cyst and documented a short intraduodenal common channel (3 mm) and narrow irregular intrapancreatic bile duct (Figure 1b). The papilla and pancreatic duct were normal. There appeared to be multiple small filling defects within the cyst suggestive of cystolithiasis though cyst filling at ERCP was minimized to prevent cholangitis.

At operation, the cyst and gallbladder were excised and a roux-en-Y hepaticojejunostomy fashioned. Postoperative recovery was complicated briefly by a right basal pneumonia. Histopathological examination of the specimen, however, revealed an area of adenocarcinoma with squamous differentiation at the distal end of the cyst (see

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below). Pancreatoduodenectomy with the excision of the remaining bile duct was therefore undertaken. This was complicated by recurrent right basal pneumonia despite epidural analgesia, and *Enterobacter* sp. bacteraemia and a transient bile leak. The patient left hospital well after a total of 6 weeks' stay but represented with malignant ascites and severe pain 4 months later and died from carcinomatosis.

(b)(ii)

Figure 1 (a) CT scan showing large thin-walled choledochal cyst. (b) ERCP showing the choledochal cyst (straight black arrow), the 2–3 mm common pancreatobiliary channel (white arrow) and the irregular, abnormal low common bile duct (curved black arrow).

Histopathology

The resected cyst specimen was $9 \times 6 \times 5$ cm and had a 2 mm thick fibrous wall. It contained bile-stained fluid but no stones and had a macroscopically smooth lining. The gallbladder ($9 \times 2 \times 2$ cm) opened via a short straight cystic duct into the proximal cyst and contained no stones. The specimen was extensively sampled, especially within its immediate supra- and retropancreatic portion. One section showed the full thickness of the wall infiltrated by moderate to poorly differentiated adenocarcinoma with some foci of squamous differentiation (Figure 2). The presence of severe epithelial atypia of the choledochal cyst lining adjacent to the infiltrating carcinoma suggested the origin of the tumour from the cyst rather than infiltration in from the adjacent pancreas. Immunocytochemically there was no carcinoembryonic antigen production. Acute and chronic inflammation of the cyst mucosa was present throughout the entire cyst.

The pancreatoduodenectomy specimen showed the changes of recent surgery and an enlarged 1 cm node between the head of the pancreas and the wall of the duodenum. Microscopically, residual

Figure 2 Histopathological specimen showing adenocarcinoma with squamous metaplasia (H&E; magnification $\times 230$).

tumour was present within the wall of the operation wound in the posterior pancreatic head. The enlarged lymph node, detected macroscopically, and a further node were involved by metastatic carcinoma. Perineural invasion of a large degenerated nerve was also seen. No carcinoma was seen at the resection margins. The non-directly involved pancreatic head and neck were histologically normal.

Cytological examination of the ascitic fluid at representation 4 months later showed typical adenocarcinoma cells.

Discussion

Choledochal cyst is uncommon in the United Kingdom.¹³ It is an anomaly which may present at any age with the classical triad of pain, abdominal mass and jaundice, although in the adult, recurrent abdominal pain alone is the commonest presenting feature,^{5,6,14} requiring an awareness of this possibility by the ultrasonographer screening a patient with upper abdominal pain. Presentation in adult-

hood is usually associated with the development of complications such as choledocholithiasis, portal hypertension due to secondary biliary cirrhosis and carcinoma.¹⁵ The presentation with these complications is the rationale for the treatment of a choledochal cyst by excision of the cyst as opposed to simple drainage.

Neoplasm is a well-recognized complication of all forms of choledochal cysts^{11,16,17} and was first reported in Type I cysts by Irwin and Morrison in 1944,¹⁸ Type V cysts in 1968¹⁹ and in choledochoceles in 1980.¹¹ This complication is most commonly seen in Types I and IV cysts, that is cysts involving the common bile duct. In these patients a distended gallbladder may be a good clinical guide to the presence of a neoplasm.²⁰ Adenocarcinoma is by far the commonest histological type, but squamous, adenosquamous, anaplastic and sarcomatous varieties have been described.^{11,16,18,20}

The reason for the development of neoplasms in the choledochal cyst is unknown but in this and other such cases acute and chronic inflammation within the cyst is common, probably as a result of stasis with infection and the reflux of pancreatic juice into the cyst as a result of the common channel between the bile duct and the pancreatic duct.^{21,22} The long history in the present case undoubtedly predisposed to the development of the carcinoma and the difficulty in diagnosing the condition despite careful preoperative investigation emphasizes the care required in the management of this rare anomaly. Further, routine histology failed to detect the lesion, and it was not until serial section of the entire cyst had been undertaken that the true diagnosis was apparent. However, the general experience with neoplasm in cysts is that cure is rare and the short term prognosis of even apparently resected tumours is poor.^{9,10,13,15} The only hope is early cyst detection and resection⁷ as only one patient has developed neoplasia after resection.⁸ Nevertheless, resection will not eliminate the risk completely as neoplasia may arise outside the cyst in 40% of cases.

The complications of choledochal cyst are life threatening; drainage does not prevent these complications, but it is probable that early diagnosis with complete excision of the cyst down to its junction with the pancreatic duct is the minimal surgical management. Complete diagnosis requires careful imaging with the awareness of the non-specific nature of the early symptoms, and surgical treatment requires specialist surgery to ensure that prophylactic treatment has been undertaken for it is probable that once carcinoma has developed with its early lymphatic and perineural invasion, cure is unlikely by even the most radical clearance.

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