

CLINICAL NOTES

Type II intrapancreatic choledochal malignant cyst in adults: Duodenopancreatectomy

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ABSTRACT

A 62-year-old female patient was admitted for abdominal pain and vomiting. Imaging tests revealed a solid-cystic lesion at the head of the pancreas communicating with the distal bile duct. A Todani type II choledochal cyst was diagnosed with neoplastic degeneration after cytological diagnosis with endoscopic ultrasound-guided puncture. The patient was treated with a cephalic duodenopancreatectomy with curative intention.

Key words: Choledochal cyst. Cancer. Pancreatoduodenectomy.

INTRODUCTION

Choledochal cysts are a very rare entity in our environment, with an incidence of one in every 2 million. They are more common in females (1-4) and have a higher incidence in Asia (1 in every 150,000 live births).

The classification of choledochal cysts was initially reported by Alonso-Lej (5) in 1959. Today the most commonly used classification is that of Todani et al. (1), which establishes 6 types of choledochal cyst (Table I).

The most frequent types of cyst are I and IV, which make up 80 % of the total (1,4,5); the other types account

for less than 5 % each (4,6). The diverticular type, or type II, accounts for 2-3 % of cases (7). Few patients are diagnosed in adulthood and the risk of complications, mainly malignancy, increases with age (7,8).

We report a case of intrahepatic location and proven malignancy which required a cephalic duodenopancreatectomy.

CASE REPORT

A 62-year-old woman with no previous history of interest was admitted to emergencies for symptoms of abdominal pain. Laboratory tests including a haemogram, liver function tests (ALT, AST, bilirubin and alkaline phosphatase) and tumour markers (CEA and CA 19.9) were normal.

During admission, she underwent abdominal ultrasonography, which revealed a mass with a solid-cystic echo structure, some 4 cm in maximum diameter and intimately related to the common bile duct via a thin pathway (Figs. 1 and 2). The bile duct had a normal calibre, no choledocholithiasis and a slightly dilated main pancreatic duct. The study was completed with a cholangio-NMR (Fig. 3) and abdominal NMR (Fig. 4), with and without i.v. contrast,

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Table I. Todani classification

I	Saccular dilatation of the extrahepatic bile duct
II	Bile duct diverticulum
III	Dilatation of the intraduodenal portion of the bile duct
IV	Multiple cysts of both the intra- y extrahepatic ducts (type A), or multiple cysts involving just the extrahepatic duct (type B)
V	Intrahepatic cysts or Caroli disease
VI	Cystic dilatation of the cystic duct

which confirmed the previous findings of a Todani type II choledochal cyst with probable tumour degeneration.

The thoracic-abdominal-pelvic CT extension study with i.v. contrast did not detect adenopathies or distant metastases. Endoscopic ultrasound-guided fine-needle aspiration was performed. The cytopathological study of the material obtained showed abundant epithelial cellularity, which was atypical and generally arranged in three-dimensional clusters, variably cohesive and often adopting a papillaroid

pattern. These were findings compatible with a diagnosis of adenocarcinoma.

With a diagnosis of intracystic neoplastic degeneration we decided on surgical treatment. A midline supra- and infraumbilical laparotomy was performed. No adenopathies or other signs of abdominal neoplastic spread were encountered during the operation. The intrapancreatic choledochal cyst was revealed with a posterior wall intimately related to, but not infiltrating, the superior mesenteric vein. A Whipple cephalic duodenopancreatectomy was performed, which was incident-free. After presenting with severe infection of the central catheter during the postoperative period the patient was discharged at 20 days.

Macroscopic examination of the surgical specimen disclosed a 3.5 cm unilocular cyst formation in the midst of the pancreatic tissue, revealing several polypoid formations at the wall, which were villous in appearance and the largest of which had a maximum diameter of approximately 2.2 cm. The microscopic study showed epithelium with a biliary coating and variable degrees of dysplasia (biliary intraepithelial neoplasia, BilIN), often a high degree (BilIN 3). Additionally, stromal microinfiltration foci were observed. No lymphovascular invasion or perineural infiltration was detected; neither was there involvement of adjacent structures (pancreatic parenchyma, duodenum) or isolated lymph nodes.

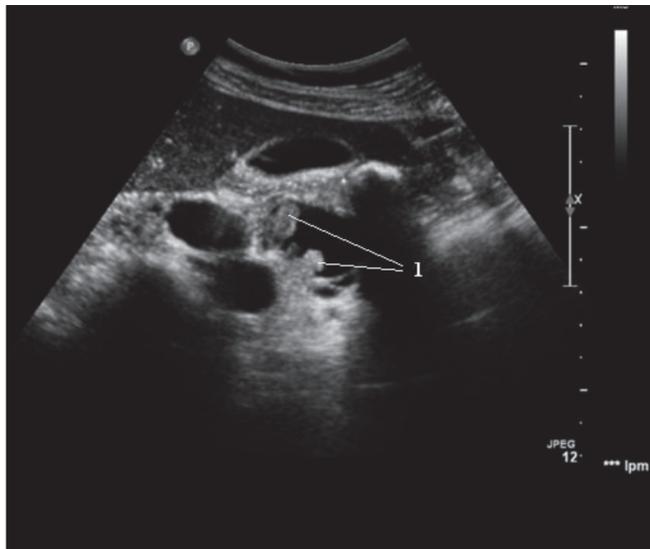


Fig. 1. Contrast-enhanced abdominal ultrasonography (arterial phase) centred on the head of the pancreas. Note the solid-cystic lesion at the head of the pancreas showing vascularisation and enhancement of the solid intracystic papillary projections (1).

DISCUSSION

Choledochal cysts are a very rare pathology in our environment and are mostly diagnosed in childhood or adolescence (only 20-30 % in adults).

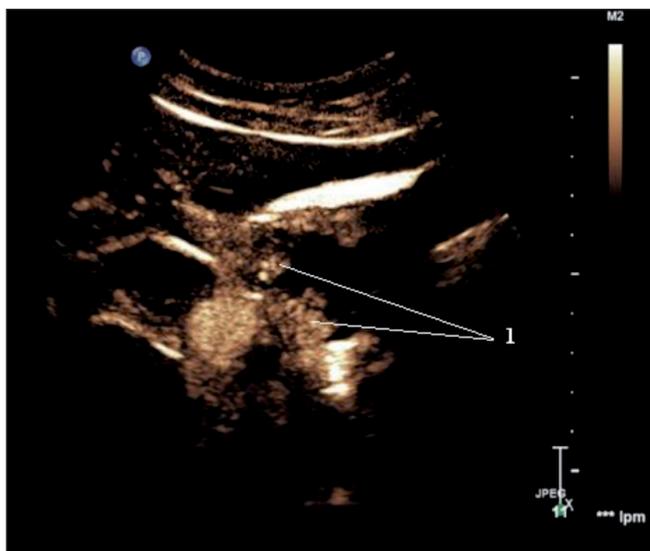


Fig. 2. Echoendoscopy showing fine-needle puncture of the internal solid papillary projections (1).

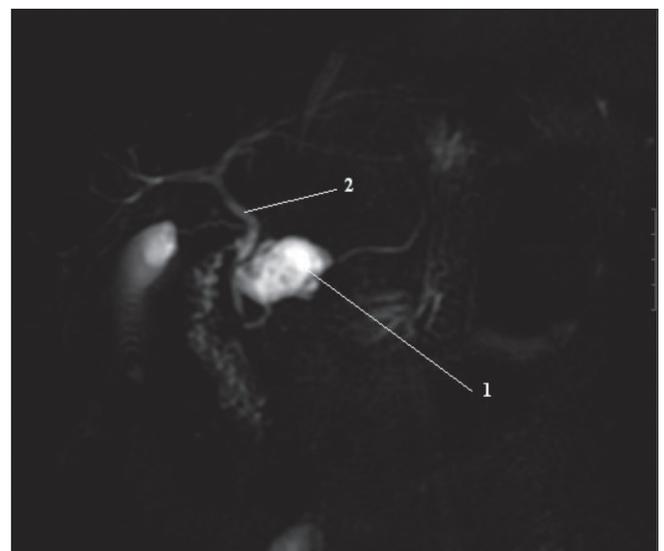


Fig. 3. Oblique coronal plane of a T2-weighted cholangio-MR sequence showing the choledochal cyst (1) and the intra- and extrahepatic bile duct (2).

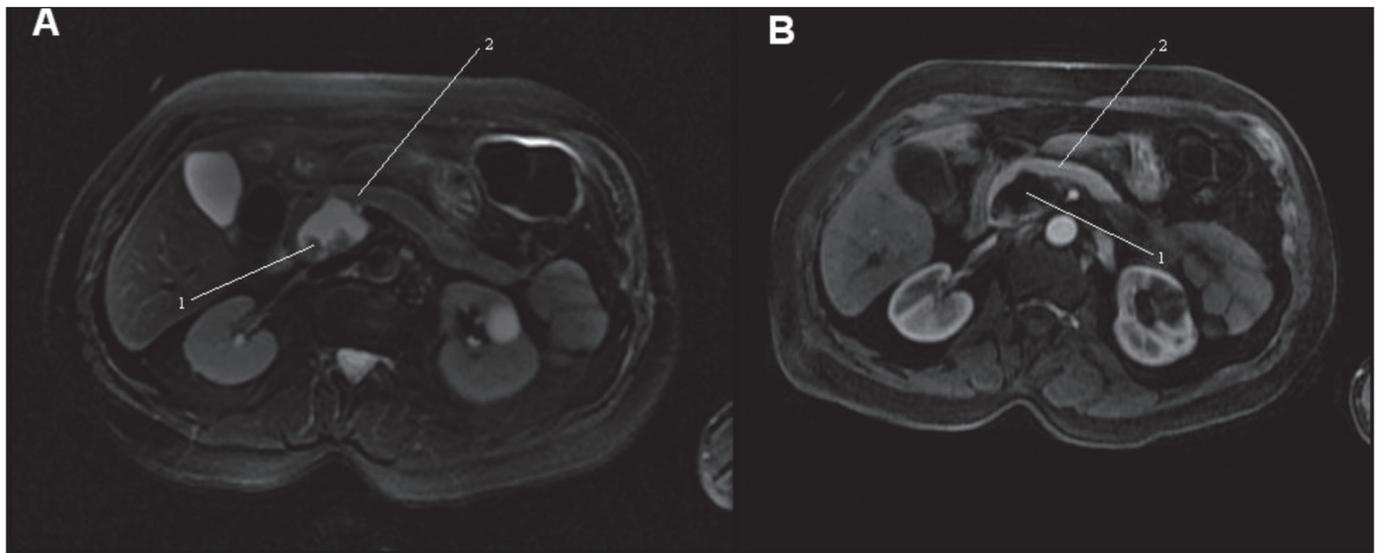


Fig. 4. NMR axial sections: A. T2-weighted sequence with fat suppression showing the cyst below the pancreatic head (2) and the intracystic papillary projections (1). B. T1-weighted 3D gradient echo sequence with fat suppression and intravenous contrast in the arterial phase showing the choledochal cyst (1) below the head of the pancreas (2).

The most common symptoms are abdominal pain in the right hypochondrium and jaundice. Adults usually have less florid symptoms, which implies a late diagnosis (5). The classic triad of abdominal pain, jaundice and abdominal tumour presented in 85 % of paediatric patients is a rarity in adults.

Diagnosis of choledochal cyst is established by imaging techniques. Ultrasound and CT usually show a cystic lesion, independent of the gallbladder, located in the subhepatic space between the duodenum and the portal confluence (4,5). As an initial imaging test ultrasound is a useful technique for assessing the biliopancreatic tree, gallbladder and pancreas. Moreover, contrast-enhanced ultrasonography provides real-time information on tumour perfusion and is useful, as in our case, for demonstrating uptake by the small intracystic papillary projections.

Cholangio-NMR is the method of choice for completely mapping the bile duct. It has a high sensitivity and specificity: some 90-100 % and 73-100 % respectively (4). In our case, it enabled the cyst to be characterised.

Malignant transformation is the most serious complication and has a very poor prognosis for survival. It occurs in less than 1 % of paediatric patients and in 10-30 % of adults; it can even reach 50 % in patients aged over 50 years following internal drainage as their only treatment (1,3,8,9).

Todani type I seems to be the most commonly associated with cancer, followed by type IV. The most common site for development of the carcinoma is the wall of the cyst, although it can occur at any level of the biliary tree.

This high probability of complications and malignant degeneration justifies surgery as elective treatment. The

best method is complete resection with a biliodigestive anastomosis; simple drainage of the cyst without complete resection is associated with the development of complications and does not reduce the malignancy potential of the disease. Complete resection of the extrahepatic bile duct, together with cholecystectomy, considerably reduces the risk of neoplastic degeneration, which drops to 0.7 % (4).

Bismuth and Krissat (7) found that seven of every eight patients with cancer associated with choledochal cysts had previously been given expectant management with internal drainage or without complete resection of the cyst, which highlights the need to select an adequate treatment once the cyst is diagnosed.

If neoplastic degeneration is shown to affect the distal portion of the bile duct, the usual techniques for treating choledochal cysts, with excision of the cyst or the whole bile duct (depending on the case) and association of a hepaticojejunostomy, are no longer valid (13,14). Metastatic disease must be ruled out initially, as well the absence of infiltration to important adjacent structures such as the portal vein, and surgery must be performed with a curative intention (R0). A Whipple cephalic duodenopancreatectomy with a sufficient safety margin is the technique of choice in cases suspected with neoplastic degeneration of the cyst (12).

Curative resections are associated with a 5-year survival rate of 25-50 %, the primary determinants of these poor results being positive margins and lymph node involvement (11). It is also important to mention the patchy nature of these lesions, as they can occur at other levels of the biliary tree and require appropriate clinical and radiological follow-up.

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