Choledochal Cyst Cholangiocarcinoma Arising from Adenoma: Case Report and a Review of the Literature

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A case of cholangiocarcinoma arising in an unresected choledochal cyst in adulthood is presented. Although typically diagnosed in pediatric population, as many as 20% to 30% of choledochal cysts can be discovered in adulthood. Unresected choledochal cyst is clearly associated with increased risk of cholangiocarcinoma. Proper surgical treatment includes cyst resection and bilioenteric anastomosis. Asymptomatic patients with choledochal cyst previously treated by biliary diversion without cyst resection present a challenging issue and should be considered for cyst resection. Association of choledochal cyst with pancreaticobiliary anomalies is reviewed. (Curr Surg 63: 281-284. © 2006 by the Association of Program Directors in Surgery.)

KEY WORDS: biliary cyst, cholangiocarcinoma, biliary obstruction, jaundice

INTRODUCTION

In this report, choledochal cyst carcinoma is described as an uncommon etiology of jaundice in adulthood. Although incidence is low in the United States, cases can be encountered by general surgeons occasionally. Estimated incidence in Western population is 1 in 15 000 live births,[1] and approximately 20% to 30% are first detected in adulthood. This condition is more frequently encountered in Asia,[2] where most experience was gained.[2-4]

Symptoms most commonly associated with choledochal cysts include right upper quadrant pain, jaundice, and cholangitis. Painless jaundice can be the presenting complaint in previously asymptomatic patients with advanced malignancy.

CASE REPORT

A 57-year-old Caucasian woman presented with 1-week history of painless jaundice. No fever and no weight loss were reported, and the patient was in no apparent distress. Surgical history included Roux-en-Y choledochojejunal anastomosis for a choledochal cyst that was performed in adolescence, and both choledochal cyst and a gallbladder were left in situ.

On physical examination, a non-tender indistinct mass was discovered in the right upper quadrant. Laboratory results included total bilirubin of 8.5 mg/dl, alanine aminotransferase of 100 IU/l, aspartate aminotransferase of 66 IU/l, and alkaline phosphatase of 566 IU/l. Ultrasonography revealed marked dilation of the biliary tree and a normal gallbladder without evidence of cholelithiasis. Magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreaticography (MCRP) identified a choledochal cyst measuring $7 \times 3.5 \times 5$ cm with a mass adjacent to its superior aspect (Figs. 1 and 2). Preoperative percutaneous transhepatic cholangiography demonstrated complete occlusion of common hepatic duct secondary to the tumor (Fig. 3). During laparotomy, both choledochal cyst with tumor and old Roux-en-Y hepaticojejunostomy were resected. Proximal resection margin was at the level of common hepatic duct bifurcation, where the cystic process terminated. The distal resection margin was located in the area of normal-appearing terminal common bile duct, and this was carefully dissected from the dorsal aspect of the pancreas avoiding an injury to pancreatic duct. A new Roux-en-Y hepaticojejunoanastomosis connecting the common hepatic duct bifurcation to the jejunal limb was constructed.

On pathologic evaluation, the size of the choledochal cyst was $9 \times 5.5 \times 5$ cm and its wall thickness was 0.3 cm. Approximately one half of its surface was lined with shaggy papillary pink mucosa. Beneath this mucosal layer, an infiltrating white mass was identified. Microscopy revealed well-differentiated infiltrating adenocarcinoma arising in predominantly villous adenoma. The tumor was confined to choledochal cyst, and no nodal involvement was identified in a total of 5 resected lymph nodes. Signs of perineural, venous, and lymphatic invasion were absent.

Postoperatively, the patient experienced a prolonged course with a subhepatic biliary abscess treated with temporary percutaneous drainage and intravenous antibiotics. She recovered

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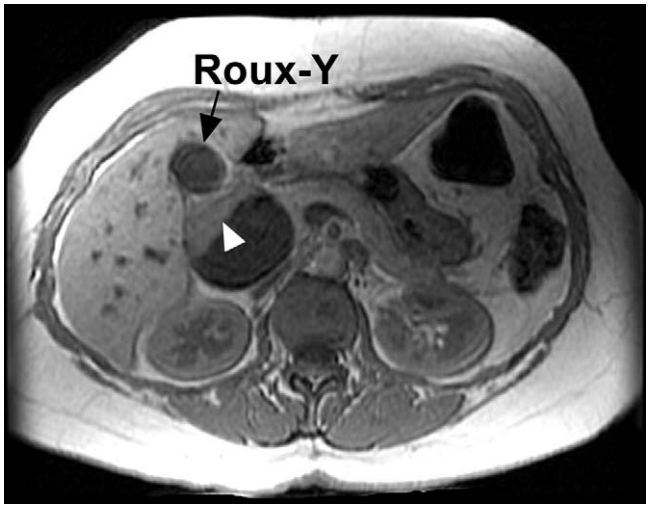

FIGURE 1. Magnetic resonance image of upper abdomen, axial image. Choledochal cyst with tumor (arrowhead) and Roux-en-Y limb of original anastomosis are demonstrated.

and is periodically observed for follow-up care. So far she is asymptomatic and her liver function tests are normal 1 year after operation.

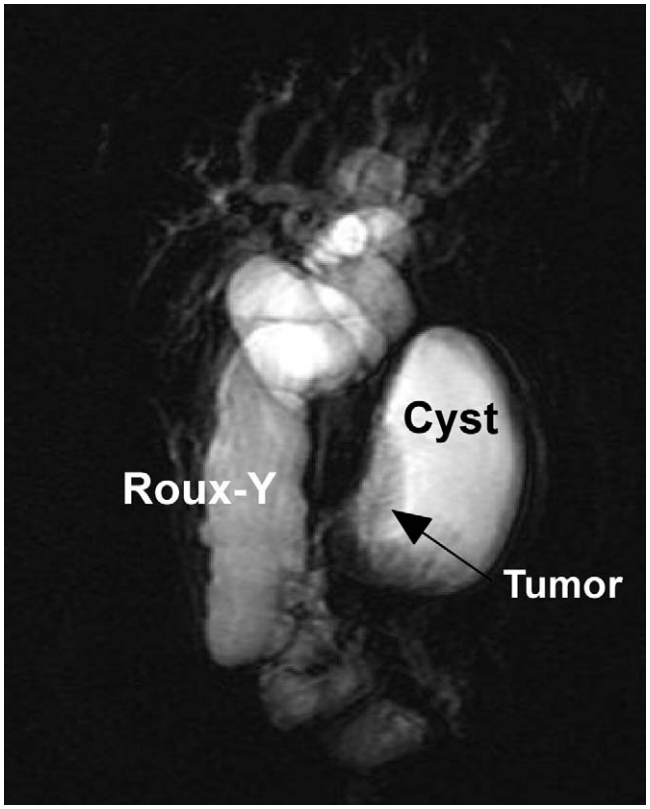

FIGURE 2. Magnetic resonance cholangiopancreaticography, reconstruction of biliary anatomy. Choledochal cyst measuring approximately 7 × 3.5 × 5 cm is identified inferiorly to the tapering of the inferior portion of the common hepatic duct with evidence of an adjacent mass (arrow). The communication with the choledochal cyst is not defined.

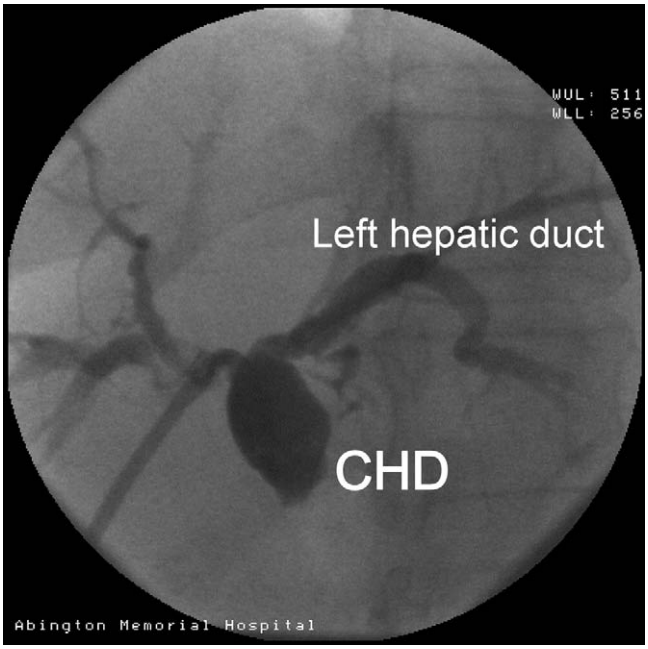

FIGURE 3. Preoperative percutanneous transhepatic cholangiogram. There is filling of the right and left hepatic ducts. Complete occlusion of the common hepatic duct (CHD) just distal to the bifurcation. There is no filling of the common bile duct or cystic duct.

DISCUSSION

Although choledochal cysts are an uncommon cause of biliary tract symptoms in adults, they constitute an important cause of jaundice in the pediatric population. The incidence has been well documented in the Asian population at 0.32%,[2] where it seems to be higher than in the Western population. There is a female predominance. Estimated incidence in Western population is 1 in 15 000 live births.[1] Although most cysts are diagnosed in children, some 20% to 30% are first detected in adults as they either become symptomatic or are incidentally found during imaging for an unrelated cause.[5]

Extrahepatic bile duct cysts were first classified by Alenso-Lej et al in 1959,[6] and revised by Todani et al[7] in 1977 with the addition of 2 types to describe intrahepatic cystic dilation (Table 1).

Radiologic diagnosis is usually made by ultrasound or computer tomography. Both delineate presence of a cyst; however,

TABLE 1. Classification of Biliary Tree Cysts[6,7]

Type	Morphologic Description
I	Cystic dilatation of the common bile duct
II	True diverticulum of the extrahepatic bile duct
III	Choledochocele (cystic degeneration limited to the intraduodenal portion of the distal common bile duct)
IV	Multiple extrahepatic biliary cysts. Type IVA (associated intrahepatic cysts) and type IVB (extrahepatic cysts only)
V	Caroli's disease. Cysts of intrahepatic biliary tree, extrahepatic biliary system normal.