CASE PRESENTATION

A 52-year-old male with no significant medical history was evaluated for acute onset of right upper quadrant pain. The pain was intermittent, sharp, and radiated to the back; there was no associated nausea or vomiting. He reported three similar episodes of pain in the past 10 days and also recalled experiencing significant abdominal pain as a child which was never evaluated. He denied any hematemesis, melena or any significant weight loss. He never smoked or used illegal drugs, but admitted to drinking one glass of wine daily.

At presentation, he was afebrile with a blood pressure of 100/71 and a heart rate of 63. His physical examination was remarkable only for moderate right upper quadrant tenderness without any rebound, hepatosplenomegaly or palpable mass. His initial laboratory values showed a normal hemogram with WBC 3.83 × 10^6/L. His liver function tests were remarkable for elevated AST (514 IU/L), ALT (474 IU/L), alkaline phosphatase (181 IU/L), and bilirubin (1.38 mg/dl). A chest radiograph was normal. A CT scan of the abdomen showed a fusiform dilatation of the CBD up to 4.6 cm in maximum transverse diameter without any associated intrahepatic biliary ductal dilatation suggesting a type 1 choledochal cyst (Figure 1). Soft tissue nodules were identified along the dependent and nondependent surfaces of the duct lumen. A 1.9 cm × 1.6 cm portacaval lymph node was also identified. The gallbladder size was normal. Pancreatic ducts appeared normal. He underwent resection of the entire extrahepatic biliary tree along with hepatico-jejunostomy with an uneventful post-surgical course. Histopathology of the resected specimen showed invasive cholangiocarcinoma without any metastasis (Figure 2). The resected enlarged lymph node demonstrated preserved architecture with reactive hyperplasia.

Choledochal cysts are generally classified using the Todani modification of the Alonso-Lej classification. Within this classification, there are five types of...
A CASE REPORT

Congenital Type 1 Choledochal Cyst

Patients with choledochal cysts have a substantial risk of cholangiocarcinoma in comparison with the general population. The incidence has been estimated to be as high as 75% in collected series from Japan in older age groups. The risk of developing cholangiocarcinoma has been reported to be greatest in patients with intrahepatic biliary involvement (types IV and V). Extrahepatic cystic involvement is best treated by excision of the entire extrahepatic biliary tree and with subsequent biliary enteric anastomosis; this appears to eliminate the risk of cholangiocarcinoma entirely. For patients with types IV and V cyst disease, liver transplantation is the only potential means of eliminating the risk of cholangiocarcinoma.

References

Figure 2. Pathology of the resected specimen of bile duct: Invasive cholangiocarcinoma extending through the muscular wall into the surrounding fibroadipose tissue.

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Send in a brief case report. No more than one double-spaced page. One or two illustrations, up to four questions and answers and a three-quarter to one-page discussion of the case. Case to include no more than two authors. A $100.00 honorarium will be paid per publication.

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