Radiographic Quiz

Answer to Radiographic quiz

Radiographic findings
Fusiform dilatation of the entire common bile duct and both common hepatic bile ducts. No evidence of obstruction or filling defects with free passage of the contrast to the duodenum.

Diagnosis
*Type I Choledochal cyst*

OVERVIEW OF THE DISEASE

Choledochal cysts
Choledochal cysts are unusual congenital anomalies of the bile ducts. They consist of cystic dilatations of the extrahepatic biliary tree, intrahepatic biliary radicles, or both. Alonso-Lej et al provided the first systematic description of choledochal cysts in 1959 based on the clinical and anatomic findings in 96 cases. The resultant system classified choledochal cysts into 3 types and outlined therapeutic strategies.

ANATOMY
The following discussion of the pertinent anatomy of choledochal cysts is based on the Todani classification.

- **Type I choledochal cysts** are most common and represent 80-90% of the lesions. Type I cysts are dilatations of the entire common hepatic and common bile ducts or segments of each. They can be saccular or fusiform in configuration. Type I cysts can be divided into 3 subclassifications, including type IA cysts, which are typically saccular and involve the entire extrahepatic bile duct (common hepatic duct plus common bile duct) or the major portion of the duct.

- **Type II choledochal cysts** (2%) are relatively isolated protrusions or diverticula that project from the common bile duct wall. They may be sessile or may be connected to the common bile duct by a narrow stalk.

- **Type III choledochal cysts** (1.5 – 5%) are found in the intraduodenal portion of the common bile duct. Another term used for these cysts is choledochocele.
for each. The classification system for choledochal cysts currently includes 5 major types (1). (See diagrams below.)

PATHOPHYSIOLOGY
The pathogenesis of choledochal cysts is most likely multifactorial. Some aspects of the disease are consistent with a congenital etiology, others with a congenital predisposition to acquiring the disease under the right conditions.

The vast majority of patients with choledochal cysts have an anomalous junction of the common bile duct with the pancreatic duct (anomalous pancreato-biliary junction (APBJ)). An APBJ is characterized when the pancreatic duct enters the common bile duct 1 cm or more proximal to where the common bile duct reaches the ampulla of Vater. Miyano and Yamataka have demonstrated such APBJs in more than 90% of their patients with choledochal cysts.

The APBJ allows pancreatic secretions and enzymes to reflux into the common bile duct. In the relatively alkaline conditions found in the common bile duct, pancreatic proteases can become activated. This results in inflammation and weakening of the bile duct wall. Severe damage may result in complete denuding of the common bile duct mucosa. From a congenital standpoint, defects in epithelialization and recanalization of the developing bile ducts during organogenesis and congenital weakness of the duct wall have also been implicated. The result is the formation of a choledochal cyst.

CLINICAL MANIFESTATIONS
Infants commonly present with conjugated hyperbilirubinemia, failure to thrive or abdominal mass. In patients older than 2 years, chronic intermittent abdominal pain is the presenting symptom 50-96% of the time and intermittent jaundice 34-55% of the time. They are 70–80% more frequent in women than men. Typical presentation: recurrent right upper quadrant pain, jaundice, and/or palpable mass.

They may be complicated by stone formation, cholangitis, pancreatitis, or rupture.

Type I choledochal cysts are often associated with recurrent pancreatitis and eventually cholangiocarcinoma. Rarely, they can show malignant degeneration. Cholangiocarcinoma develops in up to 7% of cases.

They can be associated with gallbladder aplasia, double gallbladder, annular pancreas, and biliary.

RADIOLOGICAL WORKUP AND TYPICAL FINDINGS:
Conventional Radiography:
Findings: Plain abdominal radiographs are of little use in the diagnosis of choledochal cysts. They offer no specific information related to this diagnosis. In patients presenting with abdominal pain, radiographs are frequently ordered as part of the standard workup. At most, radiographs may suggest displacement of an adjacent hollow viscus, such as the duodenum, by a mass.

Computed Tomography:
Findings: Abdominal CT scanning is useful in the diagnostic algorithm for choledochal cysts. CT is highly accurate and offers a great deal of information that is helpful, not only in confirming the diagnosis but also in planning surgical approaches.

CT scans of a choledochal cyst demonstrate a dilated cystic mass with clearly defined walls, which is separate from the gallbladder. The fact that this mass arises from or actually is the extrahepatic bile duct usually is clear from its location and its relationships to surrounding structures. The cyst is typically filled with bile, which produces water-like attenuation. Depending on the patient's age and clinical history, the wall of the cyst can appear thickened, especially if multiple episodes of inflammation and cholan-
therapy for type III choledochoceles, or choledochoceles, depends on the size of the lesion. Choledochoceles with a diameter of 3 cm or smaller may be approached endoscopically and effectively treated by means of sphincterotomy. Choledochoceles larger than 3 cm in diameter are often associated with some degree of duodenal obstruction. These cysts are excised surgically by using a transduodenal approach. If the pancreatic duct is found to be entering the choledochocele, it must be reimplanted into the duodenum after the cyst is excised.

For type IV choledochoceles, the dilated extrahepatic duct is completely excised, and a Roux-en-Y biliary-enteric anastomosis procedure is performed. No therapy is specifically directed at the intrahepatic ductal disease, except if intrahepatic ductal strictures, hepatolithiasis, or hepatic abscesses are present. In these patients, interventional radiologic techniques can be performed. If the disease is limited to specific hepatic segments or a lobe, these may be resected. A type V choledochocele, or Caroli disease, is defined only by the dilatation of the intrahepatic ducts. If dilatation is limited to a single hepatic lobe, usually the left, the affected lobe is resected. Patients who have bilobar disease and signs of biliary cirrhosis, portal hypertension, or liver failure may be candidates for liver transplantation. Patients with cholecystochal cysts require lifelong follow-up monitoring. They remain at increased risk for development of cholangiocarcinoma, even after complete excision of the cyst.

References:
1. Images from e-medicine Available at: http://www.emedicine.com/RADIO/topic161.htm