Recurrent Pyogenic Cholangitis

April 05, 2012
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A 23-year-old male with alcohol consumption with elevated liver enzymes. Ultrasound shows intrahepatic ductal dilatation involving Seg 6 RLL with multiple echogenicity with posterior shadowing represent calculi.

A 23-year-old male with alcohol consumption with elevated liver enzymes.

Ultrasound shows intrahepatic ductal dilatation involving Seg 6 RLL with multiple echogenicity with posterior shadowing represent calculi. The findings are stable since previous exam four years before.
Ultrasound shows intrahepatic ductal dilatation involving Seg 6 RLL with multiple echogenicity with posterior shadowing represent calculi, GB shows adenomyomatosis. CBD is not dilated.
MRI two months after last ultrasound — Magnetic resonance cholangiopancreatography (MRCP) shows intrahepatic ductal dilatation and structures involving Seg 6 RLL with multiple intraductal and CBD stones.

ERCP, recurrent oriental (pyogenic) cholangitis involving posterior segment of right lobe of liver with intra hepatic and CBD stones.
ERCP, recurrent oriental (pyogenic) cholangitis involving posterior segment of right lobe of liver with intra hepatic and CBD stones. CBD stones removed.

Cholangiohepatitis, recurrent pyogenic cholangitis (RPC), or oriental cholangitis is a poorly understood syndrome consisting of intrahepatic pigment stone formation with chronically recurrent exacerbations and remissions. It is endemic to Asia.

Cholangiohepatitis, or recurrent pyogenic cholangitis (RPC), is characterized by a recurrent syndrome of bacterial cholangitis that occurs in association with intrahepatic pigment stone formation, hepatic abscesses, dilatation and structuring of the intrahepatic bile duct with intrahepatic biliary obstruction. Pathologically, the intra- and extrahepatic ducts are dilated and contain soft, pigmented stone and
pus. The cause of the disease is not known, but associations with clonorchiasis, ascariasis, and nutritional deficiency have been suggested.

The calculi are typically bile pigment stones with varying amounts of calcium. The stones have a mud or paste-like consistency. The stones presumably form due to bacterial enzymes causing deconjugation of bilirubin which then precipitates as calcium bilirubinate. In addition it has been postulated that a low protein diet is seen in some Asian populations may play a role in formation of intraductal stones.

**Pathophysiology:**

Recurrent episodes of cholangitis develop as a result of pyogenic bile duct infection. Predominantly coliform-type bacteria are seeded to the biliary system, and, as a consequence, a cascade of stone formation and recurrent cholangitis is initiated. The bile ducts of patients with cholangiohepatitis are thought to be increasingly susceptible to bacterial seeding as a result of helminthic infections or as a consequence of malnutrition.

Malnutrition results in a relative deficiency of enzymes that inhibit deconjugation of conjugated bilirubin. The relatively unopposed deconjugation favors breakdown of conjugated bilirubin to its unconjugated form, which then complexes with calcium to form calcium bilirubinate. Calcium bilirubinate acts as the nidus for the formation of pigmented stones. Pigment stone formation then leads to recurrent biliary obstruction with the characteristic consequence of suppurative cholangitis. This theory of nutrition associated deficiency of beta-glucuronidase has been called into question as definite differences in enzyme levels between patients with gallstones and control subjects are difficult to demonstrate.

Infection in the biliary system from ascaris lumbricoides or from trematodes, such as clonorchis sinensis and opisthorchis viverrini, often results in significant epithelial damage. Clinical and experimental data suggest that coliforms may then result in portal bacteremia by bacterial translocation as a result of this epithelial damage. Repeated portal bacteremia, in turn, results in the cascade of events characterized by biliary stasis, obstruction, and stone formation, which consequently result as RPC. However, it is more likely that infection and malnutrition act in concert to give rise to RPC.

**Epidemiology:**

The condition is rare. It often is observed in immigrants from Southeast Asia or from other regions of the world where trematode infections are endemic. Patients who have RPC develop it over time; hence, there does not appear to be a risk of developing RPC for visitors to endemic areas. The worldwide prevalence of cholangiohepatitis is unclear largely because of the paucity of disease statistics in many parts of the world where this disease is endemic. RPC is prevalent in Hong Kong and East Asia.

Although the arrival of Asian immigrants in the United Kingdom, Australia, Europe, and the United States has resulted in more cases reported in nonendemic areas, the overall incidence in East Asia has been in decline over the last 20 years. Population surveys confirm this trend. The decline in incidence has been credited to the improved economic situation and living standards with the associated westernization of diet.

**Mortality/Morbidity:**

Morbidity is related to recurrent biliary infection and may manifest as pancreatobiliary abnormalities, such as pancreatitis, biliary fistulae, hepatic abscesses, and supplicative cholangitis. Consequences of systemic infections and remote foci of infections account for distant or nonpancreatobiliary morbidity.

**History:**

Patients may present with the following:

1. An acute attack of cholangitis,
2. A history of recurrent attacks of cholangitis typified by fevers and right upper quadrant (RUQ) abdominal pain, or
3. Complications of pyogenic cholangitis. As the disease progresses, patients may develop cholangiocarcinoma and present with constitutional symptoms, including weight loss, easy fatigability, and jaundice.

**Physical:**

No specific physical findings are evident in RPC. The history is cardinal in prompting the diagnosis. Patients often appear ill, frequently are jaundiced, and usually have tenderness in the RUQ of the abdomen. Not infrequently in RPC, upon careful physical examination, an enlarged tender gallbladder can be palpated in the RUQ.

Other findings are specific to local and systemic complications of RPC, which can include the
following:
• Rupture into the peritoneum can precipitate an acute abdomen.
• Rupture or fistulization into the abdominal wall often presents with pus drainage from cutaneous fistulae.
• Rupture into the pericardium may present with tamponade.

Causes:
The underlying mechanism of RPC is unclear. Most experts believe that RPC is initiated by helminthic infection of the bile ducts and/or sludge/stone formation from deficient glucuronidation as a consequence of profound malnutrition. The initial insult(s) to the bile ducts precipitates a cycle of biliary stone formation and infection that results in recurrent episodes of pyogenic cholangitis.

Radiographic Features:
Magnetic resonance cholangiopancreatography (MRCP) is gold standard non invasive investigation for diagnosis of oriental cholangitis. The best diagnostic clues are intra and extrahepatic biliary dilatation due to multilevel strictures and calculi within them without gall bladder calculi, combination of variable density calculi / sludge and regions of segmental liver atrophy secondary to chronic biliary obstruction.
Localized dilatation of the lobar or segmental bile ducts and increased periportal echogenicity. Segmental hepatic atrophy, and gallstones have also been reported.

Differentials:
• Cholangitis
• Cholecystitis
• Choledocholithiasis
• Primary Sclerosing Cholangitis

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References:
http://emedicine.medscape.com/article/184131
http://emedicine.medscape.com/article/365322
http://manju-imagingexpert.blogspot.com/2010/01/oriental-cholangitis
http://www.bahrainmedicalbulletin.com/march_2002
http://radiopaedia.org/articles/oriental-cholangiohepatitis

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