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Case Report

Choledochal Cyst Originating from Primary Sclerosing Cholangitis in a Child

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Choledochal cysts are common in Asian children. Primary sclerosing cholangitis (PSC), which is characterized by inflammation and fibrosis and may lead to bile duct stricture over the intrahepatic or extrahepatic bile duct, is rare in children. Here we report a case of a 10-year-old boy who presented with a choledochal cyst originating from PSC. He had suffered from repeated abdominal pain and cholangitis for 3 years. A type IV choledochal cyst was suspected from the ultrasound and computed tomography image showing a distended gallbladder and dilatation of the bilateral intrahepatic duct at the hepatic hilar area and common bile duct (CBD). During laparotomy, a markedly distended gallbladder was noted and was shown to have no communication with the CBD by intraoperative cholangiogram. Choledochal cysts with extrahepatic and intrahepatic duct dilatation at the hilar area and marked stenosis with nearly total obstruction of the distal CBD were noted. Hepaticojejunostomy was performed. The histopathologic findings demonstrated a typical PSC picture. The patient's postoperative course was uneventful for 8 months after surgery, and he received no medication during a regular follow-up.

Key words: Choledochal cyst – Primary sclerosing cholangitis – Common bile duct stenosis – Children

Choledochal cysts are congenital dilatations of the extrahepatic biliary tree. They may involve any part of the bile duct.[1] Most choledochal cysts are diagnosed in patients younger than 10 years of age.[2] Primary sclerosing cholangitis (PSC) is a chronic cholestatic liver disease that is caused by inflammation around the bile ducts and leads to fibrosis of the duct. With progression of the disease, cholestasis causes damage to the hepatocytes, and the patient may die from liver failure.

Usually, PSC occurs in adults, with an incidence of 1 per 100,000 persons/year.[3] The incidence of PSC in children is much less.[4] The diagnosis of PSC is based on a cholestatic biochemical profile and cholangiography shows characteristic bile duct changes with multifocal strictures and segmental

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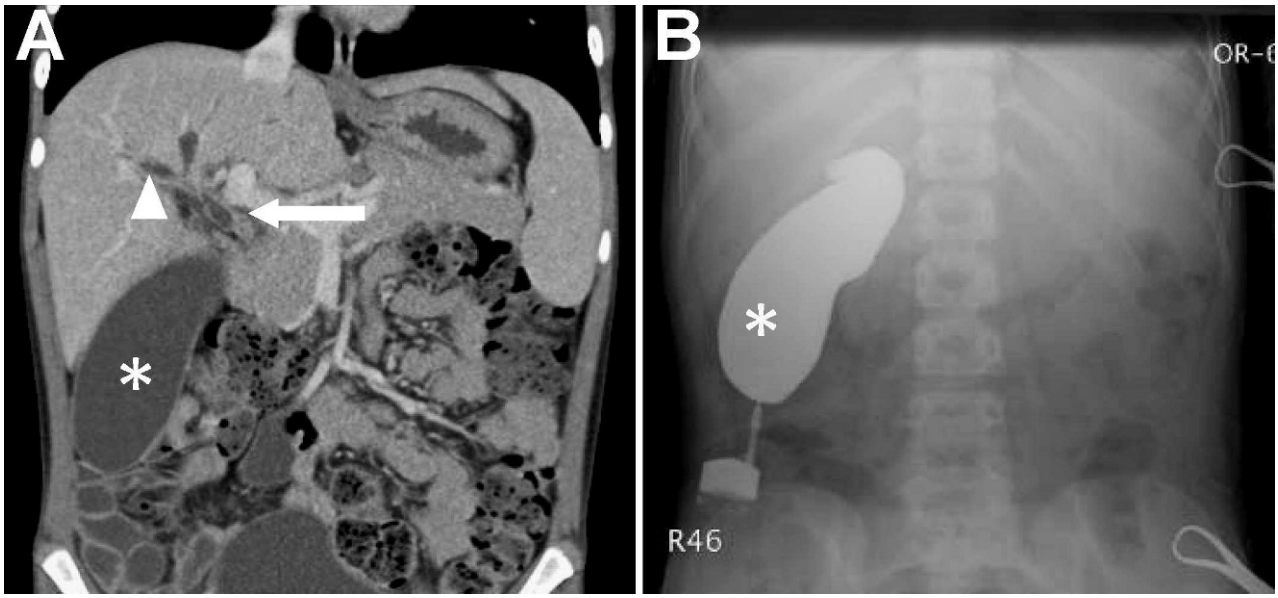

Fig. 1 (A) Preoperative magnetic resonance cholangiopancreatography showing dilated intrahepatic ducts (arrowhead) and proximal common bile duct (arrow) with a distended gallbladder (*). (B) Intraoperative cholangiogram showing a distended gallbladder (*) with no passage of contrast medium into the common bile duct.

dilatations of the hepatic duct and common bile duct (CBD). The administration of high dose ursodeoxycholic acid shows some beneficial effects in patients with PSC.[5] Surgical resection may be indicated if there is a dominant stricture over the extrahepatic duct with symptoms and signs of CBD obstruction. Liver transplantation is an option for patients with PSC-induced end-stage liver disease. Some autoimmune disease may occur in association with PSC, especially inflammatory bowel disease and autoimmune hepatitis. However, the most dreaded complication of PSC is cholangiocarcinoma.

Here we present a rare case of a choledochal cyst originating from PSC with dilated extrahepatic and intrahepatic ducts near the hepatic hilar area and a prominent distal CBD stricture in a 10-year-old boy.

Case Report

This 10-year-old boy had a history of frequent right upper quadrant abdominal pain in the past 3 years. An episode of clay-colored stool with icteric face was noted when he was 7 years old. The symptoms could be relieved by antibiotics and ursodeoxycholic acid therapy. However, the symptoms had frequently recurred since then. The abdominal sonogram showed dilated intrahepatic and extrahepatic ducts near the hilar area; the CBD also appeared to be dilated. Marked elevations of alkaline phosphatase (ALP, >2000 U/L), gamma-glutamyl transferase (GGT, >400 U/L), and hyperbilirubinemia were noted, and total bilirubin was higher than 8 mg/dL. Elevations of serum aspartate transaminase (AST, 226 U/L), alanine transaminase (ALT, 415 U/L), and anti-nuclear antibody (1:40 +) were also noted. The abdominal magnetic resonance cholangiopancreatography showed a distended gallbladder, bilateral intrahepatic duct dilatation near the hilar area, and a dilated proximal common bile duct (Fig. 1A). A type IV choledochal cyst was suspected.

During exploratory laparotomy, only very light green bile could be found in the markedly distended gallbladder. An intraoperative cholangiogram with urograffin demonstrated an isolated, distended gallbladder with no communication to the common bile duct (Fig. 1B). The upper CBD and intrahepatic duct were dilated, with nearly total obstruction of the distal CBD, which was filled with granulation and fibrotic tissue. Stenosis of the CBD was found at the distal part from the junction of the CBD with the cystic duct (Fig. 2). A choledochal cyst originating from the PSC with distal CBD obstruction was diagnosed. Cholecystectomy and excision of the proximal dilated and distal stenotic CBD, followed by a Roux-en-Y hepaticojejunostomy, were performed. The histopathologic picture of the distal CBD showed stenosis surrounded by concentric fibrous tissue, which is compatible with PSC

Fig. 2 The CBD was divided just distal to the junction with the cystic duct. A dilated proximal CBD (arrow) and a nearly total obstruction with granulation and fibrotic tissue of the distal CBD (arrowhead) were noted.

(Fig. 3A). The liver biopsy showed fibrotic expansion of portal areas. Bile duct damage with concentric fibrosis and ductular proliferation was also noted (Fig. 3B).

The patient's postoperative course was uneventful. The icteric face gradually recovered. Serum ALP, GGT, AST, ALT, and bilirubin returned to normal within 2 weeks of the surgical intervention. The patient has lived well uneventfully and medication-free for 8 months. Immunosuppressant drugs were not given during the regular follow-up period because no symptoms, signs, or evidence of autoimmune disease was seen after the surgical intervention. Regular follow-up with blood biochemistry examination (including liver enzymes and bilirubin levels) and abdominal echo every 3 months should be performed. Abdominal image examination such as magnetic resonance cholangiopancreatography should also be arranged every year after surgery to evaluate the patency of his bile duct.

Discussion

Primary sclerosing cholangitis usually occurs in adults. Sclerosing cholangitis with strong autoimmune features can occur in pediatric patients, more commonly in girls. It is reported to respond to immunosuppressive treatment and has a better prognosis than PSC in adults.[6] Dominant strictures of the CBD are uncommon, and the prevalence of cholangiocarcinoma is lower in children.[4] The treatment is similar to the treatment of PSC in adults, but immunosuppressants should be used in patients with autoimmune disease.

In the present case, the chronic cholangitis caused by PSC induced fibrosis of the bile duct, which led to stenosis of the distal CBD. During flare-ups of the disease, the inflammatory process can cause abdominal pain, fever, and elevated ALP, GGT, and serum bilirubin. Near total obstruction of the distal CBD may cause distension of the gallbladder and dilatation of the proximal CBD and the intrahepatic bile duct at the hilar area. A type IV choledochal cyst can then occur. Very light green bile in the distended gallbladder may be a sign of a partially obstructed cystic duct.

Surgical intervention is not routinely performed in patients with PSC. The indication for surgery in

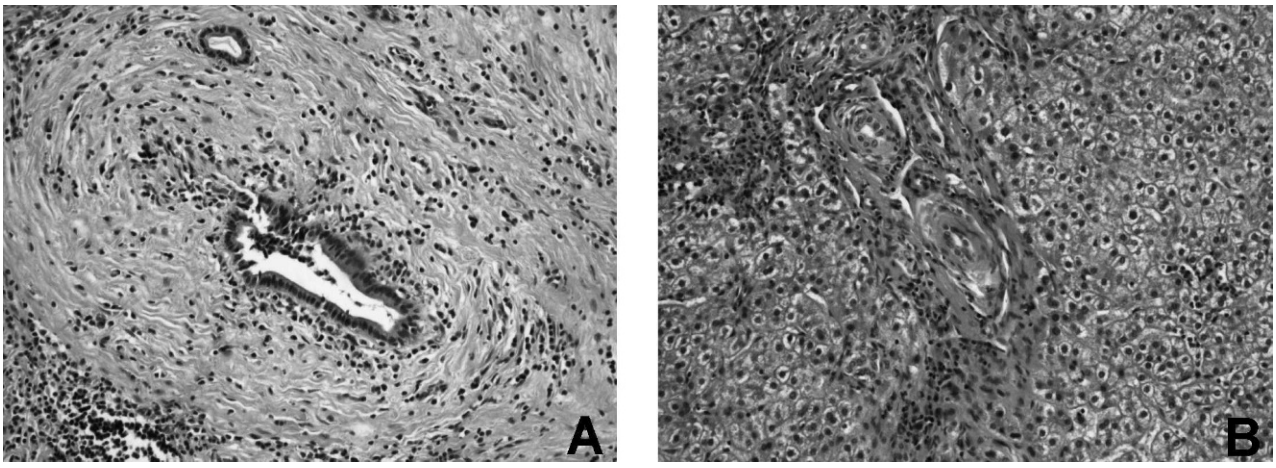

Fig. 3 Histopathologic picture showing the stenotic CBD surrounded by concentric fibrous tissue (A) and the intrahepatic bile ducts damaged by concentric fibrosis and ductular proliferation (B).

this patient was a dominant stricture of the extrahepatic duct and CBD. A ''dominant stricture'' has been defined as a stenosis with a diameter less than 1.5 mm in the CBD or less than 1 mm in the hepatic duct.[3] In the present case, the stricture of the distal CBD was causing near total obstruction. The gallbladder was severely distended, and intraoperative cholangiography showed no communication with the CBD. Excision of the CBD with bile drainage by Roux-en-Y hepaticojejunostomy was helpful in this case. There were several treatment options for bile duct strictures including stenting, surgical resection, or sphincterotomy. In the present case, surgical resection with Roux-en-Y choledochojejunostomy was selected because of the long segment of bile duct involvement and difficult passing any stent through the severe (almost complete) stricture site. The lifetime incidence of malignancy in choledochal cyst and among patients with PSC ranges from 6%–36% in the literature.[7] To differentiate between benign and malignant strictures, we could arrange yearly tumor markers, CT, or MRI for early detection of malignant change of the bile duct. In our patient, because a dominant stricture of smooth looking CBD with no marked bleeding tendency during the surgery was detected, which was followed by a histologic benign picture, confirmed after the surgical intervention. There was no malignancy found at present.

Medication is commonly used to treat PSC in adults. Ursodeoxycholic acid can decrease bile viscosity and facilitate the drainage of bile, thereby improving bile passage. It is reported that patients with PSC who undergo long-term, high dose ursodeoxycholic acid treatment show improvement on liver function tests, but there was no improvement in the survival rate.[8] High dose ursodeoxycholic acid was also reported to increase the rate of serious hepatotoxic side effects. Strong antibiotics might also be recommended if the cholangitis becomes more severe. In the present case, although the patient had received long-term ursodeoxycholic acid and antibiotic treatment, he underwent surgical intervention because of the frequent episodes of recurrent cholangitis and the image demonstrating a dilated CBD. No medication was given after the surgery because there were no signs, symptoms, or evidence of autoimmune disease during the follow-up period.

It has been reported that children with PSC might have inflammatory bowel disease or autoimmune overlap and advanced hepatic duct or CBD fibrosis at diagnosis.[9] In the present patient, the histopathology demonstrated an obstructive cholangiopathy, but there was no significant liver cirrhosis. Also, there was no laboratory evidence of autoimmune hepatitis. We concluded that the diagnosis was CBD originating from PSC without associated autoimmune disease.

The curative treatment of PSC with liver cirrhosis is liver transplantation if the impairment of liver function persists and end-stage liver disease is diagnosed. PSC will lead to liver failure in 10–15 years if the symptoms persist. The need for liver transplantation may be higher in patients in whom PSC occurs at a young age. The present patient had an uneventful postoperative course for 8 months. However, regular follow-up for a longer period will be necessary to clarify whether he needs further medication or even liver transplantation.

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