

Intrahepatic Stones from Congenital Biliary Dilatation

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ABSTRACT

Background: Congenital or primary intrahepatic bile duct (IHBD) dilatation is a rare disorder with symptoms of abdominal pain and hepatomegaly that usually presents in childhood and adolescence. Recurrent cholangitis, liver abscesses, septicemia, and biliary cirrhosis may result secondary to biliary sludge and hepatolithiasis.

Case Report: We present a case of IHBD dilatation with hepatolithiasis cured with surgical resection and discuss the management of this disease.

Conclusion: IHBD dilatation should be treated, as chronic biliary stasis and hepatolithiasis can lead to infection and recurrent cholangitis that can progress to cholangiocarcinoma. Treatment for IHBD dilatation usually involves multiple modalities including medical therapy, but ultimately resection of the diseased segments or lobes is required given the increased risk of malignancy.

INTRODUCTION

Congenital or primary intrahepatic bile duct (IHBD) dilatation is a rare disorder with symptoms of abdominal pain and hepatomegaly that usually presents in childhood and adolescence. Abnormal development of the ductal plate results in dilatation of the biliary tree, predisposing the biliary passages

to stagnation of bile. Recurrent cholangitis, liver abscesses, septicemia, and biliary cirrhosis may result secondary to biliary sludge and hepatolithiasis. The condition can progress to cholangiocarcinoma. We present a case of IHBD dilatation with hepatolithiasis and discuss the management of this disease.

CASE REPORT

A 47-year-old female with a history of pancreatitis and gout was referred to our institution for evaluation of abdominal pain and an abnormal magnetic resonance cholangiopancreatogram (MRCP). The patient had acute onset abdominal pain 1 week prior to the visit for which she was hospitalized. The pain, accompanied by nausea and vomiting, was described as severe and localized to the epigastrium. Laboratory investigation was notable for an increased total bilirubin of 1.5 mg/dL; remaining liver function tests and pancreatic enzymes were normal. Carcinoembryonic antigen and carbohydrate antigen 19-9 were also normal. A right upper quadrant ultrasound revealed tubular and nodular areas of echotexture within the lateral segment of the left hepatic lobe. The common bile duct measured 8 mm, and the gallbladder was unremarkable. MRCP demonstrated focal IHBD dilatation in the left lateral hepatic segment with associated parenchymal atrophy.

Subsequent endoscopic retrograde cholangiopancreatogram (ERCP) revealed dilatation of the common bile duct to 10 mm. The left main hepatic duct was diffusely dilated with an irregular cavitation, multiple segmental stenoses, and intraluminal filling defects (Figure 1). The common bile duct was explored with the SpyGlass probe (Boston Scientific) and showed evidence of sludge and erythema; the left intrahepatics were not accessible. Biopsies and brushings of the left main hepatic duct were negative for malignancy. Endoscopic ultrasound demonstrated multiple stones in the left intrahepatic ducts, and the left hepatic lobe appeared hypoechoic. Fine needle aspiration revealed marked acute inflammation with atypia but no evidence of malignancy. Postintervention computed tomography (CT) scan of the abdomen

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Figure 1. Endoscopic retrograde cholangiopancreatogram revealed significant left-sided biliary dilatation with intra-ductal stones (arrow).

revealed persistent biliary duct dilatation in the left lobe of the liver with evidence of high attenuation material related to stones, calcification, or debris.

The patient was referred for surgical resection with a plan for left hepatectomy. Initial operative inspection revealed a severely atrophied left lobe (Figure 2). Several perihepatic lymph nodes were dissected that were negative for malignancy. After removal of the left hepatic lobe, the resected specimen was dissected, further revealing hepatic duct ectasia with hepatolithiasis (Figure 3). Intraoperative cholangiogram demonstrated normal filling of the common and right hepatic ducts. The patient tolerated the procedure well, and her postoperative course was uncomplicated. Final pathology indicated congenital ductal dilatation with hepatolithiasis; no evidence of malignancy was found.

DISCUSSION

Congenital or primary IHBD dilatation was first described by Le Naour in 1941¹ but was further classified by Caroli et al and Guntz et al.^{2,3} Intrahepatic ductal dilatations belong to the spectrum of congenital bile duct cysts classified by Todani et al, in particular, type V choledochal cysts.⁴ Guntz et al subclassified the type V cysts into saccular IHBD dilatation of the peripheral ducts (type I), fusiform IHBD dilatation of the large ducts (type II), and saccular IHBD dilatation of the large ducts (type III).³ The intrahepatic bile duct cysts in Caroli disease fall under type I in this classification system.^{2,3} The



Figure 2. Initial operative inspection noted severe atrophy of segments 2 and 3 of the liver.

malformation may involve the entire intrahepatic biliary system but is more frequently unilobar, involving only the left side of the liver.⁵⁻⁷ The predisposition to the left side may be because of a selection bias, as patients selected for surgery usually have unilobar disease determined to be resectable.⁵

The presence of IHBD dilatation may be asymptomatic and may be an incidental finding on routine imaging. IHBD dilatation can be identified via endoscopic or noninvasive imaging. Patient presentation may dictate the initial use of endoscopic imaging, but noninvasive modalities such as CT and magnetic resonance imaging can identify the dilatation of the intrahepatic bile ducts as well as intrahepatic stones with focal atrophy of the surrounding tissue.⁶

The ductal dilatation causes a predisposition to stone formation because of biliary stasis that may subsequently cause pain and progress to cholangitis.⁵⁻⁷ Additionally, biliary stasis also predisposes patients to the formation of intrahepatic stones. The combination of biliary stasis, intrahepatic stones, and cholangitis can lead to chronic irritation of the biliary

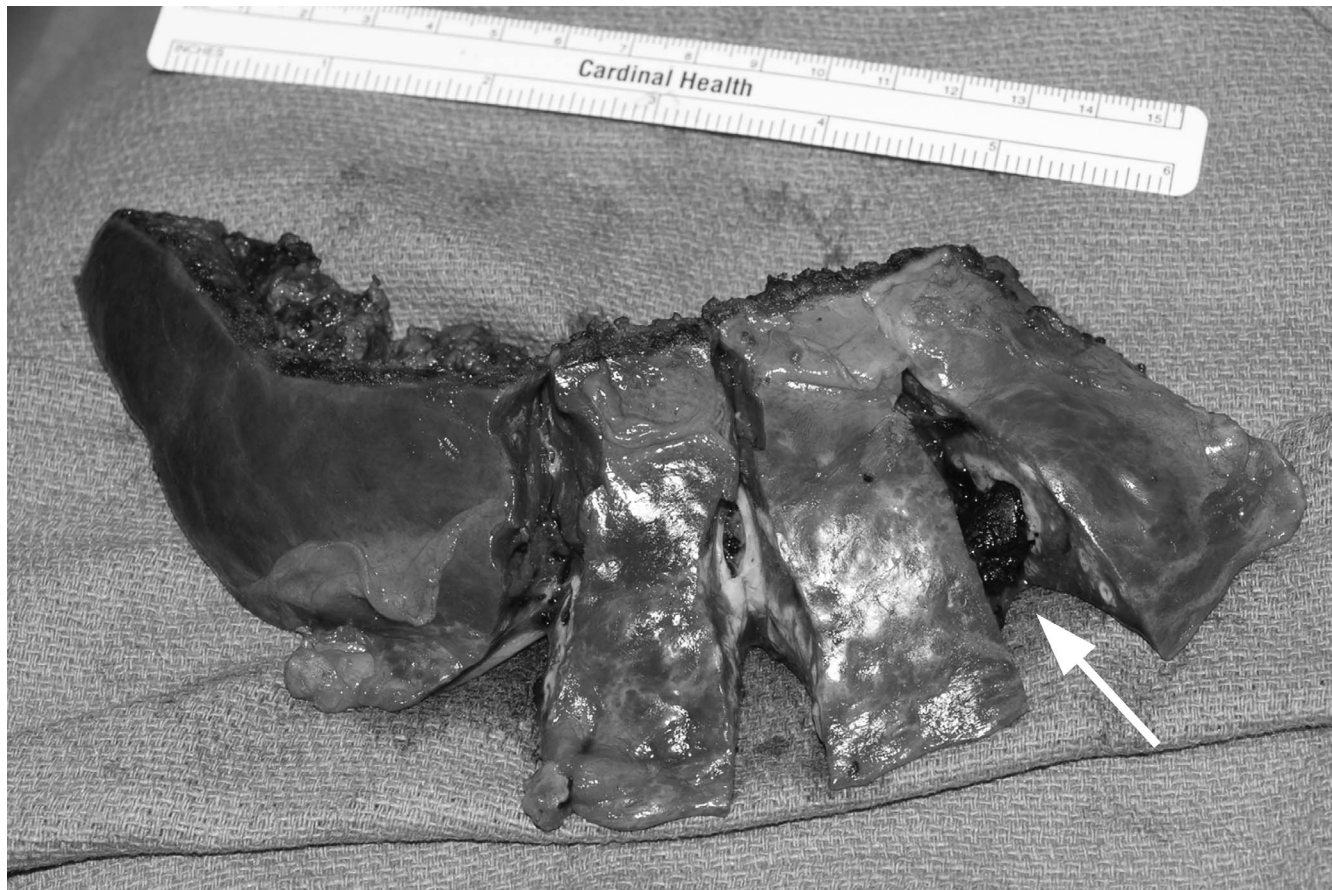


Figure 3. Large intrahepatic stones (arrow) were seen once the specimen was dissected.

mucosa, as well as parenchymal atrophy of the surrounding liver. The chronic irritation can progress to mucosal hyperplasia with chronic proliferative cholangitis and finally progress to cholangiocarcinoma.⁵⁻⁷ Indeed, the incidence of cholangiocarcinoma in this population is reported to be 7%-25%.^{5,7}

The treatment of IHBD dilatation is challenging and complex. Management depends on the extent of the disease and the preoperative condition of the patient. The goal of treatment is complete removal of all stones and the affected segment of liver. Several methods, such as interventional ERCP, shock wave lithotripsy, antibiotics, and ursodeoxycholic acid have been unsuccessful in curing the condition because of recurrent stones and cholangitis.⁷ Endoscopic and surgical drainage procedures, creating bilioenteric anastomoses, have also been reported but are not viable long-term options.⁵

In cases of unilobar disease, open or laparoscopic surgical resection of the diseased segments of liver is safe and effective. Resection removes the hepatolithiasis and reduces the risk of recurrent cholangitis with progression to cholangiocarcinoma.⁵⁻⁷ Extrahepatic extension of disease to the main biliary

convergence may require partial resection of the extrahepatic bile ducts to clear the disease.⁵ In cases of bilobar disease, radical liver resection and liver transplantation are viable options, with transplantation the treatment of choice for patients with concomitant hepatic cirrhosis or fibrosis.⁵ Intraoperative ultrasound can help determine the topographical extent of disease to find the margin of resection. An intraoperative cholangiogram can identify areas of filling defects that may represent intrahepatic stones and guide their removal.^{5,6}

CONCLUSION

IHBD dilatation should be treated, as chronic biliary stasis and hepatolithiasis can lead to infection and recurrent cholangitis that can progress to cholangiocarcinoma. The treatment usually involves multiple modalities including medical therapy, but ultimately resection of the diseased segments or lobes is required given the increased risk of malignancy. If evidence of hepatic fibrosis or cirrhosis is present, transplantation is the treatment of choice.

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