ACG CASE REPORTS JOURNAL

CASE REPORT | BILIARY

Role of Cholangioscopy in a Patient With Hepatolithiasis and Caroli Disease

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ABSTRACT

Caroli disease is an infrequent congenital pathology that is part of the spectrum of fibrocystic diseases, characterized mainly by malformation of bile ducts. These patients often have stones and cholangitis, which respond poorly to conventional endoscopic retrograde cholangiopancreatography. To date, there is little evidence on the usefulness of cholangioscopy in this disease, so we describe the experience of performing cholangioscopy in a patient with hepatolithiasis and Caroli disease.

INTRODUCTION

Caroli disease is an infrequent congenital pathology that is part of the spectrum of fibrocystic diseases, characterized mainly by bile duct malformation that manifests as cystic segmental dilatation of the intrahepatic bile ducts[1] that can compromise the entire liver or be limited to 1 hepatic lobe.[2] Its clinical presentation is variable, and in up to 60% of cases, it is associated with kidney disease, where its presentation is related to the dilation of the collecting ducts.[3] This structural alteration of the bile duct increases the risk of lithiasis, cholangitis, and liver abscesses,[1] and as a consequence, these patients require multiple biliary interventions throughout their lives, with conventional endoscopic retrograde cholangiopancreatography (ERCP) being the usual therapeutic method with limitations for the resolution of obstructive symptoms due to the technical difficulty secondary to the biliary anatomy of these patients. The performance of cholangioscopy in patients with Caroli disease is poorly described in the literature; however, it is a valuable alternative for the management of these complications, so we describe the experience and usefulness of performing this procedure in a patient with this medical condition.

CASE REPORT

A 44-year-old man with a medical history of renal transplantation in 2004 and Caroli disease, with recurrent cholangitis in the context of unresolved hepatolithiasis presented to the emergency department with a 6-day history of abdominal pain and fever. He previously required ERCP on 2 occasions at another institution without resolution of lithiasis, with the last ERCP performed 1 year before.

Physical examination was remarkable due to abdominal pain on the right hypochondrium and epigastrium, without signs of peritoneal irritation. Laboratory tests (Table 1). Magnetic resonance cholangiopancreatography showed multiple defects due to hepatolithiasis with changes of atrophy of the right hepatic lobe and alteration of the anatomy of hepatic bile ducts (Figure 1).

Because of the unsolved hepatolithiasis by conventional methods, it was decided to take this patient to cholangioscopy using the *SpyGlass* (Boston Scientific) direct visualization system that showed saccular changes of the intrahepatic bile ducts associated with multiple giant left lithiasis that excluded the right bile duct because of massive hepatolithiasis. Therefore, electrohydraulic lithotripsy and extraction of fragments with SpyGlass Retrieval Basket (Boston Scientific) were performed, with subsequent drainage of cholangitis, cleaning of the bile duct, and insertion of plastic stents selectively to the right and left ducts (Figure 2). Four weeks later,

ACG Case Rep J 2022;9:e00935. doi:10.14309/crj.0000000000000935. Published online: January 5, 2023

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Table 1. Laboratory tests

Laboratory test	Result	Reference range
White blood cell count (10^3 cells/μL)	3.35	5.0–10
Hemoglobin (g/dL)	7.01	13.5–18
Platelet (10^3 cells/μL)	141	150–450
Aspartate aminotransferase (U/L)	21	5–34
Alanine aminotransferase (U/L)	25	0–55
Alkaline phosphatase (U/L)	289	40–150
Gamma-gutamyl transferase (U/L)	321	12–64
Total bilirubin (mg/dL)	0.5	0.2–1.5
Direct bilirubin (mg/dL)	0.3	0.05
Amylase (U/L)	53	28–100

another session of cholangioscopy was performed, where electrohydraulic lithotripsy was performed again, and the hepatolithiasis resolved. Follow-up was indicated at 4 months.

DISCUSSION

Caroli disease is a rare autosomal recessive congenital pathology, with a prevalence of 1 in 1,000,000 people in the general population,[4] affecting both men and women with similar frequency, with most of them being diagnosed before the age of 30 years.[5] This pathology is part of the spectrum of fibrocystic diseases characterized mainly by the malformation of the bile ducts that manifests as cystic segmental dilatation of the intrahepatic bile ducts[1] and can compromise the entire liver or may be limited to either the right or left hepatic lobes.[2]

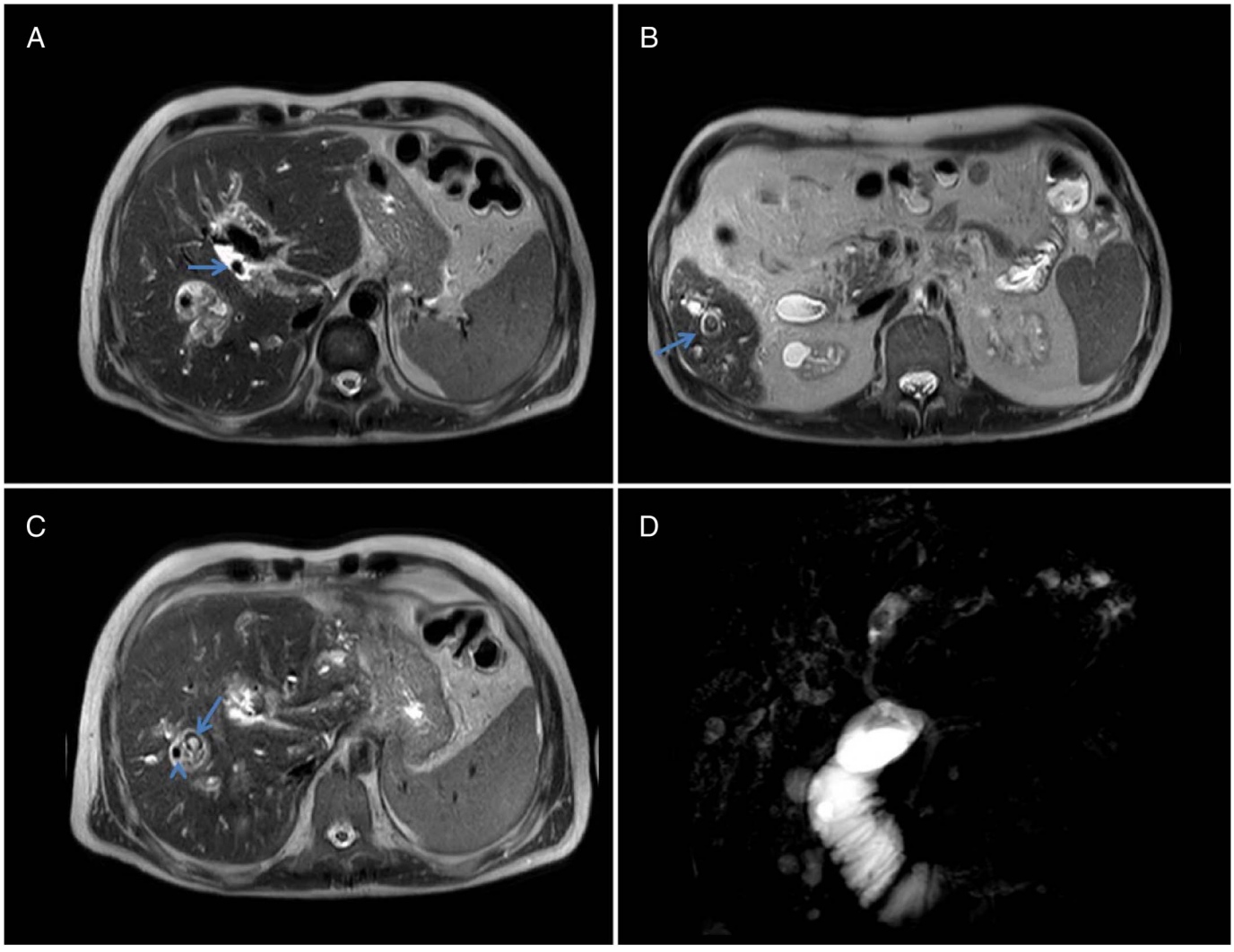

Figure 1. T2-weighted images obtained from cholangioresonance, showing slight lobulation of the hepatic contours and atrophy of the right hepatic lobe, associated with the loss of normal morphology of the intrahepatic bile ducts, with the presence of irregular cystic dilations and some defects inside them that are consistent with stones (marked with arrows in A, B, and C). (C) An image is also observed inside the intrahepatic bile duct that corresponds to the portal vein central point sign found in Caroli disease (indicated by the arrowhead). (D) Cholangiographic sequence showing multiple defects in the intrahepatic bile duct and the cystic dilatations described in the previous images.

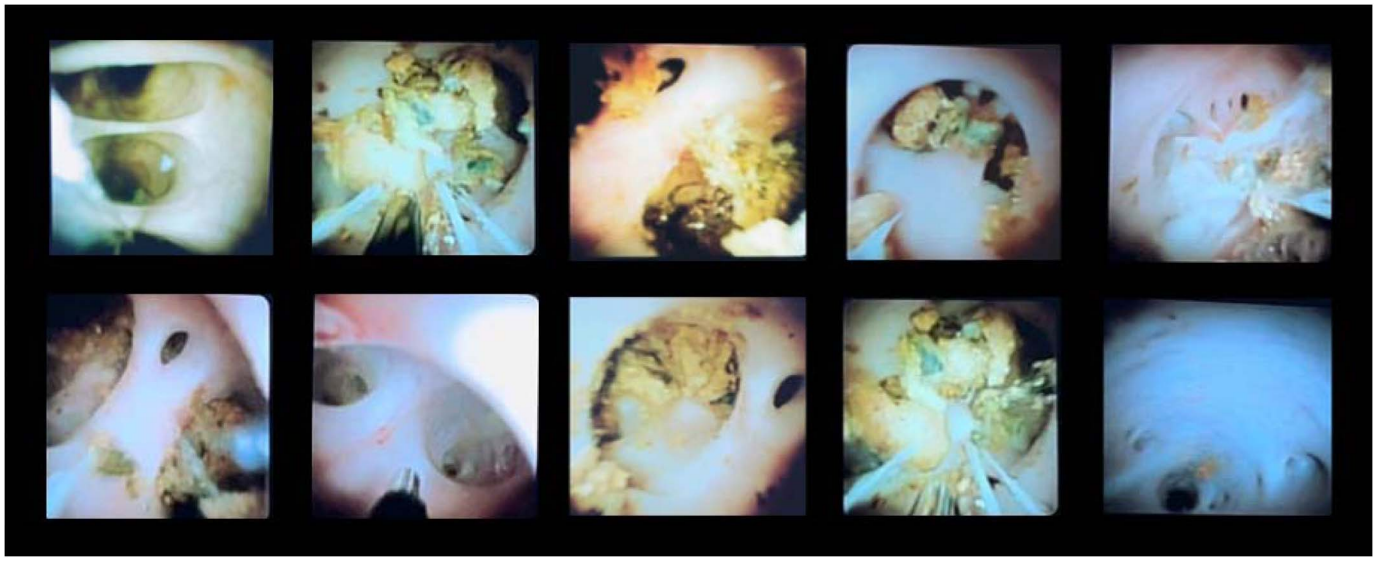

Figure 2. Cholangioscopy with evidence of multiple hepatolithiasis, extraction of stones with electrohydraulic lithotripsy, and extraction of fragments with a basket, with subsequent resolution of hepatolithiasis.

The disease has 2 presentations, the first is called Caroli disease type I or simply Caroli disease, which is characterized only by cystic segmental dilation of the bile ducts. The second is called type II Caroli disease, also called Caroli syndrome, which, in addition to saccular alterations of the hepatic ducts, is associated with liver fibrosis and even cirrhosis with manifestations of portal hypertension,[2] as is the case of our patient.

This structural alteration of the bile duct increases the risk of lithiasis, cholangitis, and liver abscesses,[1] and as a consequence, these patients require multiple therapeutic biliary interventions throughout their lives. However, resolving hepatolithiasis in these patients with conventional methods, such as ERCP, is complex, and given the usefulness of direct vision tools of the bile duct, cholangioscopy has been proposed as an effective method for the treatment of intrahepatic lithiasis.[6–10] This patient had previously undergone ERCP without achieving complete resolution of the intrahepatic lithiasis; therefore, cholangioscopy was performed using the *SpyGlass* (Boston Scientific) system, with resolution of the lithiasis through electrohydraulic lithotripsy in a total of 2 sessions.

As a consequence, cholangioscopy is a useful therapeutic procedure in the management of patients with hepatolithiasis and structural anatomical alterations, such as Caroli disease, as part of their comprehensive management.

DISCLOSURES

Author contributions: X. Morales collected the data, reviewed the literature, drafted the manuscript, and contributed to data interpretation and is the article guarantor. L. Jiménez-Hermida contributed to data interpretation. G. Hernández-Cely and JM Pérez extensively revised the manuscript, took care of the patient, and contributed to data interpretation. All authors read and approved the final manuscript.

Financial disclosures: There is no funding support for this manuscript. G. Hernández-Cely has a competing interest with Boston Scientific. All the other authors declare that they have no competing interests.

Informed consent was obtained for this case report.

Received May 25, 2022; Accepted November 14, 2022

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