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Case report

Endoscopic Management of Hepatic Cyst Secondary to Caroli Disease: A New Era—A Case Report

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Abstract

Introduction: Caroli disease is a rare condition characterized by segmental saccular dilatations of the intrahepatic biliary tree, which generally involve the entire hepatic parenchyma. One of its main features is the presence of multiple hepatic cysts throughout the liver parenchyma, although extrahepatic biliary involvement is uncommon. **Case report:** We present the case of a woman with a history of a hepatic cyst in the left lobe. Given her history of Caroli disease, she was admitted with a 10-day clinical course consisting of cholestatic jaundice, weight loss, and pruritus. Magnetic resonance cholangiography revealed multiple saccular dilatations in the intrahepatic bile ducts and a hepatic cyst arising from the biliary tree. **Discussion:** The diagnosis of Caroli disease was primarily established through endoscopic procedures, which demonstrated multiple hepatic cysts and a giant hepatic cyst. In this case, endoscopic methods were employed, resulting in successful treatment with lower morbidity. **Conclusions:** This case report highlights that Caroli disease, being a rare and highly complex condition, has seen limited advances in its treatment. We therefore report on a new era of endoscopic management that enables disease resolution with reduced morbidity and mortality.

Keywords

Caroli disease; intrahepatic bile ducts; cholestasis; jaundice; natural orifice endoscopic surgery.

INTRODUCTION

Jacques Caroli first described Caroli Disease in 1958. This disease is a congenital malformation of the intrahepatic bile ducts characterized by ectasia and dilation of the intrahepatic ducts, which may involve the biliary tract in either a focal or multifocal manner[(1)]. It is a low-prevalence condition, occurring in approximately fewer than one case per 1,000,000 inhabitants[(2)]. The clinical presentation of this disease varies, but it is mainly characterized by right upper quadrant abdominal pain, jaundice, fever, pruritus, and the development of multiple hepatic cysts, which are managed through interventional radiology; however, they carry a high risk of superinfection due to repeated punctures[(2–9)]. Management of complications associated with Caroli disease is primarily performed through percutaneous drainage to resolve the collections that form within the liver. Patients often require multiple punctures performed at different stages to complete drainage and achieve resolution of the septic process[(8,9)]. Consequently, endoscopic surgical techniques have been described in which collection drainage can be achieved through the placement of stents, resulting in lower morbidity.

In caroli Disease, hepatic damage or liver failure is characterized by the presence of hepatic cysts mainly associated with primary hepatic fibrosis, and in rare cases these hepatic

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cysts contain tissues dependent on the intrahepatic biliary tree[3]. Therefore, the present case describes the management of a giant hepatic cyst communicating with the intrahepatic biliary tree, treated through endoscopic surgery.

CLINICAL CASE

A 51-year-old female patient was admitted to the emergency department with a one-week history of generalized jaundice associated with headache, nausea, choluria, and acholia, which had intensified over the previous three days. The patient had a history of a hepatic cyst diagnosed in 2013, which was resected laparoscopically in 2022, with immunohistochemical findings consistent with a simple hepatic cyst. On physical examination, the following vital signs were recorded: blood pressure, 132/77 mm Hg; heart rate, 78 beats per minute (bpm); oxygen saturation (SpO_2), 98%; and temperature, 36.5 °C. Generalized jaundice and tenderness on palpation of the right upper quadrant without signs of peritoneal irritation were noted. A biliopancreatic profile was requested, yielding the following results: total bilirubin, 8.2 mg/dL; direct bilirubin, 7.2 mg/dL; indirect bilirubin, 1.2 mg/dL; lipase, 9 mg/dL; amylase, 25 mg/dL; alkaline phosphatase, 245 mg/dL; aspartate aminotransferase (AST), 52 IU/L; and alanine aminotransferase (ALT), 99 IU/L. Based on these findings, an intermediate risk of choledocholithiasis was established according to the 2019 ASGE criteria, and magnetic resonance cholangiography was requested (**Figure 1**), which identified a giant complex hepatic cyst in the left hepatic lobe measuring 57 × 66 mm in diameter.

Given the obstructive etiology of the jaundice syndrome, an endoscopic retrograde cholangiopancreatography (ERCP) was performed, revealing an angulated and stenotic common bile duct in its middle third, with dilation of the hepatic and intrahepatic ducts. Contrast imaging additionally demonstrated a large hepatic cyst measuring greater than 12 cm, communicating with the left hepatic ducts, with evidence of uncontrollable biliary, hemorrhagic, and purulent drainage through the papilla. During the procedure, a 10 French (Fr) × 12 cm biliary stent was placed.

Subsequently, an endoscopic ultrasound (**Figure 2**) was performed, revealing a complex left hepatic cyst with thick walls, irregular contours, and dense internal fluid. An endoscopic cystogastrostomy was performed with placement of a 10 × 15 mm Hot Axios stent, followed by drainage of the cyst. An *in situ* biliary stent was also observed. A second endoscopic ultrasound was then performed, during which a cystogastrostomy was carried out with placement of a 10 mm × 15 mm Hot Axios stent (**Figure 3**), inserted through a subcardial approach (**Figure 4**), resulting in drainage of 350 mL of hematopurulent content. Findings suggested monolobar left-sided hepatic Caroli disease and severe acute cholangitis.

DISCUSSION

Caroli disease belongs to a family of fibrocystic ductal plate disorders, which also includes Autosomal Recessive Polycystic Kidney Disease, Congenital Hepatic Fibrosis, Autosomal Dominant Polycystic Kidney Disease, biliary hamartomas, and mesenchymal hamartomas[1]. Its etiology remains unknown; however, it appears to be associated with an autosomal recessive inheritance pattern[2].

When symptomatic, Caroli disease usually manifests with recurrent episodes of right upper quadrant abdominal

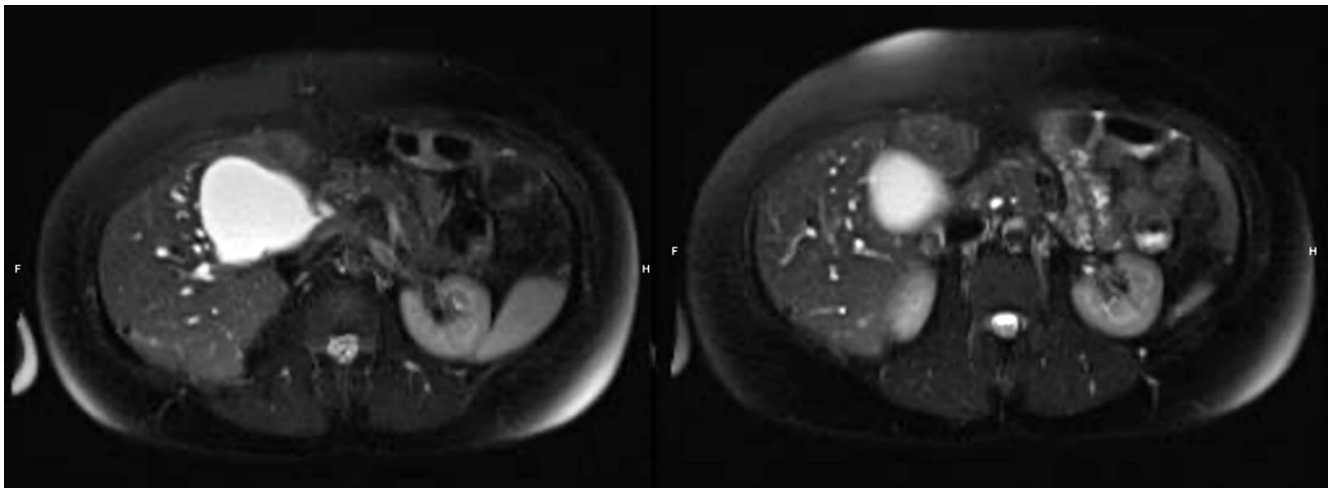


Figure 1. Magnetic resonance cholangiography showing evidence of a giant hepatic cyst with multiple hepatic cysts and collections. Author's file.

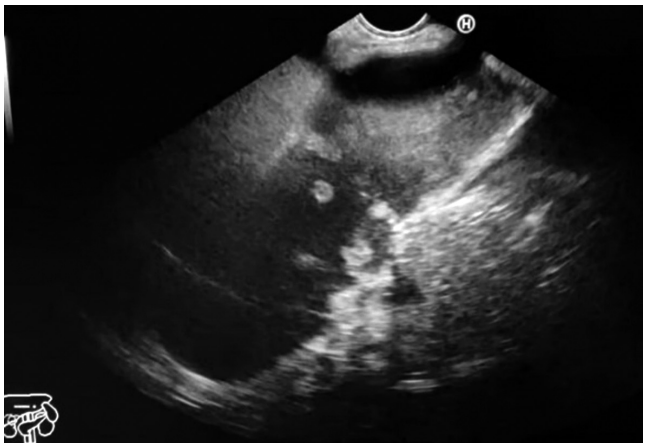


Figure 2. Giant hepatic cyst and multiple cysts identified by endoscopic ultrasound. Author's file.

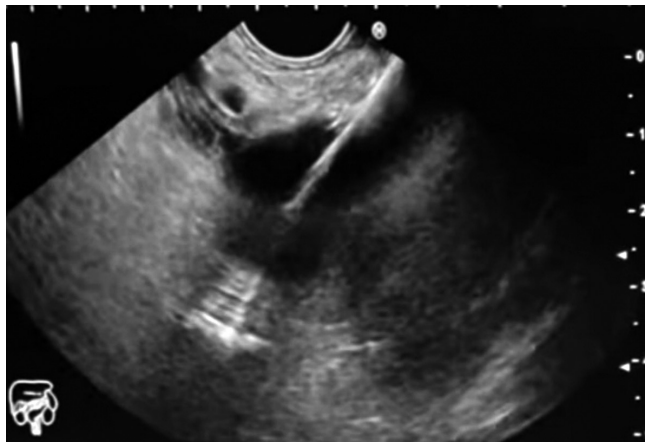


Figure 3. Endoscopic cystogastrostomy with placement of a Hot Axios stent. Author's file.

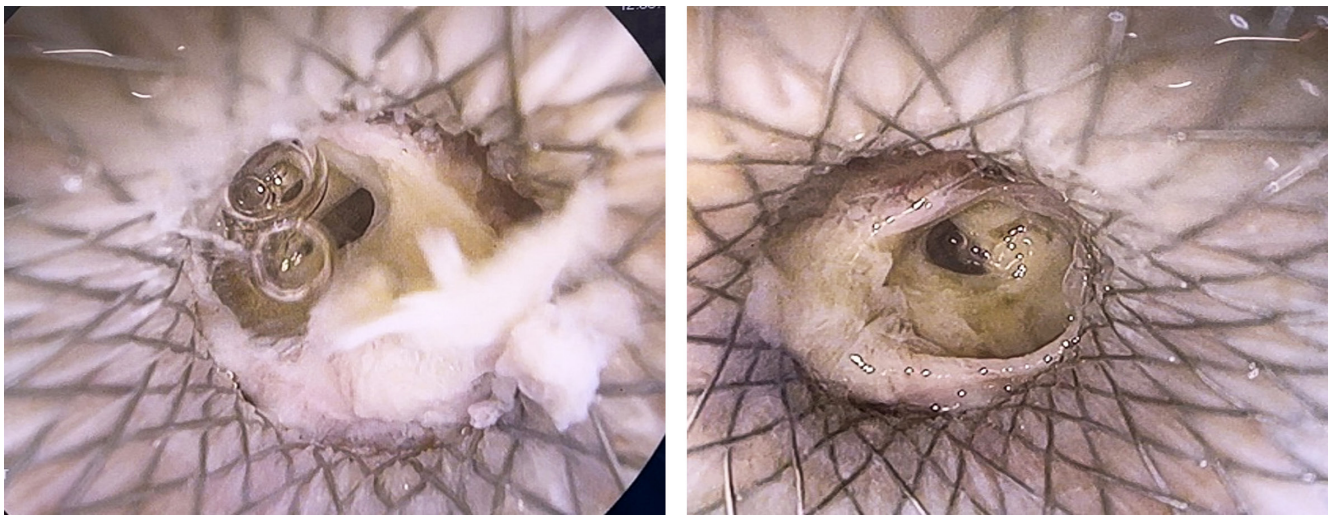


Figure 4. Subcardial endoscopic cystogastrostomy with placement of a 10 × 15 mm Hot Axios stent, showing evidence of intrahepatic ducts. The patient was transferred to the intermediate care unit for management of Tokyo III acute cholangitis. She showed favorable clinical progression and was transferred to the general ward 72 hours after admission to the unit. The patient continued to respond adequately to endoscopic management with the Hot Axios stent, with no evidence of purulent secretion drainage, resolution of acute cholangitis, and subsequent satisfactory recovery. Author's file.

pain, which may be accompanied by jaundice and pruritus. Recurrent bacterial cholangitis is the most common form of presentation and may occur with abdominal pain, fever, and chills[2]. In light of the above, the present case had a relatively typical presentation, with manifestations including abdominal pain, jaundice, choluria, and acholia.

The diagnosis of Caroli disease is based primarily on imaging findings demonstrating distal dilatations dependent on the intrahepatic biliary ducts through ultrasonography, computed tomography, or, as in this case, magnetic resonance cholangiography. Imaging characteristics include cystic dilation of the bile duct, and the presence of adjacent intrahepatic calculi secondary to biliary stasis is also a typical finding[1]. Differential diagnoses include conditions causing severe biliary dilation, as well as polycystic liver disease and congenital hepatic fibrosis[1]. Demonstration of communication between the cysts and the biliary tree is essential for establishing the diagnosis of Caroli disease[4].

Considering that there are currently no management guidelines for Caroli disease[2], treatment is individualized and based on associated comorbidities or complications, such as cholangitis, hepatic abscesses, hepatolithiasis[4], and the development of neoplasms such as cholangiocarcinoma, among others. Consequently, management involves multi-

disciplinary interventions including endoscopy, interventional radiology, and surgery[4]. On the one hand, medical therapy with ursodeoxycholic acid is implemented in some cases, such as cholestatic jaundice syndromes[2]; on the other hand, analgesics and antibiotic therapy are used in cases requiring these medications[3]. Surgical management appears to provide better outcomes in the presence of complications, risk of malignancy, or extensive disease, which may justify surgical intervention, potentially extending to hepatectomy[2]. Surgical management includes segmental or lobar hepatectomies and, in cases such as the diffuse form of Caroli disease, may ultimately require liver transplantation[1,3,4].

Among minimally invasive therapeutic options, transhepatic access or percutaneous drainage performed by the interventional radiology service has been described, as well as hepatogastric drainage guided by endoscopic ultrasonography using lumen-apposing metal stents such as the Hot Axios[6]. The latter option is particularly useful in patients with proximal biliary obstruction and altered surgical anatomy, for example[5].

Complications associated with the use of metal stents include migration rates of up to 21.2%, which are directly proportional to the duration of use; bleeding rates ranging from 0% to 10% of patients have also been reported[6].

Considering the overall incidence of malignant transformation in Caroli disease of 7% and the persistent risk of recurrence, annual follow-up with imaging studies and serial blood testing is maintained[2,3].

CONCLUSIONS

Caroli disease is a rare condition with low incidence. It usually presents with nonspecific symptoms, although one of the main findings is the presence of multiple hepatic cysts, which may be incidental findings. However, when symptomatic, patients present with an obstructive biliary syndrome leading to a clinical picture of acute cholangitis. Initial management has traditionally been performed through percutaneous drainage by interventional radiology; however, the new era has begun to adopt endoscopic drainage management, resulting in lower mortality and morbidity among patients.

Informed Consent

The patients completed informed consent forms in which they authorized the use of their medical records and diagnostic images for research purposes and case reporting. The case reports and methodology were approved by the institutional scientific ethics and research committee under Approval Record No. 023/22 dated May 17, 2021, in compliance with Resolutions No. 8430 of 1993 (risk-free study) and No. 2378 of 2007 issued by the Ministry of Social Protection, as well as in accordance with the provisions of the UNESCO Universal Declaration on Bioethics and Human Rights.

Conflict of Interest

The researchers declared that there were no conflicts of interest of any kind.

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