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Case Report

Caroli disease presenting with recurrent cholangitis and secondary small bowel obstruction: a case report

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ABSTRACT

Caroli disease is a rare congenital hepatobiliary disorder characterized by non-obstructive dilatation of the intrahepatic bile ducts and classified as Todani type V choledochal cyst. Although congenital in origin, the disease is often diagnosed in adulthood due to its indolent course and nonspecific symptoms, frequently after complications such as recurrent cholangitis have occurred. We report a case of a 46-year-old male presenting with recurrent epigastric pain, abdominal distension, fever, jaundice, nausea, vomiting, and bowel movement disturbance. Laboratory findings revealed leukocytosis, direct hyperbilirubinemia, and cholestatic liver enzyme elevation consistent with acute cholangitis. Abdominal radiography demonstrated features of partial small bowel obstruction. Magnetic resonance cholangiopancreatography (MRCP) with contrast revealed multifocal cystic dilatation of the intrahepatic bile ducts without extrahepatic obstruction, consistent with Caroli disease. The patient was managed conservatively with intravenous antibiotics, ursodeoxycholic acid, bowel decompression, and supportive care, resulting in gradual clinical improvement. This case highlights the diagnostic value of MRCP and demonstrates that conservative management can be effective in adult Caroli disease complicated by recurrent cholangitis and secondary small bowel obstruction.

Keywords: Caroli disease, Recurrent cholangitis, Small bowel obstruction

INTRODUCTION

Caroli disease is a rare congenital disorder of the biliary tract caused by ductal plate malformation, resulting in non-obstructive, segmental, or diffuse dilatation of the intrahepatic bile ducts.[1,2] It is classified as Todani type V choledochal cyst and may occur as an isolated condition or as part of Caroli syndrome when associated with congenital hepatic fibrosis.[3]

Despite its congenital nature, Caroli disease frequently manifests in adulthood due to slow progression and nonspecific clinical features.[1,4] Patients often present with recurrent abdominal pain, fever, and jaundice related to repeated episodes of cholangitis caused by chronic bile stasis within dilated ducts.[4] Radiological imaging is central to diagnosis, with magnetic resonance cholangiopancreatography (MRCP) currently considered the modality of choice. MRCP allows detailed visualization of the biliary tree, demonstrating characteristic multifocal intrahepatic ductal dilatation while excluding extrahepatic obstruction.[5-7]

This case report describes an adult patient with Caroli disease presenting with recurrent cholangitis complicated by secondary small bowel obstruction, emphasizing diagnostic challenges and management considerations.

CASE REPORT

A 46-year-old male presented to the emergency department with persistent epigastric pain, abdominal distension, nausea, vomiting, fever, and reduced oral intake. The pain was continuous, moderate in intensity, and had recurred multiple times, leading to repeated hospital admissions. The patient also reported difficulty

passing stool and flatus, dark-colored urine, and episodes of cold sweating.

Previous abdominal computed tomography (CT) scan and magnetic resonance imaging (MRI) were reported as unremarkable. The patient had no history of chronic illness or surgery. However, he reported daily alcohol consumption and a high-fat diet, which may exacerbate biliary stasis and inflammation. There was no relevant family history of hepatobiliary disease. Physical examination revealed icteric sclera, epigastric and right upper quadrant tenderness, abdominal distension, and hepatomegaly palpable approximately two fingerbreadths below the costal margin. Laboratory investigations showed leukocytosis with neutrophil predominance, elevated direct bilirubin, increased alkaline phosphatase, and elevated transaminases, consistent with cholestasis and systemic infection.

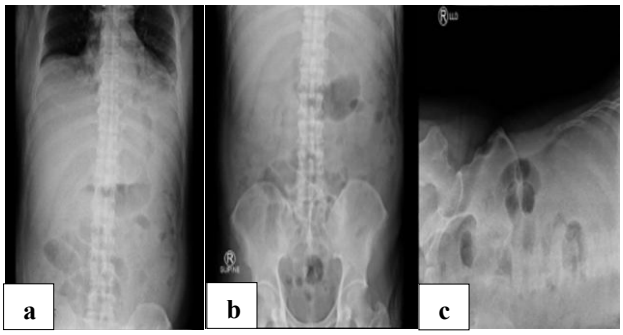

Figure 1 (a-c): Plain abdominal radiography (BNO three-position) result of the patient, showing dilated small bowel loops with a herringbone appearance and air distribution reaching the distal colon.

Plain abdominal radiography (BNO three-position examination) demonstrated dilated small bowel loops with a herringbone appearance and air distribution reaching the distal colon, consistent with partial small bowel obstruction. Abdominal MRI and MRCP with contrast examination revealed multiple cystic dilatations of varying sizes within the intrahepatic bile ducts without evidence of intra- or extrahepatic biliary obstruction, consistent with Todani type V choledochal cyst (Caroli disease). Additional findings included acalculous cholecystitis, mild bilateral pleural effusion, and a small renal cyst.

During hospitalization, the patient underwent an 11-day course of treatment and supportive care with close clinical and laboratory monitoring. Initial therapy consisted of intravenous fluid resuscitation with Ringer lactate, proton pump inhibitor therapy with Lansoprazole, antiemetic treatment using Ondansetron, and empirical broad-spectrum antibiotic therapy with Ceftriaxone, which was later escalated to a higher dose due to persistent leukocytosis and clinical signs of infection. Additional supportive therapy included Paracetamol for fever and analgesia, as well as gastrointestinal agents such as antacids and prokinetics (domperidone).

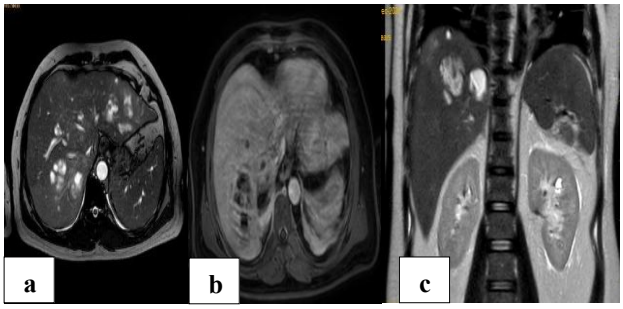

Figure 2 (a-c): Abdominal MRI with contrast image result of the patient showing multiple cystic lesions, acalculous cholecystitis, and small cyst in upper pole of left kidney.

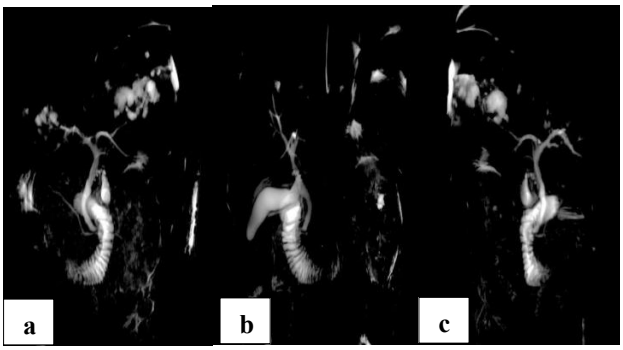

Figure 3 (a-c): MRCP with contrast image result of the patient also showing multiple cystic lesions, various size in intrahepatic bile duct suggestive of choledochal cysts type 5 (Caroli's disease), and acalculous cholecystitis.

In the initial phase (day 1–3), the patient remained symptomatic with persistent abdominal pain, nausea, leukocytosis, and the development of jaundice, consistent with ongoing cholangitis. Hepatobiliary-directed therapy with Ursodeoxycholic acid was initiated to improve bile flow and reduce cholestasis. By day 4–5, symptoms progressed with worsening abdominal distension and obstipation, and imaging confirmed partial small bowel obstruction (SBO). Multidisciplinary management was initiated and escalated with bowel rest, nasogastric tube decompression, and rectal interventions including fleet enema, along with continued intravenous therapy. During this phase, oral medications were temporarily withheld, while laxatives such as Lactulose and additional rectal agents were administered to facilitate bowel movement.

From day 6 onward, gradual clinical improvement was observed. Abdominal distension decreased, bowel function began to return (flatus followed by defecation), and inflammatory parameters showed a downward trend. By day 7–8, the patient demonstrated significant improvement in gastrointestinal symptoms with resolution of obstruction signs. Jaundice and abdominal pain progressively subsided. In the recovery phase (day 9–11), the patient subsequently tolerated gradual reintroduction of oral intake, and previously withheld medications were

resumed. Vital signs were stable, and no recurrence of obstructive symptoms were observed. The patient was discharged on day 11 in stable condition with significant clinical improvement.

At discharge, the patient was prescribed oral Lansoprazole, Domperidone, Paracetamol, sucralfate, and lactulose. The patient was advised for regular outpatient follow-up in the internal medicine clinic. On 1-week follow-up, the patient remained clinically stable with no recurrence of cholangitis or bowel obstruction symptoms. Long-term management planning includes periodic monitoring of liver function, imaging surveillance for disease progression, and evaluation for potential definitive management options. Given the diagnosis of Caroli disease, the patient was counseled regarding the risk of recurrent cholangitis and potential complications, including biliary cirrhosis and cholangiocarcinoma. Further evaluation for hepatobiliary surgical intervention or liver transplantation may be considered if the disease progresses or complications recur.

DISCUSSION

Caroli disease is a rare congenital hepatobiliary disorder resulting from ductal plate malformation, leading to non-obstructive dilatation of the intrahepatic bile ducts.[1,3,8] Although congenital, clinical manifestation frequently occurs in adulthood due to slow disease progression and nonspecific symptoms, often after complications such as cholangitis have developed.[1,4] In this patient, recurrent abdominal pain, fever, jaundice, and laboratory findings of leukocytosis and cholestatic liver enzyme elevation are consistent with adult Caroli disease complicated by acute cholangitis.[4]

Importantly, MRCP demonstrated no evidence of extrahepatic biliary obstruction, which has significant implications for management. MRCP played a decisive role in establishing the diagnosis by demonstrating multifocal cystic dilatation of the intrahepatic bile ducts consistent with Todani type V choledochal cyst.[5,7] This modality remains the diagnostic gold standard due to its ability to delineate biliary anatomy noninvasively and exclude alternative causes of biliary obstruction. In the absence of biliary obstruction or stones, invasive biliary drainage procedures such as endoscopic retrograde cholangiopancreatography (ERCP) are not routinely indicated.[6,7] This supports the decision to pursue conservative medical management rather than urgent biliary decompression in this patient. Previous studies have emphasized that cholangitis in Caroli disease frequently arises from intrahepatic bile stasis rather than mechanical obstruction, making antibiotic therapy and supportive care the cornerstone of acute management.[9,10]

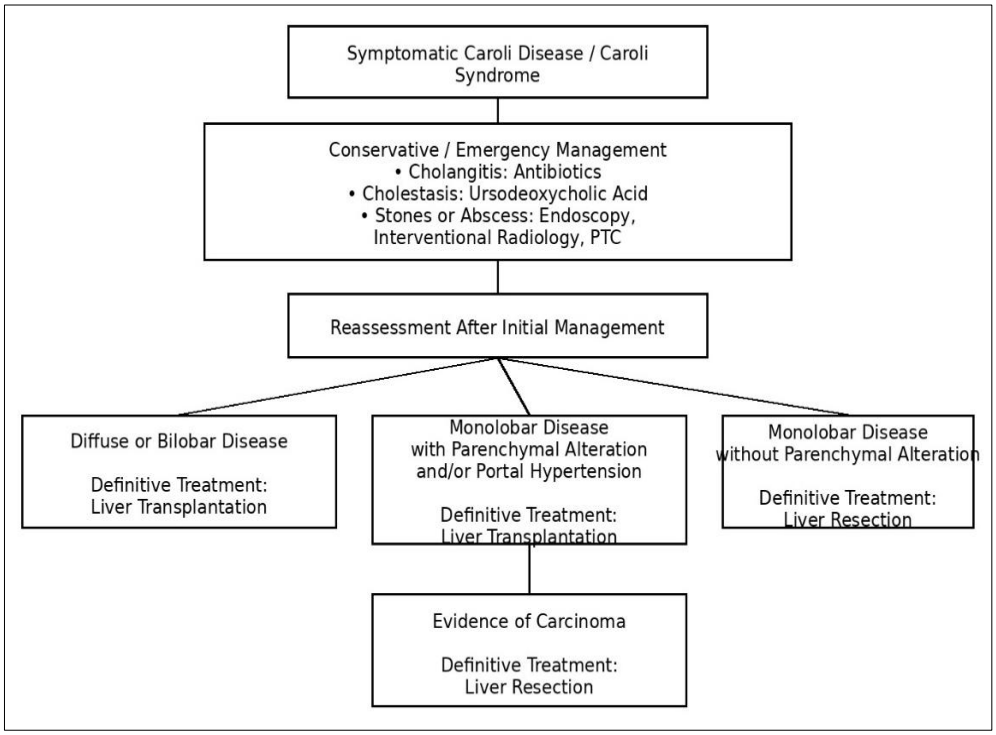

Figure 4: Algorithm for the treatment of symptomatic Caroli disease or Caroli syndrome.[12]

An additional important finding in this case was partial SBO identified on BNO examination. Although SBO is not a classic manifestation of Caroli disease, several mechanisms may explain this association. Chronic hepatomegaly and marked intrahepatic bile duct dilatation can exert a mass effect on adjacent intra-abdominal structures, potentially impairing intestinal motility.[10] Furthermore, recurrent cholangitis and biliary sepsis may induce a systemic inflammatory response, leading to electrolyte imbalance, splanchnic hypoperfusion, and

secondary paralytic or sub-obstructive ileus.[10,11] In this patient, the presence of abdominal distension, vomiting, impaired bowel movement, and radiographic evidence of dilated small bowel loops suggests that SBO was likely secondary rather than primary mechanical obstruction. The clinical improvement following nasogastric decompression, bowel rest, and resolution of infection further supports a functional or inflammation-related mechanism, consistent with previously reported cases of secondary ileus associated with severe hepatobiliary disease.[11]

Management of Caroli disease depends on disease distribution and severity. In this case, acute management focused on controlling cholangitis with intravenous fluids, early broad-spectrum antibiotics, and close monitoring, in accordance with international guidelines.[4] The absence of biliary obstruction on MRCP justified a conservative approach without urgent biliary drainage.[9] Ursodeoxycholic acid (UDCA) was administered as part of long-term management. UDCA has been shown to improve bile flow, reduce cholestasis, and potentially decrease the frequency of recurrent cholangitis episodes in patients with Caroli disease and other cholestatic liver disorders. Although evidence is limited to observational studies, UDCA is commonly recommended as adjunctive therapy in diffuse Caroli disease managed conservatively.[2] Surgical intervention is reserved for localized disease, refractory recurrent cholangitis, or advanced complications such as portal hypertension or secondary biliary cirrhosis.[3] Liver transplantation remains the definitive treatment for diffuse disease with liver failure.[3,12] The favorable response in this patient supports evidence that medical therapy can effectively control acute complications. Nevertheless, long-term surveillance remains mandatory due to the increased risk of recurrent cholangitis, hepatolithiasis, secondary biliary cirrhosis, and cholangiocarcinoma in adult Caroli disease.[1]

CONCLUSION

Caroli disease should be considered in adult patients presenting with recurrent cholangitis-like symptoms and unexplained cholestatic liver abnormalities. MRCP plays a crucial role in establishing the diagnosis. Conservative management can provide favorable short-term outcomes in selected patients presenting with recurrent cholangitis and secondary small bowel obstruction; however, long-term surveillance remains mandatory due to the risk of recurrent infection and malignancy.

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