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Caroli's Disease Presents as Acute Cholangitis and Recurrent Jaundice: A Case Report

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Abstract: ***Background:*** ***Caroli's disease is a rare congenital disorder characterized by dilatation of intrahepatic bile ducts, bile stasis, and bacterial infections. While frequent episodes of cholangitis are common in Caroli's disease, it represents a life-threatening condition that might lead to sepsis and multi-organ failure if left untreated. The presentation of disease may vary from abdominal pain, cholangitis, or end-stage liver disease; an enlarged liver is usually seen. Caroli's disease is type V in Todani's classification of choledochal cysts, which categorizes them into five types based on their characteristics and anatomical involvement. The Global prevalence was less than 1 in 100,000 births. Male-to-female ratio 1:1.8. Age at diagnosis: Most patients present before age 30. Familial occurrence tends to be sporadic, but familial cases occur in 10-20% of cases. We present a case of Caroli's disease with cholangitis, highlighting the diagnostic approach, clinical course, and treatment strategies. Caroli's disease is often misdiagnosed as uncertain cholestasis until complications like cholangitis occur. ERCP contrast injection can trigger cholangitis in patients with biliary dilatation. We advise limiting ERCP use to therapeutic purposes and employing low-pressure contrast techniques when necessary. Case Presentation: We present a case of a 41-year-old male with a long history of recurrent jaundice and a cholecystectomy 10 years ago, who came to the emergency room with a 6-day complaint of fever, right upper quadrant abdominal pain, and jaundice. Radiological studies showed the characteristic dilation of the bile duct, leading to the diagnosis of Caroli's disease. The patient commenced on fluid resuscitation, IV antibiotics Cefotaxime, and was followed by biliary drainage via stent placed by ERCP, along with administration of ursodeoxycholic acid. Conclusion: This case displays the importance of considering Caroli's disease in the differential diagnosis of patients with recurrent episodes of acute cholangitis.***

Keywords: Caroli's Disease, ERCP, Todani's Classification of Choledochal Cysts

Nomenclature:
RUQ: Right Upper Quadrant
CBD: Common Bile Duct
ERCP: Endoscopic Retrograde Cholangiopancreatography
LFTs: Liver Function Tests
CT: Computed Tomography

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I. INTRODUCTION

Caroli's disease is a rare congenital disorder characterised by cystic dilatation of intrahepatic bile ducts, with narrow or regular segments between the dilatation and the normal extrahepatic bile ducts [1].

In Caroli's syndrome, it is associated with congenital hepatic fibrosis.

In 60% to 80% of patients, it is associated with medullary sponge kidney [2]. Biliary stasis in intrahepatic ducts is a predisposing factor for cholangitis, bacterial infection, abscess formation, and septicemia.

The presentation of disease may range from abdominal pain to cholangitis to end-stage liver disease. The liver is usually enlarged.

Caroli's disease is classified as type V in Todani's classification of choledochal cysts [3], which categorises choledochal cysts into five types based on their characteristics and anatomical involvement.

The Global prevalence was less than 1 in 100,000 births [4].

Demographics:

Male-to-female ratio: 2:1 to 3:1 (Journal of Clinical Gastroenterology).

Age at diagnosis: Most patients present before age 30.

Familial occurrence: tends to be sporadic, but familial cases are reported in 10-20% of cases.

We present a case of Caroli's disease with cholangitis, highlighting the diagnostic approach, clinical course, and treatment strategies.

II. CASE PRESENTATION

A 41-year-old male came to the emergency department with a history of fever, right upper quadrant (RUQ) pain, and Itching. The pain was described as dull, intermittent, and worsening over the last two weeks. There was a notable history of intermittent jaundice over a long time, which had been previously diagnosed as uncertain cholestasis, but with no precise diagnosis.

A. Medical History

The patient's medical history was significant for recurrent jaundice and generalized body itching, which had started many years ago, during which the patient had undergone a cholecystectomy. The patient had no significant history of alcohol consumption, hepatitis B or C, or other liver diseases. There was no family history of cholestasis, biliary disorders, or genetic diseases.

B. On Examination

On physical examination, the patient appeared to be in moderate distress due to pain. Vital signs revealed fever, tachycardia, and normal blood pressure. Examination of the abdomen revealed tenderness in the right upper quadrant, with a mildly palpable liver; the liver span was 16cm, and there was no palpable mass or signs of ascites. The patient exhibited visible jaundice with yellowing of the sclera.

C. Laboratory Investigations

i. Initial Laboratory Results Showed

Elevated liver enzymes

AST: 355.2 U/L, ALT: 149.48 U/L, alkaline phosphatase: 249.68 U\L and GGT: 97.59 U\L.

Bilirubin levels: Total bilirubin of 12.08mg/dl, Direct bilirubin of 7.39mg/dl,

White blood cell count: Elevated 12x103 UI, suggesting an inflammatory response, Leukocytosis consistent with acute infection,

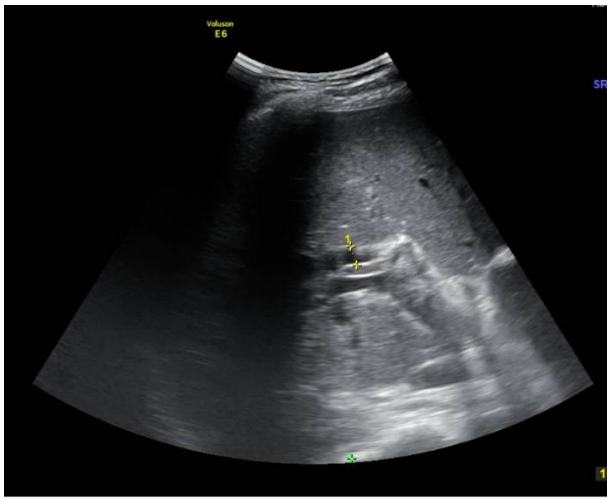
Amylase level was 51.52 U/L, and lipase levels were 58,78 U/L, excluding pancreatitis.

Inflammatory markers were elevated; ESR: 64 Mm\Hr., CRP: 24.86 Mg\dl. Electrolytes and Kidney function tests were within normal limits.

A. Imaging Studies

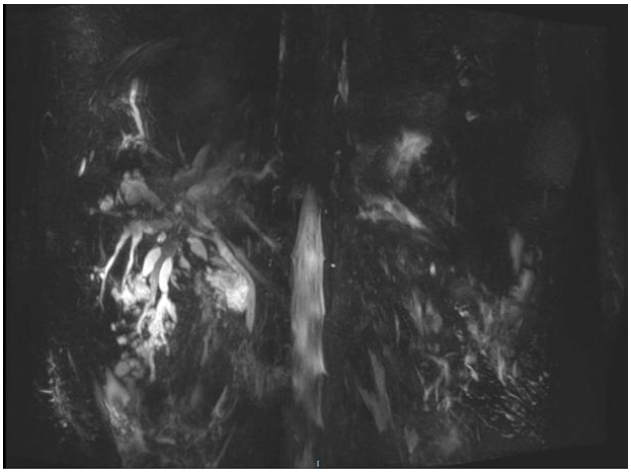
i. An Abdominal Ultrasound Revealed:

Liver was moderately enlarged in size, especially the right lobe, 22 cm, moderate right hepatic lobe intra-hepatic bile radical dilatation, and significant dilatation of the common bile duct CBD (Figure 1).



[Fig.1: Ultrasonography (US) Shows Intrahepatic Dilatation of The Right Liver Loop]

To further evaluate the biliary tree, the patient underwent MRCP (Magnetic Resonance Cholangiopancreatography), which showed: There is dilatation of the intra-hepatic ducts, worse within the right hepatic duct and the tributaries, resulting in diffuse beading and structuring/stenosis with peri-ductal tissue thickening and soft tissue-like focal areas (Figure 2). Features are consistent with a diagnosis of Caroli's disease. There was no evidence of gallstones or pancreatic lesions.



[Fig.2: Magnetic Resonance Pancreatocholangopgraphy MRCP) Shows a Characteristic of Diffuse Intrahepatic Bile Duct Dilatation]

We requested an ERCP to confirm the diagnosis and for drainage, which revealed dilated segmental intrahepatic biliary radicles, and a metallic stent was placed.

B. Diagnosis

Based on the clinical presentation, imaging findings, and recurrent episodes of jaundice and cholangitis, the patient was diagnosed with Caroli's disease presenting as acute cholangitis and recurrent jaundice [6].

III. MANAGEMENT

A. Acute Management

Upon presentation, the patient was promptly started on empiric broad-spectrum IV antibiotics to address the suspected acute cholangitis. Cefotaxime was started and later tailored based on culture and sensitivity results. Supportive care, including intravenous fluids, pain management, and antipyretics to control fever, was also initiated [7].

B. Diagnostic and Therapeutic Interventions

Endoscopic Retrograde Cholangiopancreatography (ERCP): Given the severity of cholangitis and suspicion of biliary obstruction, the patient underwent ERCP for diagnostic and therapeutic purposes. Stent placement in the bile duct was performed to ensure adequate drainage of bile from the intrahepatic ducts and prevent further cholangitis episodes [8].

C. Long-Term Management

Antibiotics: After initial resolution of the acute cholangitis episode, the patient was transitioned to oral antibiotics based on culture and sensitivity results.

i. *Biliary Drainage:* The placement of a biliary stent, following the ERCP procedure, was intended to prevent further biliary stasis and reduce the risk of recurrent cholangitis.

ii. *Surveillance and Follow-Up:* Given the chronic nature of Caroli's disease and the risk of recurrent infections, regular follow-up was scheduled, and the first visit was scheduled 1 week after discharge, then for every 3–6 months:

iii. *Liver Function Tests (LFTs:* Including bilirubin levels, Ultrasound or MRCP to monitor for changes in the biliary ducts or the development of new complications, Screening for Cholangiocarcinoma with appropriate imaging and possibly liver biopsy, as Caroli's disease is associated with an increased risk of Cholangiocarcinoma in the long term [9].

iv. *Liver Transplantation:* The patient was informed of the potential need for liver transplantation in case of progressive liver failure or the development of Cholangiocarcinoma, which can complicate Caroli's disease [10]. At the time of reporting, the patient did not require a transplant, but this remains a consideration for future management, depending on disease progression.

IV. DISCUSSION

Caroli's disease is a rare congenital condition that leads to abnormal dilatation of the biliary tract within the liver. It's usually associated with recurrent episodes of acute cholangitis due to biliary stasis in dilated cysts. This picture can pose a diagnostic challenge because it resembles other causes of cholestasis and biliary obstruction, as in our patients who had undergone cholecystectomy for the same presentation before the diagnosis was established [11,12].

Our case focuses on clinical manifestation, diagnostic approach, and management of Caroli's disease in patients with recurrent episodes of acute cholangitis.

Caroli's disease is usually diagnosed using imaging modalities such as ultrasonography (US), computed tomography (CT), or magnetic resonance cholangiopancreatography (MRCP), which reveal characteristic findings of intrahepatic bile duct dilatation and cystic formation. In our case, MRCP confirmed the diagnosis, showing typical findings of segmental bile duct dilation without distal obstruction. This is consistent with the pathophysiology of Caroli's disease, where the bile ducts are abnormally dilated due to a congenital disability in the bile duct wall.

Patients with Caroli's disease are at risk of cholangitis due to the stasis of the bile, enhancing bacterial overgrowth and infection. In our patients, the features of acute cholangitis were clear in the clinical setting. However, acute cholangitis can be managed with antibiotic therapy and biliary drainage; the underlying condition of Caroli's disease necessitates long-term management to prevent further episodes [13].

Early intervention to control the infection is crucial in improving patient outcomes. The management of cholangitis in Caroli's disease requires a precise approach, including broad-spectrum antibiotics, endoscopic or percutaneous drainage for biliary obstruction, and regular follow-up to monitor liver function [14].

Surgical intervention with hepatic resection if the disease is confined to an isolated liver segment, or liver transplantation may be considered in patients with diffuse disease and progressive liver failure or recurrent cholangitis despite conservative measures [5].

Our patient responded well to antibiotic therapy and ERCP with biliary stent drainage, with improvement in symptoms and liver function.

V. CONCLUSION

Caroli's disease should be suspected in patients with recurrent episodes of cholangitis, particularly when imaging reveals unexplained biliary dilatation. Timely recognition and precise diagnosis, using advanced imaging techniques such as MRCP, are essential for effective management and minimizing potential complications.

Clinicians should maintain a high index of suspicion for this rare condition in similar presentations to ensure timely diagnosis and optimal patient outcomes.

Further research into the long-term outcomes of patients with Caroli's disease, particularly regarding the risk of biliary malignancy, is mandatory.

DECLARATION STATEMENT

As the article's author, I must verify the accuracy of the following information after aggregating input from all authors.

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