

## Case Report

# Caroli's disease: a diagnostic challenge

**Padmini Yadav\*, Shailesh Adhikari, Narendra Pandit,  
Lalijen Awale, Keerthi Vasan,  
Sarada Khadka**

Department of Surgery, B.P. Koirala Institute of Health Sciences, Dharan, Sunsari, Nepal

**Received:** 30 August 2018

**Accepted:** 28 September 2018

### \*Correspondence:

Dr. Padmini Yadav,

E-mail: [padmeeneyadav@gmail.com](mailto:padmeeneyadav@gmail.com)

**Copyright:** © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

## ABSTRACT

Caroli's disease is a rare congenital hepatobiliary disease characterized by multifocal segmental dilatation of intrahepatic bile ducts affecting all or parts of the liver. It predisposes to biliary stasis and consequent lithiasis, cholangitis, abscesses, and septicemia. Sometimes it is difficult to diagnose and differentiate it from other similar disease conditions. 60-year-old female presented with features of recurrent cholangitis with hepatolithiasis and multiple cyst in liver, cholelithiasis was planned for cholecystectomy and drainage procedure. Patient underwent open cholecystectomy with common bile duct (CBD) exploration with T-tube placement due to intraoperative instability. Post-operative T tube cholangiogram was done. Post-operative T tube cholangiogram showed bilobar major duct cystic dilatation with predominant left lobe involvement, with few cysts containing calculi. Patient was planned for left hepatectomy with bile duct excision but patient refused to undergo aforementioned surgery. Therefore ERCP and sphincterotomy was done in view of poor performance status. Caroli's disease being a rare disease is sometimes difficult to diagnose and treat in an old age patient with bilobar involvement and poor general conditions.

**Keywords:** Caroli's disease, Diagnostic challenge, Old age, Right upper abdominal pain

## INTRODUCTION

Caroli's disease is a rare congenital malformation of the intrahepatic bile ducts characterized by duct ectasia and dilation, which may involve the biliary tract in a focal or multifocal manner. Caroli's disease is less common than Caroli's syndrome, and both are extremely rare with an approximate prevalence of less than one in 1,000,000 inhabitants.<sup>1</sup> Caroli's disease combined with congenital hepatic fibrosis and/or renal cystic disease is referred to - Caroli's syndrome.<sup>2</sup> It is currently included in group V of the Todani classification of biliary tract cystic diseases and was first described by the French gastroenterologists, Jacques Caroli et al., in 1958.<sup>3</sup> The clinical course can be asymptomatic in the first two decades from birth, in fact, typical symptoms may scarcely arise throughout the

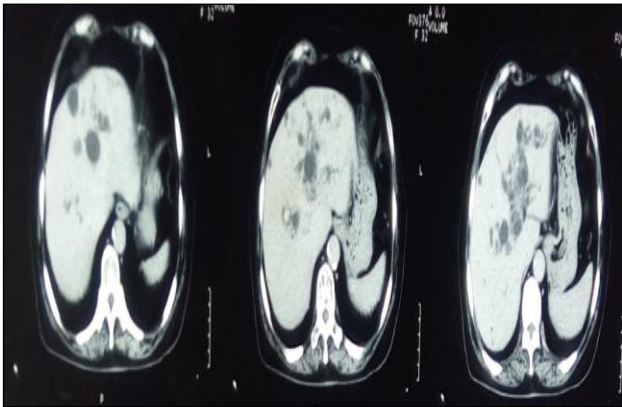
patients' life.<sup>4</sup> Caroli's disease may diffusely affected the liver or be localized to one lobe or segment. Caroli's disease predispose to biliary stasis and intrahepatic stones formation leading to cholangitis, liver abscesses, septicemia, and ultimately to secondary biliary cirrhosis.<sup>5</sup> Most used diagnosis methods are ultrasound, computed tomography (CT), magnetic resonance cholangiopancreatography (MRPC).

Endoscopic cholangiopancreatography (ERCP) can be used both as diagnostic and or therapeutic tool; a liver biopsy is rarely needed.<sup>6</sup> We describe a case of Caroli's disease presenting with right upper abdominal pain. We believe the present reports are of medical significance since they relate to patients with atypical presentations and diagnosed at an unusually late age. It serves also as a

reminder that although rarely, Caroli's disease may present later in life, have atypical presentations and be masked by non-specific clinical findings. These reports hope to add to the existing knowledge base of this very rare disease. This case is unusual in its very late presentation in 6th decade of life as opposed to previous reports in third and fourth decade of life.

## CASE REPORT

A 60-year-old female was admitted to the department of surgery (B.P. Koirala Institute of Health Sciences (BPKIHS), with complain of pain over right upper abdomen for 4 months, associated with intermittent pruritus of body.



**Figure 1: Multiple cystic dilatation of intrahepatic biliary radicle of both lobe of liver.**

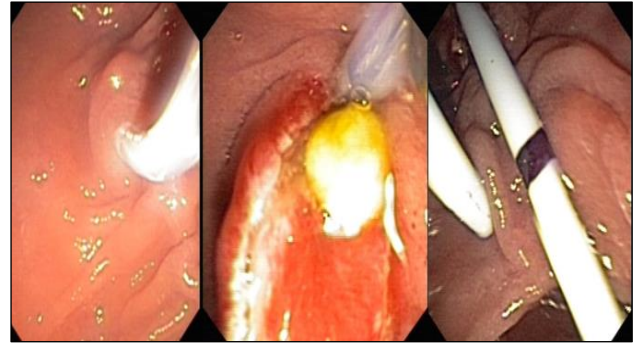
There was no history of jaundice, nausea, vomiting, chills, fever or any other comorbid factors like diabetes, hypertension. On examination, only positive finding was scratch marks over anterior abdominal wall and chest region.



**Figure 2: T tube cholangiogram showing distal CBD calculi with no flow of contrast in duodenum, with bilobar major cystic dilatation with few cysts containing calculi.**

The laboratory tests showed normal blood parameters and liver function test. The ultrasound of the abdomen

revealed hepatolithiasis with multiple hepatic cyst, cholelithiasis and bilateral renal cortical cyst. CT scan revealed multiple cystic dilatation of bilateral kidney with intrahepatic biliary radicle dilatation of both lobe of liver [Figure 1]. MRCP couldn't be done due to presence of metallic implant in left pelvis and hip region for acetabular and intertrochanteric fracture.



**Figure 3: ERCP with retrieval of retained stone and placement of stent.**

Patient was planned for cholecystectomy with drainage procedure. Intraoperatively distended gallbladder containing multiple calculi with few calculi in CBD was found and patient underwent open cholecystectomy, CBD exploration but patient went unstable with marked hypotension, so T tube placement was done. Patient was discharged after 12 days of hospital stay.



**Figure 4: Healthy scar mark in postoperative period.**

On discharge, patient's vitals were stable. She was taking orally, and wound was healthy with T-tube in situ. She then presented to surgical OPD after 1 month with distension of abdomen for 2 days associated with multiple episodes of vomiting and pain over right upper abdomen. On subsequent examination, abdomen was soft, non-tender with no organomegaly. There was healthy scar mark with T tube in situ. T tube cholangiogram was done which showed distal CBD calculi with no flow of contrast in duodenum, with bilobar major cystic dilatation with few cysts containing calculi [Figure 2].

Diagnosis of Caroli's disease left predominant was made. Patient was planned for left hepatectomy with bile duct excision but refused, hence was planned for ERCP with sphincterotomy [Figure 3].

The patient had a favorable postoperative evolution and was discharged on the 7th day [Figure 4].

## DISCUSSION

Fibrocystic liver diseases are believed to arise from the abnormal or arrested development of the embryonic ductal plates, as evidenced by the presence of persistent ductal plate-like structures in these conditions.<sup>7</sup> The ductal plate of the human fetal liver is a double-layered, wreath-like, keratin 19 (K19)-expressing bile duct structure surrounding venous branches and directly juxtaposed to the limiting plate of the hepatocytic parenchyma. After further remodeling, the ductal structures are normally separated from the limiting plate and incorporated into the portal tracts, subsequently forming mature bile ducts and ductules. The remodeling process of the ductal plates begins at the hilum and progresses to the periphery of the liver, and arrested or deranged remodeling of the ductal plate results in ductal plate malformation.<sup>8</sup> The leading symptom of the disease are cholestasis, Right upper quadrant pain and abdominal mass.<sup>9</sup> Liver function tests in Caroli's disease may be normal or there may be reversible increases during episodes of cholangitis.<sup>10</sup> Although the radiologic and pathologic features of Caroli's disease are characteristic, its clinical presentation is often non-specific.<sup>11</sup> Radiological diagnosis of Caroli's disease has, in the past, been difficult before surgery. Occasionally, mottling of contrast medium over the hepatic surface after cholangiography may indicate the intrahepatic cyst dilatations.<sup>12</sup> The cholangiographic features of Caroli's disease are well established as saccular or fusiform dilatation of the intrahepatic bile ducts. Irregular bile duct walls, strictures, and stones may be present.<sup>13</sup> The pure type of Caroli's disease sometimes may be difficult to distinguish from advanced cases of intrahepatic biliary stone disease, recurrent bacterial cholangitis (Oriental cholangiohepatitis), choledochal cysts, communicating hepatic abscesses, and primary sclerosing cholangitis.<sup>14</sup> The most serious complication of CD is the development of biliary tract cancer. The diagnosis of cancer in the setting of CD is difficult. Endoscopic biopsy, with brushing and washing yields a diagnosis in less than half of cases of bile duct cancers.<sup>15</sup> Because hepatic resection can be curative in Caroli's disease, the diagnosis is essential. Early treatment protects the patient from complications, which include calculus formation, cholangitis, abscess, and cholangiocarcinoma. Imaging is essential in planning the surgical treatment, which can consist of enterostomy, segmental or lobar hepatic resection, or liver transplantation.<sup>16</sup> Because of the slow and usually silent progress of Caroli's syndrome along with its rarity and fatal complications, it should be considered in the differential diagnosis of recurrent

cholangitis of unknown cause.<sup>17</sup> We have to consider the disease complications when choosing the treatment, if liver transplantation is not available. Cholangitis should be treated with antibiotics, while for intrahepatic lithiasis ursodeoxycholic acid may be useful. Biliary obstructions require drainage which can be performed endoscopically, radiologically or surgically, with consideration, however, of the high risk of recurrent cholangitis.<sup>18</sup> Patient was planned for left hepatectomy with bile duct excision, but patient refused to undergo aforementioned surgery. Therefore, ERCP and Sphincterotomy was done in view of poor performance status. To conclude, Caroli's disease being a rare disease is sometimes difficult to diagnose and treat in an old age patient with bilobar involvement and poor general conditions.

## CONCLUSION

Caroli's disease being a rare disease is sometimes difficult to diagnose and treat in an old age patient with bilobar involvement and poor general conditions.

## ACKNOWLEDGEMENT

Authors would like to thank Dr. Jeevan Kumar Sharma (MS Ortho, Spine fellow ISIC) for helping them in making this case report.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

## REFERENCES

- Correia PC, Morgado B. Caroli's Disease as a cause of chronic epigastric abdominal pain: two case reports and a brief review of the literature. *Cureus*. 2017;9(9):1-11.
- Mi X, Li X, Wang Z, Lin L, Xu C, Shi J. Abernethy malformation associated with Caroli's syndrome in a patient with a PKHD1 mutation: a case report. *Diagn Pathol*. 2017;12(1):61.
- Gore RM. Anomalies and anatomic variants. *Textbook of Gastrointestinal Radiology*. 1994:1851-60.
- Zhao G, Shan Q, Lv Z. Caroli's disease in two siblings. *Int J Clin Exp Pathol*. 2016;9(8):7977-81.
- Seddik H, Basr H, Aitbihi B, Adioui T, Massit H, Benkirane A. Case Report Monolobar caroli's disease: a report of three cases. *Int J Gastroenterol Hepatol Transpl Nutr*. 2017;2(1):23-5.
- Félix N, Palma F. Caroli's disease: a management of a rare condition during pregnancy. *Glo Adv Res J Med Med Sci can*. 2017;6(7):149-55.
- Desmet V. Congenital diseases of intrahepatic bile ducts: variations on the theme "ductal plate malformation." *Hepatology*. 1992;16:1069-83.
- Jang M, Lee Y, Kim H. Cholangiocarcinoma arising in Caroli's disease. *Clin Mol Hepatol*. 2014;20:402-7.

9. Tariq H. Caroli's disease or syndrome? *IJGE*. 2001;1(1):49-50.
10. Vlachogiannakos J, Potamianos S, Triantos C, Makri I, Imvrios G, Patsiaoura K. Monolobar Caroli's disease complicated by cholangiocarcinoma in a 70-year-old man, previously asymptomatic. *Gastrointest Endosc*. 2004;60(2):297-300.
11. Mall JC, Ghahremani GG, Boyer JL. Caroli's disease associated with congenital hepatic fibrosis and renal tubular ectasia: a case report. *Gastroenterol*. 1974;66(5):1029-35.
12. Bass BEM, Funston MR, Shaft MI, Hospital GS, Africa S. Caroli's disease: an ultrasonic diagnosis. *Br J Radiol*. 1977;50:366-9.
13. Levy AD, Rohrmann CA, Murakata LA, Lonergan GJ. Caroli's disease: radiologic spectrum with pathologic correlation. *Am J Roentgenol*. 2002;179(4):1053-7.
14. Miller J, Sechtin G, Campbell L, Pieters C. Imaging Findings in Caroli's Disease. *AJR*. 1995;165(8):333-7.
15. Abdalla EK, Forsmark CE. Monolobar Caroli's disease and cholangiocarcinoma. *HPB Surg*. 1999;11(C):271-7.
16. Guy F, Cognet F, Dransart M, Cercueil J-P, Conciatori L, Krausé D. Caroli's disease: magnetic resonance imaging features. *Eur Radiol*. 2002;12(11):2730-6.
17. Yonem O, Bayraktar Y. Clinical characteristics of Caroli's syndrome. *World J Gastroenterol*. 2007;13(13):1934-7.
18. Iancu C, Bodea R, Muresan T, Iancu D, Boruah P, Al Hajjar N. Diffuse form of Caroli's disease: therapeutical approach in a female patient with recurrent cholangitis. *J Gastrointest Liver Dis*. 2010;19(4):457-60.

**Cite this article as:** Yadav P, Adhikari S, Pandit N, Awale L, Vasan K, Khadka S. Caroli's disease: a diagnostic challenge. *Int Surg J* 2018;5:3750-3.