Journal of Gastroenterology, Hepatology and Endoscopy

Case Report
Published: 21 Nov, 2022

Caroli's Disease with Portal Vein Thrombosis: To Anti-Coagulate or Not?

*Li J[1] and Zhou Z[2]**

[1]Department and Institute of Infectious Disease, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, China

[2]Department of Gastroenterology, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, China

Abstract

A 32-year-old male presented with hematemesis for 5 days, which was accompanied by left lower abdominal quadrant pain and fever. The physical examination revealed mild tenderness in the lower abdominal quadrant without peritoneal irritation. The axillary temperature was 39.0°C and there was a surgical scar on the abdomen.

Keywords: Carol's disease; Caroli's syndrome; Polycystic kidney disease; Portal vein thrombosis; Cholangitis; Gastrointestinal bleeding; Endoscopic therapy

Introduction

Caroli's Disease (CD) is characterized by cystic dilatation of the intrahepatic biliary tree [1,2]. The approximate incidence of CD is less than one in 10,00,000 inhabitants [3]. Caroli's Syndrome (CS), is associated with liver cirrhosis, portal hypertension, esophageal varices, ascites and splenomegaly [4]. Congenital dilatation of the bile ducts results in bile retention, with bile duct stones and cholangitis as associated complications [5]. As a frequent and serious complication in patients with Liver Cirrhosis (LC), Portal Vein Thrombosis (PVT) may further aggravate liver damage and portal hypertension [6]. However, patients with CD and LC may carry a high risk of bleeding due to coagulopathy, leading to difficulties in implementation for anticoagulation treatments for PVT. Hence, we present the patient diagnosed with Caroli´s disease suffered from LC and PVT. The case report aims to increase knowledge with regard to this sporadic congenital disease and facilitate earlier diagnosis, timely treatment, and informed prognosis.

Case Presentation

A 32-year-old male presented with hematemesis for 5 days, which was accompanied by left lower abdominal quadrant pain and fever. The physical examination revealed mild tenderness in the lower abdominal quadrant without peritoneal irritation. The axillary temperature was 39.0°C and there was a surgical scar on the abdomen. The patient had undergone a splenectomy in 2008 because of splenomegaly and hypersplenism. It is worth mentioning that the patient had a family history of cholangiocarcinoma.

Blood routine examination revealed that the white blood cell count was 53.78 × 10^9/L, the neutrophil count was 47.13 × 10^9/L, hemoglobin was 71 g/L and the platelet count was 748.0 × 10^9/L. The biochemistry test results were as follows: Alanine aminotransferase 16 U/L, bilirubin 6.2 μmol/L, albumin 26.8 g/L, alkaline phosphatase 138 U/L, creatinine 146 μmol/L, eGFR 48.5 ml/ (min/1.73 m^2). Coagulation function testing results showed the prolonged prothrombin time and activated partial thromboplastin time (14.8 and 48.6 s, respectively). Other laboratory tests suggested an increased D-dimer levels (9.09 ug/ml) and a positive stool occult blood test.

Magnetic Resonance Cholangiopancreatography (MRCP) showed cystic dilatation of the intrahepatic ducts and polycystic kidney disease (Figure 1). The "central dot sign", which is highly specific for CD, was also seen in this patient (Figure 2). Cirrhosis and the thrombus located in the portal vein, superior mesenteric vein and splenic vein were observed on contrast-enhanced Computer Tomography (CT) image.

Due to relatively poor general condition of the patient, surgery was not performed. This patient had clinical evidence of bacterial cholangitis, so he was treated with antibiotics. Besides, the patient

OPEN ACCESS

Correspondence:
Zhenzhen Zhou, Department of Gastroenterology, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430030, Hubei Province, China, E-mail: janeychow@126.com

Received Date: *22 Oct 2022*
Accepted Date: *17 Nov 2022*
Published Date: *21 Nov 2022*

Citation:
Li J, Zhou Z. Caroli's Disease with Portal Vein Thrombosis: To Anti-Coagulate or Not?. J Gastroenterol Hepatol Endosc. 2022; 7(1): 1109.

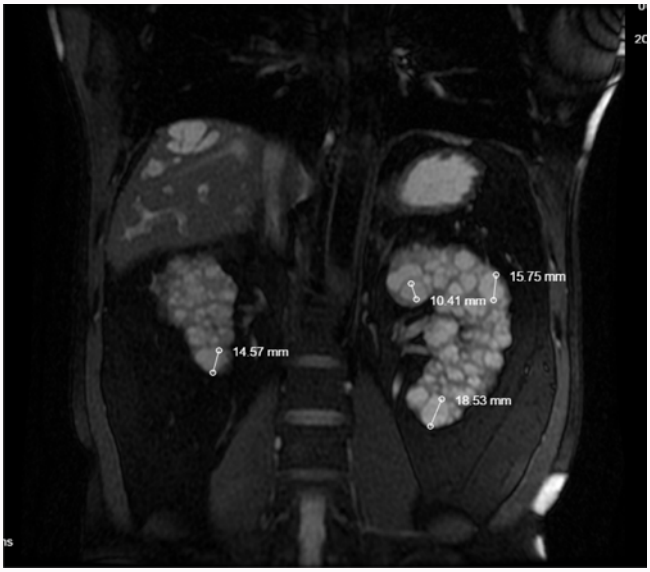

Figure 1: MRCP shows cystic dilatation of the intra-hepatic ducts and polycystic kidney.

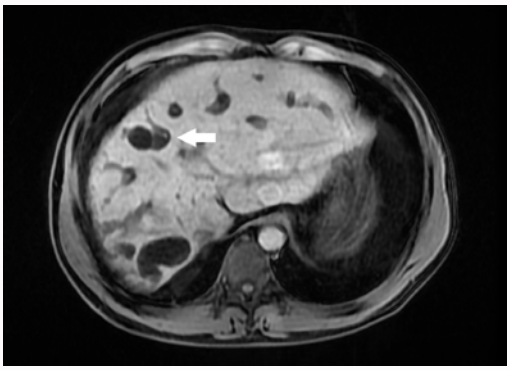

Figure 2: MRCP showed irregular cystic dilatation of intrahepatic bile ducts and "central dot sign" (arrow).

suffered from esophageal and gastric variceal bleeding. Therefore, he underwent successful treatment with Endoscopic Sclerotherapy (ES) and Endoscopic Variceal Ligation (EVL) but anticoagulatory therapy. After treatment, his biochemistry test results were improved.

Considering that this patient was already progressing to liver cirrhosis and had a family history of cholangiocarcinoma, liver transplantation is recommended in order to improve prognosis [4]. However, the patient decided not to undergo surgery due to economic difficulties. The patient has been followed-up for 9 years. There have been no other serious events as of September 2022.

Discussion

Caroli's disease is a very rare congenital disorder. Recurrent abdominal pain, jaundice and bacterial cholangitis are the most common manifestations of Caroli´s disease while Caroli's syndrome manifests as congenital hepatic fibrosis, liver cirrhosis, portal hypertension, esophageal varices [7,8]. The main clinical manifestations of this case were upper gastrointestinal bleeding, cholangitis and PVT. For one thing, hyperplasia of abnormal fibrous tissue and occlusion of small sized intrahepatic veins result in a reduced portal flow velocity, making patients more prone in developing PVT [9]. For another, this patient underwent splenectomy, which is a common cause of local vessel injury for PVT in China. It has been shown that the incidence rate of PVT after splenectomy is up to 50% in patients with liver cirrhosis [10].

Imaging, such as Magnetic Resonance Imaging (MRI), CT and ultrasound, plays a crucial role in the diagnosis of CD. The "central dot sign" is suggestive of Caroli disease [11,12]. MRCP also has been found effective for the diagnosis [13]. In addition, portal vein thrombosis appears as a non-enhancing filling defect in the portal vein lumen on CT and MRI [14].

Regarding therapeutic options, due to its rarity, no universal consensus on the best treatment planning has yet been reached. Liver resection is recommended as a primary treatment option for monolobar CD, while liver transplantation should be considered in patients with bilobar CD [15,16]. Treatments for CS are focused on alleviating symptoms caused by portal hypertension. Endoscopic injection therapy and band ligation have been widely used for the management of bleeding upper gastrointestinal tract varices. Although relatively infrequent, some researchers found that endoscopic therapy was an effective technique for the treatment of gastro-esophageal varices in a patient with Caroli's syndrome [17]. Alternatively, patients with clinical evidence of bacterial cholangitis should be treated promptly with antibiotics. Management of PVT in patients with CD and LC is controversial, especially the anticoagulation therapy. Patients with LC display an increased thrombotic risk, paradoxically associated with a bleeding tendency. In recent years, a panel of experts released the consensus for management of portal vein thrombosis in liver cirrhosis, which states that major indications for anticoagulation therapy include acute symptomatic PVT, candidates for liver transplantation, and thrombosis extension into the mesenteric veins while anticoagulation therapy should be postponed for patients with a recent history of bleeding, high-risk gastroesophageal varices and severe thrombocytopenia [18].

Conclusion

Caroli´s disease exhibits a low incidence rate worldwide, but it should be considered in patients presenting with unexplained liver cirrhosis, recurrent abdominal pain and cholangitis. The diagnosis of Caroli´s disease mainly relies on imaging examinations. Liver transplantation, as an optimal therapy for CD, is limited by a scarcity of donor organs. Based on the management experience and outcomes of the present case, endoscopic therapy is recommended for variceal bleeding in CD patients. In patients with portal vein thrombosis, anticoagulant therapy should be decided according to the specific circumstances of the patient. Antibiotics are also key allies in the treatment because patients with CD are susceptible to recurring cholangitis.

References

1. He Y, Yu Z, Chen W. Type IV choledochal cyst with polycystic kidney disease: A case report. BMC Gastroenterol. 2020;20(1):306.
2. Shi W, Yang AM. Caroli disease: An update on pathogenesis. Chin Med J (Engl). 2021;134(23):2844-6.
3. Millwala F, Segev DL, Thuluvath PJ. Caroli's disease and outcomes after liver transplantation. Liver Transpl. 2008;14(1):11-7.
4. Fahrner R, Dennler SG, Inderbitzin D. Risk of malignancy in Caroli disease and syndrome: A systematic review. World J Gastroenterol. 2020;26(31):4718-28.

5. Yuan C, Liu J, Zhou X, Wu C. Caroli's disease associated with biliary papillomatosis and cholangiolithiasis. Endoscopy. 2018;50(10):E276-8.

6. Faccia M, Ainora ME, Ponziani FR, Riccardi L, Garcovich M, Gasbarrini A, et al. Portal vein thrombosis in cirrhosis: Why a well-known complication is still matter of debate. World J Gastroenterol. 2019;25(31):4437-51.

7. Wang ZX, Li YG, Wang RL, Li YW, Li ZY, Wang LF, et al. Clinical classification of Caroli's disease: An analysis of 30 patients. HPB (Oxford). 2015;17(3):278-83.

8. Almohtadi A, Ahmed F, Mohammed F, Sanhan M, Ghabisha A, Al-Moliki L. Caroli's disease incidentally discovered in a 16-years-old female: A case report. Pan Afr Med J. 2022;41:204.

9. Zocco MA, Stasio ED, Cristofaro RD, Novi M, Ainora ME, Ponziani F, et al. Thrombotic risk factors in patients with liver cirrhosis: Correlation with MELD scoring system and portal vein thrombosis development. J Hepatol. 2009;51(4):682-9.

10. de'Angelis N, Abdalla S, Lizzi V, Esposito F, Genova P, Roy L, et al. Incidence and predictors of portal and splenic vein thrombosis after pure laparoscopic splenectomy. Surgery. 2017;162(6):1219-30.

11. Choi BI, Yeon KM, Kim SH, Han MC. Caroli disease: Central dot sign in CT. Radiology. 1990;174(1):161-3.

12. Perricone G, Vanzulli A. Education and imaging. Hepatology: "Central dot sign" of Caroli syndrome. J Gastroenterol Hepatol. 2015;30(2):234.

13. Lewin M, Desterke C, Guettier C, Valette PJ, Agostini H, Franchi-Abella S, et al. Diffuse versus localized Caroli disease: A comparative MRCP study. AJR Am J Roentgenol. 2021;216(6):1530-8.

14. Intagliata NM, Caldwell SH, Tripodi A. Diagnosis, development, and treatment of portal vein thrombosis in patients with and without cirrhosis. Gastroenterology. 2019;156(6):1582-99 e1.

15. Moslim MA, Gunasekaran G, Vogt D, Cruise M, Morris-Stiff G. Surgical management of Caroli's disease: Single center experience and review of the literature. J Gastrointest Surg. 2015;19(11):2019-27.

16. Yamaguchi T, Cristaudi A, Kokudo T, Uldry E, Demartines N, Halkic N. Surgical treatment for monolobular Caroli's disease Report of a 30-year single center case series. Biosci Trends. 2018;12(4):426-31.

17. Wang S, Xiao M, Hua L, Jia Y, Chen S, Zhang K. Endoscopic therapy for gastro-oesophageal varices of Caroli's syndrome: A case report. J Int Med Res. 2020;48(2):300060519877993.

18. Hepatobiliary Disease Study Group, C.S.o.G.C.M.A. Consensus for management of portal vein thrombosis in liver cirrhosis (2020, Shanghai). J Dig Dis. 2021;22(4):176-86.