Caroli's Syndrome in a Post Renal Transplant Patient: Case Report and Review of the Literature

Muhammad Z. Bawany, Osama Alaradi[1], Ali Nawras[2]

Department of Medicine, [2]Division of Gastroenterology and Hepatology, University of Toledo Medical Center Toledo Ohio, [1]Division Gastroenterology and Hepatology, Henry Ford Hospital, Detroit, MI, USA

Address for correspondence:
Dr. Muhammad Z. Bawany, University of Toledo Medical Center, 3000 Arlington Avenue Mail Stop 1150, Toledo, OH, USA 43614.
E-mail: muhammad.bawany@utoledo.edu

ABSTRACT

Caroli's syndrome is characterized by bile duct ectasia in association with hepatic fibrosis. It is usually transmitted in an autosomal recessive fashion and has been well documented to be associated with autosomal recessive polycystic kidney disease and occasionally with autosomal dominant polycystic kidney disease. However, there has been only few case reports published with Caroli's syndrome diagnosed postrenal transplantation.

Key Words: Bile duct ectasia, Caroli's syndrome, liver transplant

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Caroli's syndrome is characterized by bile duct ectasia in association with hepatic fibrosis.[1,2] Caroli's disease is a congenital disorder characterized by multifocal, segmental dilatation of large intrahepatic bile ducts. The condition is usually associated with renal cystic disease of varying severity. If it is only characterized by bile duct ectasia without apparent hepatic abnormalities then it is called Caroli's disease, as opposed to Caroli's syndrome in which there is hepatic fibrosis. It is usually transmitted in an autosomal recessive fashion and has been well documented to be associated with autosomal recessive polycystic kidney disease (ARPKD) and occasionally with autosomal dominant polycystic kidney disease (ADPKD).[3] However, there has been only few case reports published with Caroli's syndrome diagnosed postrenal transplantation.

CASE REPORT

We report a case of 52-year-old Caucasian male who was referred to us for the evaluation of elevated liver enzymes.

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His medical history was significant for renal transplantation 13 years ago outside our institution secondary to end-stage renal disease due to ADPKD. He had no history of smoking or alcohol use. At presentation, his vital signs were stable except for the temperature of 99 F. Rest of the physical examination was unremarkable.

At his first presentation liver function tests (LFTs) showed albumin of 3.7 g/dl, bilirubin 1.7 mg/dl with direct bilirubin 0.6 mg/dl, alkaline phosphatase 188 IU/l, ALT 50 IU/l, and AST 40 IU/l. Patient's prior LFT's were normal. Ultrasound of the liver revealed dilated biliary tree with sludge in gallbladder. CT scan of abdomen revealed marked intrahepatic biliary dilatation [Figure 1] with common bile duct measuring 1 cm. Patient had ERCP, which showed dilatation and stricturing of the intrahepatic biliary tree and multiple stones within biliary tree that were removed after performing sphincterectomy.

Subsequently, the patient presented to our institution with a complaint of repeated bouts of low-grade fever associated with right upper quadrant abdominal pain and mildly elevated liver enzymes. CT scan of abdomen revealed persistent biliary tree dilatation. ERCP revealed multiple saccular dilatations within the intrahepatic biliary tree (left more than right) [Figure 2]. Multiple balloon sweeps were performed; several stone fragments with sludge were removed. Patient continued to have recurrent cholangitis, therefore, he was eventually evaluated for liver transplant

and finally received orthotopic liver from a deceased donor. Pathology of patient's native liver revealed cavernous biliary ectasia with broad bands of fibrosis consistent with Caroli's syndrome.

DISCUSSION

The molecular pathogenesis of Caroli's syndrome is incompletely understood. However, frequent association of Caroli's syndrome with ARPKD suggest role of fibrocystin (the protein encoded by the ARPKD gene PKHD1) in the pathogenesis.[4] Presentation of caroli's syndrome varies depending on the age of onset and type of inheritance. Caroli's syndrome can also lead to cirrhosis of the liver in some patients,[5] as opposed to our patient, who did not have signs of cirrhosis.

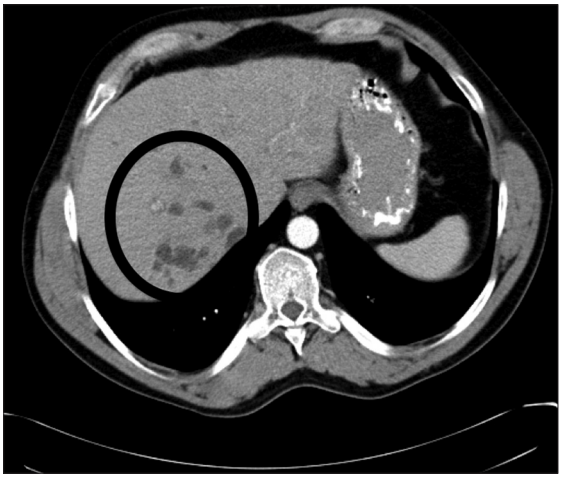

Figure 1: CAT scan showing saccular dilatation of biliary tree mostly in the right lobe of the liver

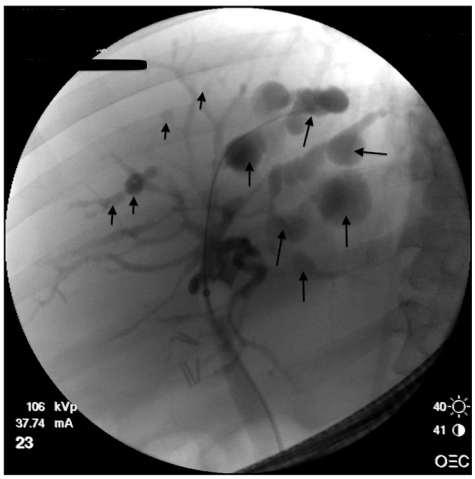

Figure 2: ERCP showing diffuse saccular dilatation of biliary tree (left>right)

To our knowledge, there are only five cases reported of Caroli's disease postsolid organ transplant in English literature,[6-10] all of the solid organ transplants were kidney transplant, just like in our patient. Review of these articles showed the primary cause of renal transplant was either ARPKD (three out of five) or ADPKD (two out of five) [Table 1].

Treatment is basically supportive and needs to be individualized. Conservative therapy with antibiotics might fail because of immunosuppressive state in post-transplantation patient, which was considered to be the cause of failure in three out of five patients reported with the history of renal transplantation. Surgery is an option for those who develop recurrent cholangitis. Liver transplantation is the treatment of choice for refractory cholangitis and diffuse involvement of liver,[11] like in our patient. However if the disease is localized in one segment or lobe of the liver, lobectomy then can be considered.[12]

CONCLUSION

Caroli's syndrome in a patient with a history of renal transplantation carries high risk of recurrent or refractory cholangitis due to immunosuppressive state, therefore, surgical therapy, i.e., liver transplantation, needs to be considered early in the course of treatment if the conservative therapy fails.

Learning point: Although, there is enough literature about Caoli's syndrome in autosomal recessive polycystic kidney disease; there are only two reports of Caroli's syndrome in patients with autosomal dominant poycystic kidney disease diagnosed after renal transplantation.

Table 1: Caroli's syndrome in postsolid organ transplant

Age	Sex	*AR vs AD	Liver involvement	Type of treatment	Follow up in months
13	Male	AR	Diffuse	Conservative	–
25	Female	AR	Diffuse	Transplant within 4-5 months	48
52	Male	AR	Diffuse	Conservative	–
54	Male	AD	Diffuse	Choledocoduodenal anastomosis	18
73	Male	AD	Left>right lobe	Left lateral segmentectomy with cholecystetomy	–

*AR: Autosomal recessive polycystic kidney disease, *AD: Autosomal domiannt polycystic kidney disease

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