Monolobar Caroli's Disease: Report of Two Cases within a Family

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Caroli's disease is a congenital disorder characterized by segmental communicating saccular dilatation of larger intrahepatic bile ducts. This malformation may occur in association with autosomal dominant or recessive polycystic kidney disease of varying severity. This study reports the successful management of two cases with monolobar Caroli's disease within a family by partial hepatic resection. Case one was a 74 year-old female, who had a known diagnosis of autosomal dominant polycystic kidney disease, and clinically manifested repeated episodes of bacterial cholangitis and septicemia. Abdominal computed tomography and magnetic resonance cholangiopancreatography confirmed the presence of saccular dilatation of intrahepatic bile ducts confined to the left lobe, hepatolithiasis and choledocholithiasis. Case two was a 43 year-old male, who was the son of case one, had autosomal dominant polycystic kidney disease, and clinically manifested acute bacterial cholangitis. Abdominal computed tomography demonstrated the presence of dilated left intrahepatic bile ducts, hepatolithiasis and choledocholithiasis. Both patients underwent cholecystectomy, choledocholithotomy and left lateral segmentectomy and the post-operative recovery was uneventful in both cases. There was no recurrence of cholangitis during a 14-month and a four-month follow-up, respectively. The diagnosis of Caroli's disease was established by imaging studies. Magnetic resonance cholangiopancreatography is preferred and it could provide a noninvasive, safe and accurate diagnosis of Caroli's disease. Partial hepatic resection may be curative in patients with Caroli's disease confined to a single lobe or segment of the liver.

Key words: Caroli's disease, autosomal dominant polycystic kidney disease, hepatic resection

Caroli's disease is a rare congenital disorder characterized by segmental communicating saccular dilatation of larger intrahepatic bile ducts. This malformation may occur in association with either renal cystic disease of varying severity or congenital hepatic fibrosis.[1,2] When it is associated with congenital hepatic fibrosis, it is named Caroli's syndrome. Both Caroli's disease and Caroli's syndrome have been described in the same family. In cases, Caroli's disease is transmitted in an autosomal recessive inheritance and is associated with autosomal recessive polycystic kidney disease (ARPKD).[1] Rare cases associated with autosomal dominant polycystic kidney disease (ADPKD) have been reported.[3] Caroli's disease may be multifocal and diffuse, or localized to a single lobe or segment of the liver. In the diffuse type, saccular dilatation of the segmental bile ducts affects the whole intrahepatic biliary tree, and in the localized type, they are often

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