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CASE REPORT

Monolobar Caroli's disease with renal cysts: Case report

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KEYWORDS

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Abstract Caroli's disease is autosomal recessive, non-obstructive dilatation of intrahepatic biliary ducts. The exact etiology is unclear. Two variants of Caroli's disease are well known-simple; in which bile ducts are dilated without hepatic fibrosis and the second type which is associated with congenital hepatic fibrosis along with its sequelae, also known as Caroli's syndrome. Simple Caroli's disease without hepatic fibrosis is quite rare. The importance of recognizing this disease as a cause of biliary stasis is its frequent association with lithiasis, recurrent cholangitis, liver abscesses, cirrhosis and cholangiocarcinoma.

1. Introduction

Caroli's disease or communicating cavernous ectasia of the intrahepatic bile ducts is a rare autosomal recessive disorder which occurs due to the abnormal development of the intrahepatic bile ducts secondary to intrauterine vascular insult (1). It is characterized by non-obstructive dilatation of the intrahepatic biliary radicles. Caroli's disease may be diffuse or may be localized to a lobe or a segment of the liver, usually the left (2). The disease is recognized in its two forms-simple (classic); in which the intrahepatic biliary duct dilatation is seen without hepatic fibrosis and Caroli's syndrome (fibrous); in which there is hepatic fibrosis in addition that may be accompanied by the cascade of events that follow portal hypertension. In contrast enhanced computed tomography (CECT), the 'central dot sign' representing the vascular bundle in dilated biliary duct is pathognomonic. Early recognition of the disease and its

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complications is required for timely management of the patient.

2. Case report

A 27-year-old female presented with intermittent right upper quadrant pain for the past five months. There was no history of fever or jaundice. The blood tests were unremarkable except for slightly raised levels of SGOT and SGPT. The general physical examination was also normal with no hepatomegaly or splenomegaly. On ultrasound of the abdomen, multiple tortuous tubular anechoic structures were seen in the right lobe of the liver. These were communicating with the right biliary tree (Fig. 1a). On Doppler imaging, the right portal vein branches were accompanying these dilated channels and some of these dilated channels were also showing traversing septae (Fig. 1b). The left biliary ducts, common hepatic duct, common bile duct and gall bladder were normal. Contrast enhanced CT abdomen showed saccular dilatation of biliary radicals which were confined to the right lobe (Fig. 2a and b). A small enhancing dot representing portal vein branch was seen in the center of some of these cystic dilatations giving the typical 'central dot' sign (Fig. 3). The left and extrahepatic biliary tree and the gall bladder were normal (Fig. 4). Both the kidneys showed multiple small simple cysts (Fig. 5a and b). There was no evidence of hepatic mass or portal hypertension on both Doppler and CT imaging. Esophageal varices, splenomegaly or ascites were not seen. The radiographic imaging was consistent with simple Caroli's disease in localized form. Because of increased risk of cholangiocarcinoma, the patient was offered hepatic lobectomy as the treatment which the patient had denied at present. So the patient is managed with

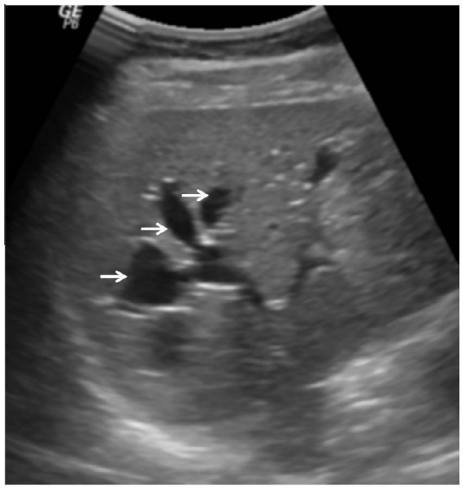

Fig. 1a Gray scale ultrasound in a 27-year-old female with monolobar Caroli's disease shows dilated, anechoic, intercommunicating biliary channels in the right hepatic lobe (white arrow).

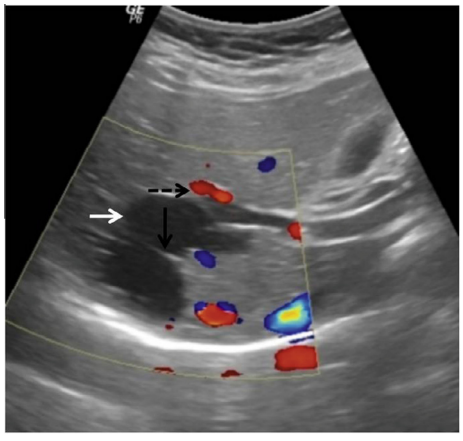

Fig. 1b Doppler ultrasound shows portal vein branches (dashed black arrow) accompanying these dilated biliary ducts (white arrow). A fibrovascular septa is also seen traversing one of these dilated ducts (black arrow).

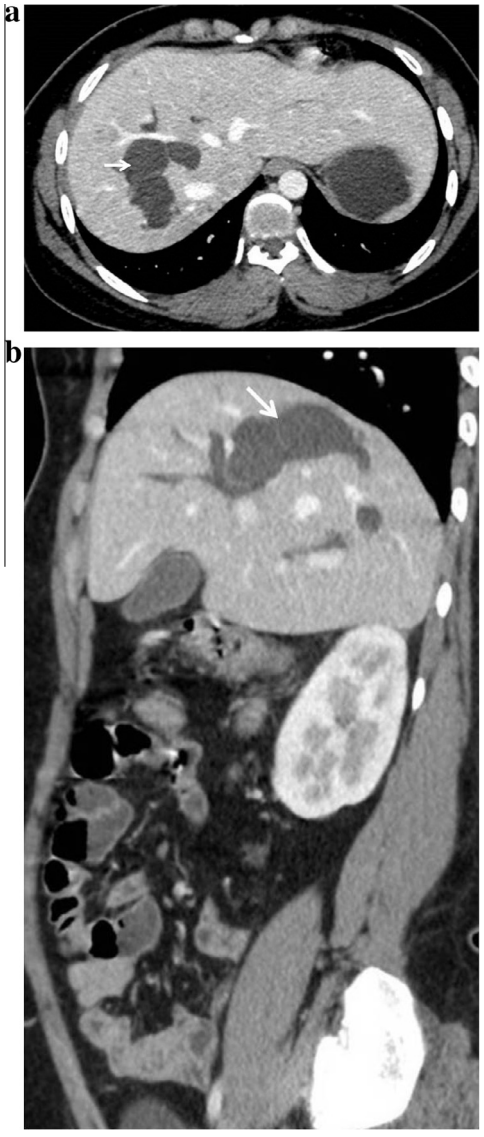

Fig. 2 (a and b) Contrast enhanced CT images (axial, sagittal) show hypoechoic saccular dilatations in right lobe of the liver communicating with right hepatic duct consistent with dilated biliary radicles (white arrow).