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Incidentally detected polycystic kidney and Caroli's disease

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Congenital cystic dilatation of the intrahepatic biliary ducts [Caroli's disease (CD)] has been seldom seen in clinical practice. It is often associated with autosomal recessive polycystic kidney disease and congenital hepatic fibrosis. Here we report an image of a female asymptomatic patient with CD and polycystic kidney disease.

A 51-year-old female patient with a one month history of pruritus visited our hospital for a routine health check up. She was undergoing treatment with her dermatologist for pruritus only with a prescription for local application. Her past medical history was unremarkable. Clinical examination was normal with only scratch marks. Laboratory investigations revealed normal hemogram, mildly elevated serum creatinine 1.3 (0.6-1.0) and normal electrolytes. Liver function test showed normal bilirubin with alanine aminotransferase 64 (5-30), AST 55 (5-30) and alkaline phosphatase 260 (30-120) gamma-glutamyltransferase 314 (5-30) with normal albumin level. All viral serology was normal. MRI/MRCP showed enlarged kidneys with parenchymal replaced by cysts along with hepatic cyst communicating with biliary tree as shown in Fig. 1.

Cystic lesions of the liver and kidney are increasingly being detected with the availability of newer imaging techniques [1]. Fibropolycystic disease can exist singly or in combination. They are usually inherited and consist of polycystic disease, congenital hepatic fibrosis, CD and choledochal cysts. CD is associated with renal involvement in up to 60%, and includes medullary sponge kidney disease or polycystic kidney disease [2]. CD can be asymptomatic or can present with cholangitis, intra hepatic stone formation or portal hypertension. CD is characterized by multifocal segmental dilatation of intrahepatic bile ducts affecting all or parts of the liver. Two forms of CD are described, the pure form and that associated with periportal fibrosis. The disease may diffusely affect the liver or be localized to one lobe or segment. Less than 20% of all reported cases of CD are monolobar type. Here we present an interesting image of CD localized mainly in the left lobe.

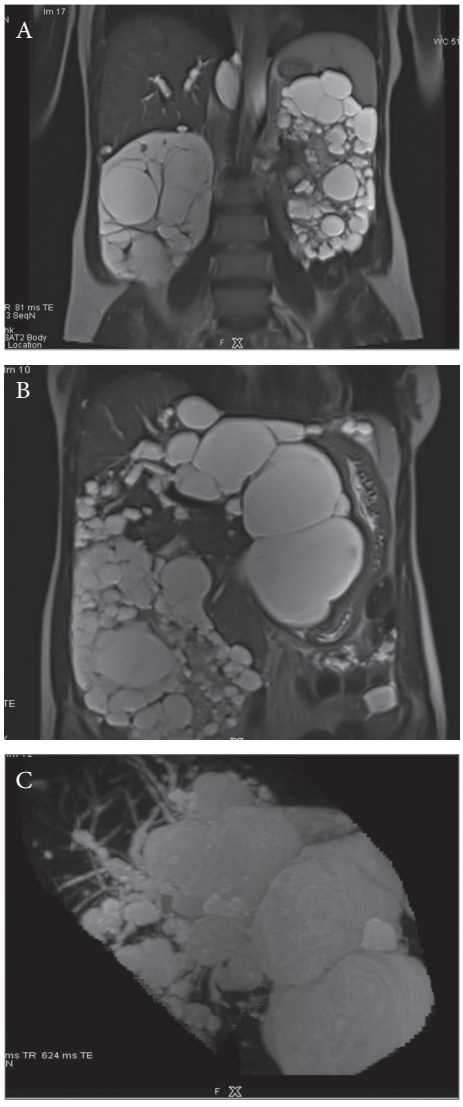

Figure 1 (A) Right and left hepatic duct with cysts in the kidneys (B) Large intrahepatic cysts in left lobe of liver with right kidney cysts (C) Cystic dilatation of intrahepatic duct due to Caroli's disease

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