

Renal cysts in Caroli's disease

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A thirty-year-old male presented with vague upper abdominal pain. There were no urinary symptoms. The general and systemic examination was unremarkable. The hemoglobin was 8.8 g/dl, total leukocyte count was 11 200/cumm, blood urea was 25 mg/dl and serum creatinine was 0.9 mg/dl. The urine examination was normal. Contrast-enhanced computed tomography (CECT) of abdomen revealed dilated intra hepatic biliary ducts and presence of "central dot" sign representing intraluminal portal vein in dilated intrahepatic bile ducts in the right lobe of liver suggestive of Caroli's Disease [Figure 1]. Axial CECT showed multiple cysts in the lower poles of both kidneys [Figure 2].

Caroli's disease is a rare, complex autosomal recessive congenital disorder characterized by multiple focal segmental saccular dilatations of the intrahepatic bile ducts, a predisposition to biliary calculi and cholangitis and an association with various forms of cystic renal disease. The disease is common in childhood and in the second to third decades of life. Caroli's disease can be associated with varying degrees of renal cysts, renal tubular ectasia, medullary sponge kidneys, autosomal recessive kidney disease, nephrocalcinosis, interstitial fibrosis and renal failure.^[1-3] To differentiate from autosomal dominant polycystic kidney disease where-in the liver and kidneys are affected, the presence of a central dot sign in CECT helps in pointing the diagnosis toward Caroli's disease. In our evaluation, we found renal cysts in both the kidneys.

References

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DOI: 10.4103/0971-4065.62089



Figure 1: Contrast-enhanced computed tomography of abdomen showing dilated intrahepatic biliary ducts with air foci and presence of "central dot" sign representing intraluminal portal vein in dilated intrahepatic bile ducts in right lobe of liver

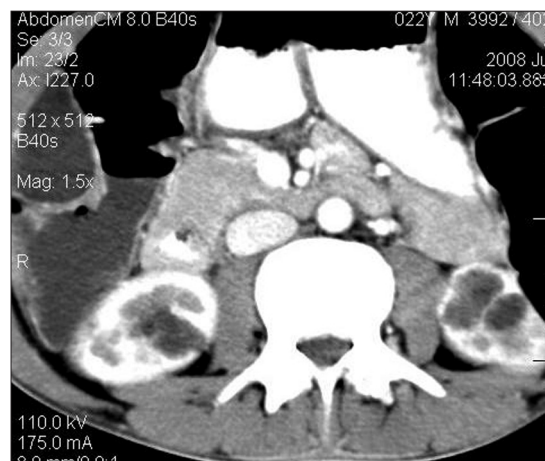


Figure 2: Axial contrast-enhanced computed tomography showing multiple cysts in the lower poles of both kidneys

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Source of Support: Nil, Conflict of Interest: None declared.