Clinical Quiz

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An infant with renal and hepatic cysts

Clinical Presentation

A 2-week-old Saudi boy presented with a history of abdominal distension. He was a product of full term baby with normal vaginal delivery. His birth weight was 3.1 kg. The antenatal history was normal. The family revealed that his cousin had congenital hepatic fibrosis. His past medical history was otherwise, noncontributory. His physical examination demonstrated mild abdominal distension. No hepatosplenomegaly was noted, otherwise, systemic examination was unremarkable. The laboratory findings revealed a normal complete blood count. The electrolytes, liver function tests, renal function, and coagulation study were normal. The abdominal ultrasound showed increased echogenicity in both kidneys. The infant underwent magnetic resonance cholangiopancreatography (MRCP) (**Figure 1**).

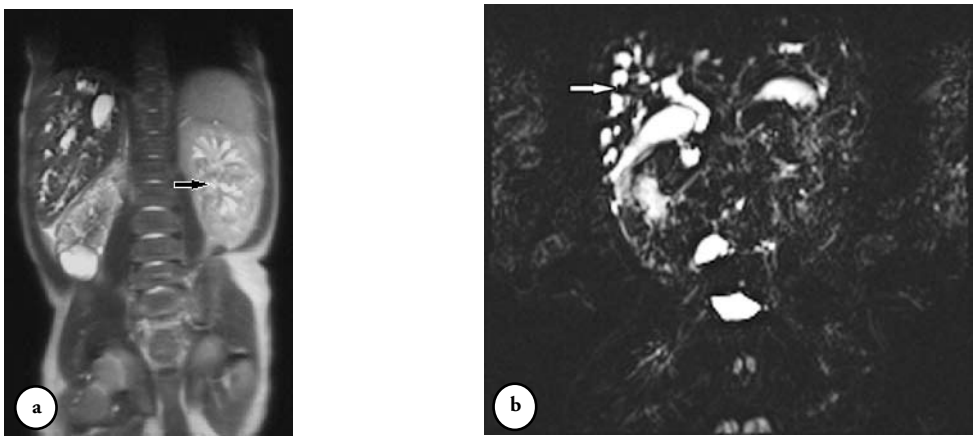

Figure 1 - The magnetic resonance cholangiopancreatography of the: **a**) kidney and **b**) liver.

Clinical Quiz

Questions

1. Describe the abnormalities in Figure 1.

2. What is the diagnosis?

3. What are the complications?

4. What is the treatment option?

Answers and Discussion

1. ***Abnormalities.*** **Figure 1a** of MRCP showed abnormalities in the kidney with demonstrated multiple cortical cysts (black arrow).The largest one measured 1.4 cm x 1.4 cm. **Figure 1b** of MRCP showed abnormalities in the liver with diffused dilatation of intrahepatic ducts, mainly seen in the right lobe of liver (white arrow). One of the cysts demonstrated central dot sign, and it was communicating to the duct. The common bile duct is mildly dilated.
2. ***Diagnosis.*** The diagnosis is Caroli disease (CD). The CD is a rare congenital disorder characterized by multifocal, segmental dilatation of large intrahepatic bile ducts.[1] The CD or Caroli's syndromes (the more common variant) are transmitted in an autosomal recessive fashion, and are associated with autosomal recessive polycystic kidney disease (ARPKD).[1] The CD and Caroli syndrome are part of a broader spectrum of diseases linked by a common pathogenetic mechanism called ductal plate malformation.[1]
3. ***Complications.*** The complications of CD include cholangitis, sepsis, gallstone, portal hypertension, liver failure, and high risk of cholangiocarcinoma.[2]
4. ***Treatment.*** The management of CD is largely supportive.[3] Ursodeoxycholic acid can decrease the frequency of cholelithiasis. Broad-spectrum antibiotic coverage is indicated in cases of cholangitis. Fat soluble vitamin supplementation can improve cholestasis and liver transplantation is indicated in decompensated cirrhosis.[3]

References

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