US AND CT OF CAROLI DISEASE: REPORT OF TWO CASES

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ABSTRACT

Caroli disease is a rare congenital disorder with multiple cystic dilatation of the intrahepatic bile ducts. We report two pediatric patients with cystic dilatation of intrahepatic bile ducts accompanying congenital hepatic fibrosis. CT scans of the liver showed tiny dots with strong contrast enhancement inside dilated intrahepatic bile ducts ("central dot signs"). These dot signs in CT scans corresponded to intraluminal portal veins in sonograms. Ultrasonography and CT scans are very effective noninvasive methods to diagnose this disease.

INTRODUCTION

Caroli disease is a disease of segmental nonobstructive dilatation of the intrahepatic bile ducts. It is a rare congenital abnormality, in which there can be many malformations of the hepatobiliary system. These range from isolated cystic dilatation alone of the intrahepatic bile ducts to hepatic fibrosis with cystic dilatation of the intrahepatic bile ducts.[1] We describe two children with the latter.

CASE REPORTS

Case 1.- A 10-month-old girl had fever, anemia, and a distended abdomen. Her temperature was 39°C. On palpation, the liver was enlarged. Laboratory studies showed a hemoglobin concentration of 9.3 g/dL, a white blood cell count of 11,900, and an alkaline phosphatase of 386 IU/L (normal 23-98). Creatinine was normal. Abdominal ultrasonograms showed hepatomegaly and saccular dilatation of the intrahepatic bile ducts (Fig. 1A). Increased echogenicity of the renal parenchyma of both kidneys was also noted. Abdominal CT scans showed hepatomegaly. Saccular dilatation of intrahepatic bile ducts, more severe in the right hepatic lobe, was seen. Central dot signs and bridge formation across the dilated lumina of bile ducts were seen (Fig. 1B). A slightly increased density of the renal medullas in noncontrast CT scans and striation of the renal medullas in contrast CT scans were seen (Fig. 1C). Due to persistent cholangitis, the patient underwent hepatectomy of the right lobe, after which the fever subsided. The pathological specimen showed saccular dilatation of intrahepatic bile ducts consistent with Caroli disease, active cholangitis, and congenital hepatic fibrosis. A microscopic study showed portal radicles surrounded by dilated bile ducts (Fig. 1D).

Case 2.- A 6-year-old boy had had a distended abdomen for 5 years. Physical examination revealed hepatomegaly, ascites, and dilatation of the superficial veins of the abdomen. Laboratory studies showed a hemoglobin concentration of 5.6 g/dL, a white blood cell count

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of 9,000, and an alkaline phosphatase of 188 IU/L (normal 23-98). Creatinine was normal. Abdominal ultrasonograms showed hepatomegaly, saccular dilatation of the intrahepatic bile ducts, intraluminal portal vein (Fig. 2A), increased echogenicity of the renal parenchyma of both kidneys, and splenomegaly. Abdominal CT scans showed hepatosplenomegaly and saccular dilatation of intrahepatic bile ducts, more severe in the right lobe (Fig. 2B). Central dot signs and bridge formation across the dilated lumina of bile ducts were seen. A slightly increased density of the renal medullas in noncontrast CT scans and striation of the renal medullas in contrast CT scans were seen (Fig. 2C). This patient did not undergo surgery, because he did not have persistent cholangitis or any other complications, except for portal hypertension.

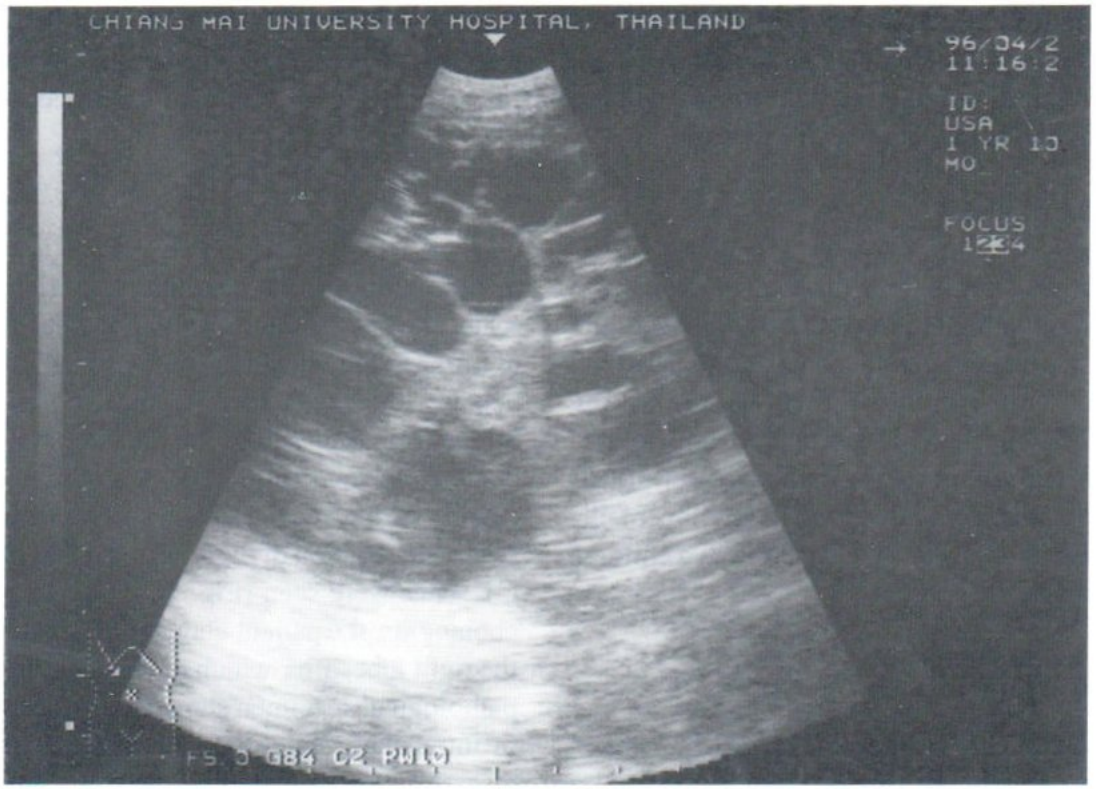

Fig. 1 Case 1
Fig. 1A Oblique sonogram of the liver shows saccular dilatations of the intrahepatic bile ducts.

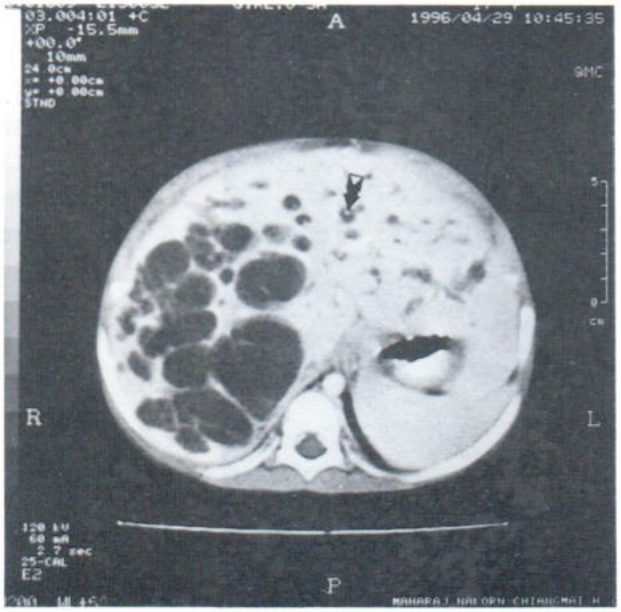

Fig. 1B Contrast CT scan shows saccular dilatation of the intrahepatic bile ducts. Several tiny dots are seen in dilated bile ducts (arrow). Bridge formation (multiple septa) across the dilated bile ducts are seen in right hepatic lobe.

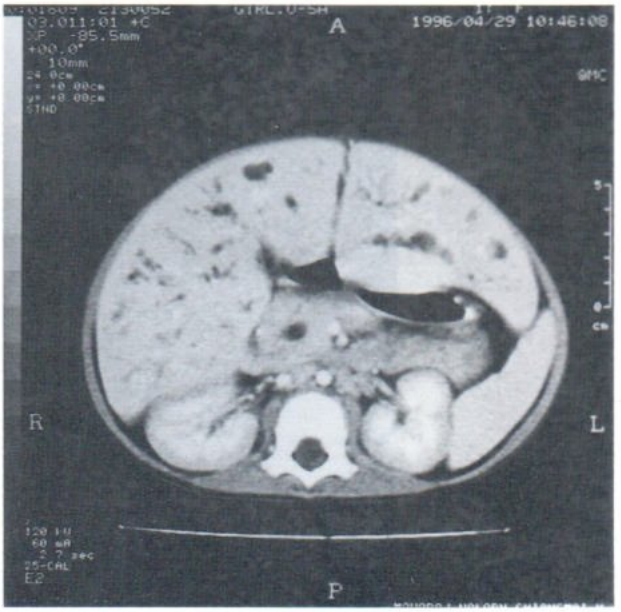

Fig. 1C Contrast CT scan shows striation of the renal medullas in both kidneys, hepatomegaly, saccular dilatation of intrahepatic ducts, and mild dilatation of the common bile duct.

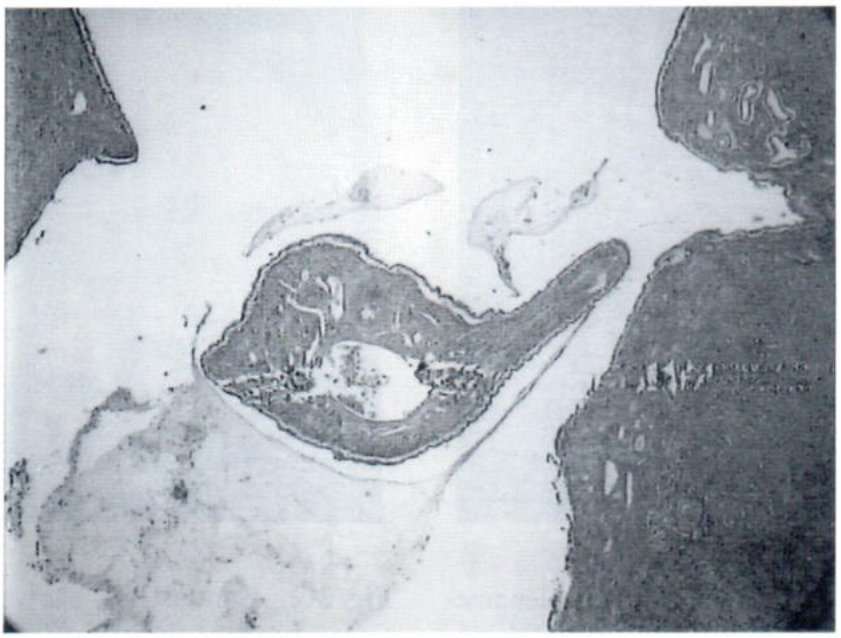

Fig. 1D Microscopic study (X40) of the liver shows dilated bile ducts surrounding a portal radicle.

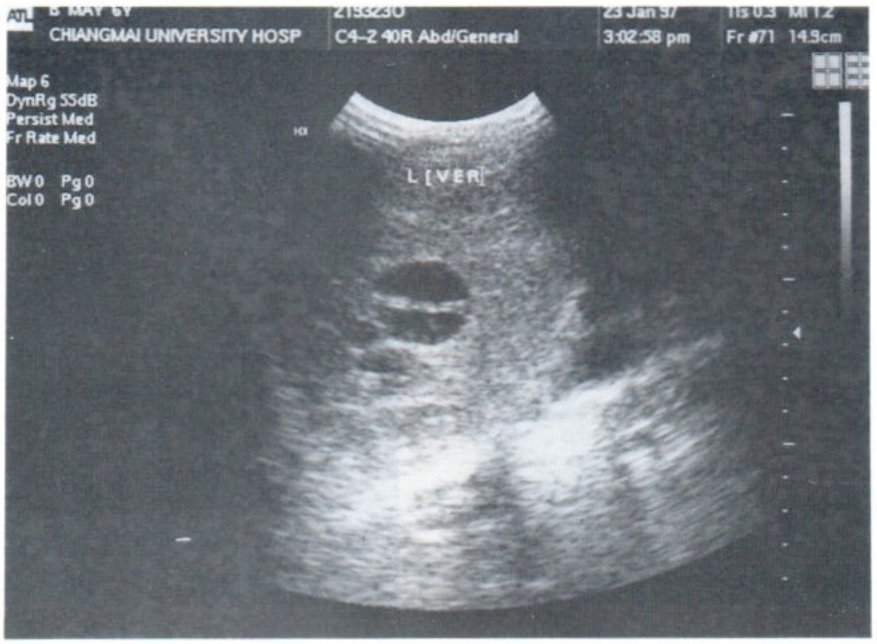

Fig. 2 Case 2

Fig. 2A Transverse sonogram of the liver shows intraluminal portal vein.

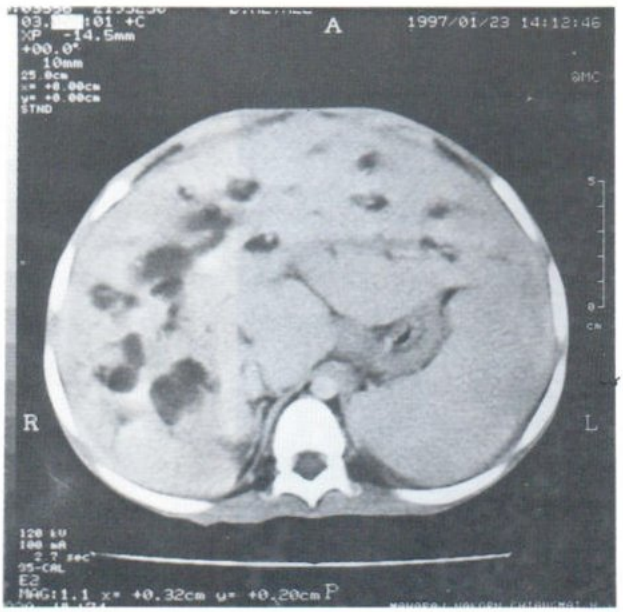

Fig. 2B Contrast CT shows hepatosplenomegaly and saccular dilatation of the intrahepatic bile ducts.

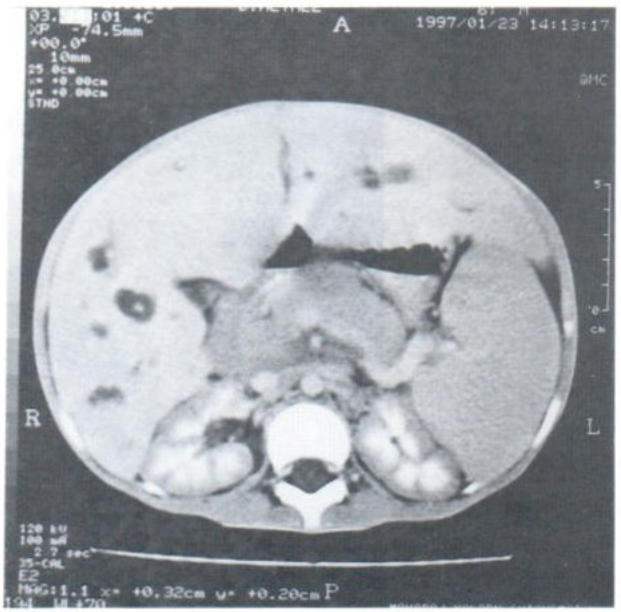

Fig. 2C Contrast CT shows hepatosplenomegaly, saccular dilatation of the intrahepatic bile ducts, central dot sign, and striation of renal medullas of both kidneys.

DISCUSSION

In Caroli disease there are congenital bile duct cysts classified as type V in Todani's system.[2] Although this disease is present from birth, most patients usually seek medical assistance for chronic recurrent upper abdominal pain or recurrent cholangitis as children or young adults. Caroli disease is associated with intrahepatic biliary calculi, hepatic abscesses, cholangiocarcinoma, hepatic fibrosis or cirrhosis.[3] There is also an association with benign renal tubular ectasia and renal cysts.[4] Patients with associated hepatic fibrosis may develop portal hypertension, while those with no hepatic fibrosis do not. Our first patient did not have portal hypertension but did have hepatic fibrosis, which was confirmed by histopathological examination. The second one had splenomegaly and dilatation of abdominal superficial veins consistent with portal hypertension. This is consistent with hepatic fibrosis although this could not be confirmed since there was no surgery. Both had evidence of tubular ectasia of the kidneys on US and CT, but the patients did not have renal biopsies. Both had normal creatinine levels.

Noninvasive imaging methods for Caroli disease are ultrasonography (US), computed tomography (CT), and nuclear scintigraphy. Invasive imaging methods for Caroli disease are endoscopic retrograde cholangiopancreatography (ERCP) and percutaneous transhepatic cholangiography (PTC). Nuclear scintigraphy, ERCP, and PTC cannot demonstrate associated abnormalities outside the biliary tract such as renal abnormalities, while US and CT can.

US and CT have been shown to be useful in detecting segmental dilatation of intrahepatic bile ducts. Marchal et al.[5] described sonographic features of Caroli disease in three patients. Besides the well-known characteristic of nonobstructive segmental dilatation of the bile ducts, they also observed portal radicles that were partially or completely surrounded by dilated bile ducts. There were similar findings in the histopathological examination of the liver of our first case and in the ultrasonogram of our second case.

Choi et al.[6] reported two adults with Caroli disease with CT scans having portal radicles surrounded by dilated intrahepatic bile ducts (the central dot sign). There were tiny dots in dilated intrahepatic bile ducts in CT scans, which enhanced strongly. These dots were not in the dependent portion of the dilated bile ducts. These intraluminal dots in CT scans corresponded to intraluminal portal veins in sonograms, which indicated that the portal radicles were surrounded by dilated bile ducts. The portal radicles were so completely enveloped by abnormal bile ducts that they appeared to lie within the lumina of the bile ducts. Bridge formation across dilated intrahepatic ducts resembling internal septa in intrahepatic ducts was also seen. This is consistent with walls of insufficiently resorbed, malformed ductal plates that surround the portal radicles.

The central dot sign was also reported in a patient with periductal cysts;[7] however the cysts did not communicate with the dilated ducts, which excluded Caroli disease.

CONCLUSION

Our cases demonstrate that US and CT scans are good noninvasive imaging methods for revealing the abnormalities of the hepatobiliary system and other organs in patients with Caroli disease. The abnormalities seen in both methods correlated well with the pathological specimens.

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