Carolī’s Disease: Hepatic Arterial Color Doppler Signals in the Communicating Dilated Bile Ducts

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— Abstract —

Three siblings with congenital dilatation of the intrahepatic bile ducts (Carolī’s disease) are presented. Bile duct pathology was associated with congenital hepatic fibrosis and polycystic renal disease in all three patients. On color Doppler imaging (CD imaging), multiple small color Doppler signals were observed in or near the vascular radicles within the dilated bile ducts, besides other well-known sonographic findings such as bile duct dilatations, biliary calculi. Dopper frequency spectral analysis confirmed all these color Doppler signals as arterial origin in all patients, showing pulsatile wave pattern. Although portal venous radicles are well known in conventional sonograms or computed tomography(CT), continuous wave patterns were not detected in all patients. In addition to previously reported sonographic findings about Carolī’s disease, color Doppler signals showing arterial wave pattern in or around the portal venous radicles within dilated ducts are another helpful diagnostic criteria and these findings are easily depicted on routine sonograms with color mapping.

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INTRODUCTION

Since the first report by Carolī et al(1), there have been several reports dealing with Carolī’s disease or congenital dilatation of the intrahepatic bile ducts(2-9). Recently new diagnostic modalities of ultrasonography(US)(10-13) and computed tomography(CT)(14-17) were applied for the diagnosis of this disease entity.

Besides congenital dilatation of intrahepatic bile ducts(bile cysts), sonograms of the liver showed portal venous radicles, which were variably described as intraluminal bulbar protrusions or bridge formation across dilated lumina(2). The pathologic features of portal radicles, partially or completely surrounded by dilated bile ducts, were also reported with US(10) and CT(14).

By looking into vascular patency and the status of blood flow within vascular radicles (or portal radicles), color Doppler Imaging(CD imaging) or conventional pulsed Doppler US may play an additional role in this disease entity. We describe newly observed color Doppler signals representing arterial wave patterns within the dilated bile ducts, which may be another diagnostic clue in diagnosing Carolī’s disease and/or Grumbach’s disease.

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CASE REPORTS

Case 1.

A 33 year-old man was admitted because of recurrent fever and right upper quadrant pain. On physical examination, the liver was palpable 3cm below the right costal margin. He had had several episodes of fever, chills, and right upper quadrant pain. Past history revealed splenomegaly from birth and he had suffered from easy fatigability. He and his younger brother (Case 2) were diagnosed as having congenital spherocytosis. Due to splenomegaly, pancytopenia, and increased peripheral ovalocytes (26%), splenectomy was done at a private hospital three years ago.

The following abnormal laboratory values were obtained: white blood cell count, 13,800/mm$^3$; mildly elevated serum alkaline phosphatase, 272IU/L. Serum total bilirubin was normal. Urinalysis was normal.

Abdominal CT and US revealed dilatations of intrahepatic bile ducts in the entire liver with a hypertrophied left lobe. Vascular radicles in the ductal lumen were also seen. Intrahepatic stones and portal vein thrombosis were also evident. Bilateral small renal cysts were associated. CD imaging demonstrated multiple color Doppler signals within dilated saccular bile ducts, appearing within or along portal radicles. Doppler spectral analysis confirmed these color signals as arterial origin by demonstrating a pulsatile wave pattern. Wedge biopsy of the liver confirmed Caroli’s disease, associated with congenital hepatic fibrosis (Fig. 1).

Case 2.

The younger brother, 27 years of age, had a palpable spleen from birth. He had a history of poliomyelitis with residual sequelae including scoliosis. He also received splenectomy under the impression of congenital spherocytosis at a private hospital three years ago. At operation, a nodular and enlarged liver was found. He was admitted this time with hematemesis. Gastric endoscopy disclosed severe esophageal varices. Laboratory tests were normal. Pathologic slide review was not available in this patient. Abdominal CT and US showed moderately severe dilatation of the intrahepatic bile ducts in both

Fig. 1. (Case one). (a) Enhanced CT shws multiple small bile cysts of dilated bile ducts mainly in the right lobe. Multiple septations are seen within cysts. (b) Transverse CD image of dilated bile ducts in the right lobe demonstrates color signals near the vascular radicle and across the lumen. (c) By light microscopy, dilated and distorted bile ducts are arranged along the fibrous septation of nodules. Within the expanded area, bile duct is irregularly dilated.
Fig. 2. (Case two). (a) Enhanced CT of case 2. Similar features to figure 1(a) are seen. (b) Oblique CD image displays a few dilated bile ducts. Color signal is depicted near the vascular radicle(arrow). (c) Pulsatile wave pattern is obtained in Doppler spectral analysis, identifying the color signal as arterial origin.

Fig. 3. (Case three). (a) Enhanced CT reveals multiple dilated bile ducts in the right lobe and the medial segment of the left lobe. Note the intraluminal linear density within the cyst of medial segment(arrow). (b) Transverse duplex CD image of the same cyst in the medial segment shows vascular radicle. Doppler spectral analysis at the tip reveals arterial pulsatile wave. (c) Longitudinal CD images of(b) demonstrate color signals, encoded red, around bulbar protrusion. (d) Arterial wave form is appeared in a Doppler spectral display in the red color signal.
lobes with small dilated saccular bile ducts in the right lobe. The left lobe of the liver was hypertrophied. Bilateral renal cysts were also found Superior mesenteric arterial portography showed multiple periportal collaterals without visualization of the main portal vein. CD imaging showed multiple color signals in dilated small bile ducts near the portal radicles, and Doppler spectral analysis revealed all these color signals to be of arterial origin(Fig. 2). Expected continuous wave patterns from the intraluminal portal venous flow within the portal radicles were not observed.

Case 3.

The eldest brother, a 35-year-old, also have had splenomegaly since birth. When he was 25 years old, he had an exploratory laparotomy because of sudden severe abdominal pain. At that time the liver was grossly cirrhotic. He did not receive a splenectomy. Thereafter he has suffered from recurrent right upper quadrant pain and fever. Abdominal CT showed dilated multiple bile ducts with intrahepatic and extrahepatic biliary lithiasis. Gallbladder stones were also found. Both kidneys showed multiple small cysts. Small dots of color Doppler signals were found in the center of vascular radicles or beside them. Spectral analysis confirmed these color signals as arterial in origin(Fig. 3). Endoscopic retrograde cholangiopancreatography confirmed the diagnosis of Caroli's disease.

DISCUSSION

Congenital bile duct dilatation is an autosomal recessive inherited disease. Pathologically, tortuous dilated, dysplastic bile ducts are found. A common additional finding is an infantile polycystic kidney. In the pathogenetic analysis of Caroli's disease by Nakanuma et al(2), they consider the malformation as a developmental abnormalities, probably a combination of overgrowth of biliary epithelium and of its supporting portal connective tissues. Disproportionate speed and extent of growth of these two tissue components in the fetus and possibly after birth might result in elongation, tortuosity, and irregular dilatation of the bile ducts, with bulbar protrusions and bridge formation of ductal walls. Jorgensen(3,4) developed the concept of the ductal plate malformation to explain the findings in autosomal recessive polycystic kidney disease and congenital hepatic fibrosis. An additional finding, visible on the macroscopic photographs, is the presence of vascular radicles including both hepatic arterial and portal venous ones, partially or completely surrounded by dilated bile ducts. These structures correspond to normal sized veins and may be hepatic arteries on sonograms, located in bulbar protrusions(6,10). But, hepatic arterial radicles are not easy to document on conventional grayscale sonograms.

To date, reported sonographic findings were dilated bile ducts with vascular radicles appearing as intraluminal bulbar protrusions, bridge formation across lumen, or portal radicles in the center of the dilated ducts, which is regarded as specific findings for the diagnosis. These findings were regarded as to support the hypothesis of arrested normal embryogenesis of intrahepatic bile ducts for the pathogenesis of this disease(10). CT can reveal findings similar to sonograms(14-17).

However, CD imaging can not only depict dilated bile ducts with intraluminal vascular radicles but also easily identify intraluminal color Doppler signals. These color signals are probably originated from small arterioles in the vascular radicles in the dilated bile ducts or in the vicinity of intraluminally located bulbar protrusions(a kind of description for vascular radicles). According to Marshal et al(10), they emphasized the presence of vascular radicles by quoting Nakanuma's previous macroscopic and microscopic photographs(2) demonstrating the portal vein and accompanying hepatic artery in the central fibrous core(so-called the portal radicles) of dilated bile ducts. In this aspect, the term of the vascular radicles seems to be more comprehensive than the portal radicles to understand and explain the embryogenesis of this disease.

The location of the arterial color signals did not match well with portal radicles: no arterial echoes could be identified in the lumen of bile ducts after turning off the color encoding. The reason for non-visualization of the hepatic arterial radicles may be explained by the fact that:relatively high velocity arteries are not imaged on gray scale sonograms because of small caliber. At present, we are lacking in proof for the explanation of the non-visualization
of the venous color signals. The lower velocity than that of arteries, inappropriate scale for velocity search, or possible portal venous thrombosis can be factors that could explain this phenomenon.

We believe these arterial color signals are very specific for the diagnosis of Caroli's disease. Although our case number were limited, we did not see such color signals in patients with hepatic simple cysts or dilated bile ducts in patients with obstructive jaundice. Some background noise signals in the lumen of hepatic cysts may mimic Caroli’s intraluminal color signals. This can be avoided with appropriate color settings prior to examination. CD imaging may play an additive or hopefully confirmative diagnostic tool in evaluating Caroli’s disease and give clues to the pathogenesis of this unique disease entity.

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<국문 요약>

Caroli’s Disease : Hepatic Arterial Color Signals in the Communicating Dilated Bile Ducts

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저자들은 선천적 간내담관 확장을 동반한 3명의 형제를 대상으로 color Doppler flow imaging(이하 CD imaging로 함)을 시행하였다. 담관 병리조직 소견상 및 임상적으로 간 섬유증과 다낭성 신질환이 세환자 모두에서 있었다. CD imaging상 담관의 색신호(color signal)를 보였고 그 외에 담관확장, 간내담석, 담액낭내의 문맥기(portal venous radicle)를 의 미하는 구상형 돌출(bulbar protrusion)을 보였다. 세명의 환자에서 세로이 발견된 색신호가 스펙트럼 분석에서 박동성 파장을 보이는 동맥인 것으로 판명되었다. 따라서 CD imaging상 동맥 색신호가 보이는 경우는 다른 간의 낭성질환에서는 볼 수 없는 이 질환의 특징적인 소견이라고 생각된다.