A CASE OF CAROLI'S DISEASE ASSOCIATED WITH CHOLANGITIS, HEPATOLITHIASIS, AND POLYCYSTIC KIDNEY DISEASE: USEFULNESS OF THE MAGNETIC RESONANCE CHOLANGIOPANCREATOGRAPHY

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Abstract: Caroli's disease is a rare congenital condition characterized by cystic dilatation of the intrahepatic bile ducts. A 74-year-old man with chronic renal failure complicated by polycystic kidney disease presented with jaundice and fever. Ultrasonography, non-contrast computed tomography, and magnetic resonance cholangiopancreatography (MRCP) were performed. Contrast medium was not administered because of the renal failure due to polycystic kidney disease. MRCP provided cholangiographic images of the biliary system. No hepatic fibrosis was observed on liver biopsied specimens. Based on the cystic dilatation of the intrahepatic bile ducts, stone formation, cholangitis, absence of hepatic cirrhosis, and association with cystic of the kidneys, a diagnosis Caroli's disease was made. (奈医誌. J. Nara Med. Ass. **50**, 46～49, 1999)

Key words: Caroli's disease, cholangitis, MRCP, polycystic kidney disease

INTRODUCTION

Caroli's disease is a rare congenital condition of the intrahepatic biliary tract which is characterized by multiple cystic dilatation of the bile ducts. It was clearly defined by Caroli et al.[1] in 1958. Various complications has been reported including bile stasis, stone formation, and biliary tract infection. Since new diagnostic techniques for evaluation of hepatic disease have been introduced, the number of reported cases of Caroli's disease has increased[2]. Recently, Pavone et al.[3] have demonstrated the utility of magnetic resonance cholangiopancreatography in the diagnosis of Caroli's disease. In this report, a patient with Caroli's disease complicated with cholangitis, hepatolithiasis, and polycystic kidney disease is discribed. The usefulness of new imaging modalities for the diagnosis of Caroli's disease is discussed.

CASE REPORT

A 74-year-old man was admitted to our hospital because of fever and pain in the right upper quadrant of the abdomen which continued for one week. He had chronic renal failure (since age 70), but no past history of biliary tract disease or hepatic dysfunction. There was no family history of hepatobiliary or polycystic kidney disease.

Physical examination revealed jaundice and dehydration. His temperature was 38.2℃, heart rate was 84 beats/min, and blood pressure was 140/85 mmHg. No lymph nodes were palpable.

いた症例は今回がはじめての報告と思われる．手術は全例，片側卵巣摘出術が行われていた．可能な限り腫瘍核出術を試みるべきであることは言うまでもないが，西田ら[5]は，卵巣腫瘍茎捻転の発症後36時間以上経過した例では腫瘍核出が可能なものはなかったと報告しており，また組織学的診断が可能なのは24時間以内であると述べている．先述したように，低年齢児では茎捻転の診断に苦慮する場合が多く，卵巣摘出術も，やむをえないと思われた．

類皮嚢胞腫の診断は比較的容易で，小児においては，とくに腹部超音波検査が有用である．近年の普及率の増加と，画像解析度の向上により，従来問題とされていた急性虫垂炎との鑑別は容易となり，嚢胞内の骨，歯牙による高輝度エコーや，毛髪による“hairball sign”が鮮明に描出されるようになったために[6]，最近では術前に確定診断される症例が多く見られる．また，本症例では尿閉を伴っていたために膀胱が緊満し，腫瘍が膀胱を圧迫する像が明確に描出され，この経時変化を観察することで，腫瘍と尿閉の因果関係を類推する一助となった．

卵巣腫瘍による尿閉の機序として，腫瘍のダグラス窩への嵌頓による膀胱頚部および尿道の機械的圧迫[7]や，腫瘍が膀胱底部を挙上させることによる引き上げ現象で尿道が延長，偏位すること[8]などが考えられている．本症例においては，右卵巣が左膀胱底部後方に存在しており，また茎捻転により膀胱と強い炎症性癒着を呈し，左膀胱底部は前上方へ偏位していた．つまり，引き上げ現象による尿閉の可能性が高いと考えられた．さらに，右卵管は両側尿管をまたぐ形で存在しており，これが尿管開口部を圧迫していた可能性も否定できない．また，導尿後すみやかに腹痛が消失したことから，膀胱の緊満によって捻転が助長され，腫瘤が膀胱に与える影響が相対的に増大することにより尿閉の解除が遅れるという，一種の悪循環に陥った可能性があり，本症例において，乏尿の原因を脱水によるものとして安易に輸液を続けたことは反省すべきである．以上のことから，小児尿閉の原因のひとつに腹腔内腫瘤があることを念頭におき，初期診断時に超音波検査を中心とした画像検査を積極的に行うべきであると思われた．

ま　と　め

幼児期に発症した卵巣類皮嚢胞腫茎捻転に尿閉を伴った稀な症例を経験し，小児期の尿閉に腫瘤性病変が原因となりうることを述べ，若干の文献的考察を加えて報告した．

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Cardiac ausculation revealed no extra heart sounds or murmurs. The liver was revealed 1/2 of a finger breadth below the costal margin at the mid-clavicular line. There was no evidence of ascites, splenomegaly, or venous dilatation of the abdominal wall.

Laboratory examination on admission revealed erythrocyte sedimentation rate of 120 mm in 1 hour, white blood cell count (WBC) of 23,250/mm^3 with 89 % neutrophils, and C-reactive protein concentration of 14.6 mg/dL. The total bililubin (TB), glutamic oxaloacetic transaminase (GOT), glutamic pyruvic transaminase (GPT), and serum creatinine (Cr) concentrations were elevated at 7.0 mg/dL, 69 IU/L, 67 IU/L and 6.5 mg/dL, respectively.

Abdominal ultrasonography (US) demonstrated cystic dilatation of the intrahepatic bile ducts, intrahepatic lithiasis, and bilateral multiple renal cysts with septae. Non-contrast computed tomography (CT) of the liver revealed multiple cysts with an unclear relationship to the intrahepatic biliary tract (Fig. 1), and polycystic kidneys with calcification of the septal wall (Fig. 2). Intravenous administration of contrast media was not performed because of the history of chronic renal failure complicated by polycystic kidney disease. Magnetic resonance cholangiography (MRCP) showed multiple intrahepatic cystic lesions with communications to the intrahepatic bile ducts (Fig. 3.). The cholangitis resolved with antibiotic treatment 2 weeks later. The WBC, TB, GOT, GPT, and Cr concentrations improved (8,710/mm^3, 1.7 mg /dL, 39 IU/L, 25 IU/L and 3.4 mg/dL, respectively).

Percutaneous transhepatic cholangiography (PTC) revealed multiple liver cysts in communication with the intracepatic bile ducts. Cytologic examination of the bile showed no malignant cells. The bile cultures were sterile. Pathologic examination of a liver biopsy specimen revealed no congenital hepatic fibrosis. The patient was diagnosed with a pure type of Caroli' s disease.

DISCUSSION

Caroli's disease is classified into two types on the basis of the existence of congenital hepatic fibrosis. The first is the pure type of Caroli's disease without congenital hepatic fibrosis, which is considered to be rare. The second type is associated with congenital hepatic fibrosis.

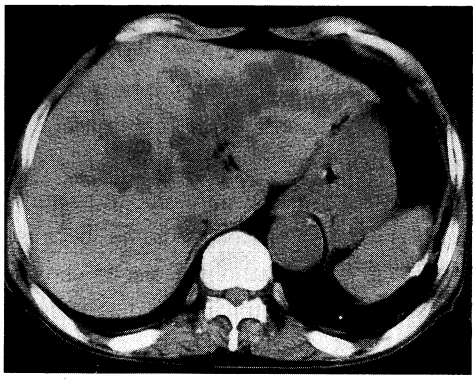

Fig. 1. Unenhanced computed tomography of the liver demonstrated cystic dilatation of intrahepatic biliary tracts.

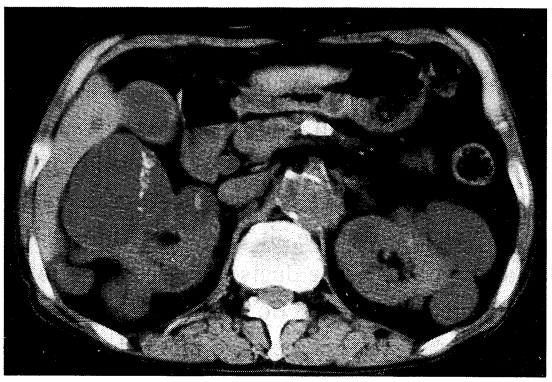

Fig. 2. Unenhanced computed tomography of the kidney demonstrated multiple cysts with calcification of septal wall.

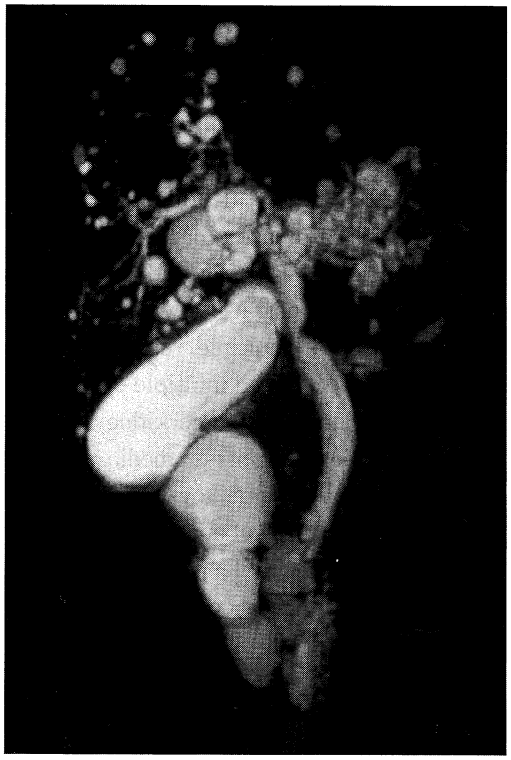

Fig. 3. Magnetic resonance cholangiography showed multiple intrahepatic cystic dilatation which communicated with intrahepatic bile ducts.

According to recent reports, both entities are closely related, representing different points along a spectrum of congenital deficits[2]. The symptomatology of the first type is due to ductal stone formation caused by bile stasis within the cystic lesions. Such patients often have relapsing cholangitis, which can lead to liver abscess and sepsis. In the second type, the clinical manifestations are mainly due to portal hypertention. Esophageal varices are often present. Polycystic kidney disease is known to be associated with Caroli's disease.

Hepatolithiasis and intrahepatic cholangiocarcinoma are the two most serious complications of Caroli's disease. Tsunoda[4] has reported hepatolithiasis in 30.6 % of patients with Caroli's disease in Japan. Long-standing bile stasis and repeated episodes of cholangitis appear to play an important role in stone formation. Tsunoda[4] has also reported cholangiocarcinoma in 8 % of patients with Caroli's disease in Japan. Irritation from stones, long-standing bile stasis, and repeated episodes of cholangitis seem to relate to the development of carcinoma. In patients with Caroli's disease, it is difficult to make a diagnosis of cholangiocarcinoma because dilatation of the bile ducts may be caused by the original disease, hepatholithiasis, or cholangiocarcinoma. Magnetic resonance imaging (MRI) appears to be useful in the diagnosis of Caroli's disease accompanied by cholangiocarcinoma[5].

Most diagnostic procedures for evaluation of biliary tract, such as PTC and endoscopic retrograde cholangiography (ERC), are invasive. PTC and ERC are the most accurate methods for the demonstration of biliary anatomy and the communications between the cystic dilata-

tions and the bile ducts. However, they are invasive, and require radiation exposure and systemic contrast medium administration. Moreover, they may lead to cholangitis.

With the recent introduction of new imaging techniques, the non-invasive diagnosis of Caroli's disease is now possible. In the present case, Caroli's disease was diagnosed using US, CT, and MRCP. US showed portal radicles partially or completely surrounded by dilated bile ducts, as reported by Marchals et al.[6]. The CT findings include strongly enhanced tiny dots in the dilated intrahepatic bile ducts, as reported by Choi et al.[7]. This has been called the central dot sign. However, the administration of contrast medium is contraindicated for the patients with renal dysfunction. This is important because the incidence of renal involvement in Caroli's disease is high. It is possible to diagnose Caroli's disease using MRCP without intravenous administration of contrast medium.

MRCP is a sensitive, noninvasive method for the diagnosis of Caroli's disease which is comparable to ERC and PTC and requires neither biliary intervention nor contrast mudium administration. Futhermore, MRCP can show the cystic lesions along the bile ducts and their communications with the biliary tract. MRCP should be considered as a most valuable method for the diagnosis of Caroli's disease.

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内視鏡的に急性期および治癒期を観察し得た腸管出血性大腸菌O157：H7による出血性腸炎の1例

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A CASE OF HEMORRHAGIC COLITIS DUE TO ENTEROHEMORRHAGIC ESCHERICHIA COLI O 157 OBSERVED ENDOSCOPICALLY AT THE ACUTE AND HEALED STAGE

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Abstract: A 49-year-old woman was admitted to the hospital because of abdominal pain and bloody diarhea. Emergent colonoscopy was performed due to continuous bloody diarrhea on the following day after admission. Although edema, erosion, ulceration and hemorrhage were observed from the sigmoid colon through the cecum, inflammatory changes were more remarkable in the ascending colon with narrowing of the colon due to severe edema. Apparent inflammatory changes were not seen in the terminal ileum. Subsequently the stool culture was positive for Escherichia coli O 157, resulting in the diagnosis of hemorrhagic colitis due to Escherichia coli O 157 infection. Biopsy specimens revealed fibrin thrombosis in many vessels, closely similar to observations in ischemic colitis. It is therefore indicated that differential diagnosis from ischemic colitis is important. Her symptoms and inflammatory parameters improved with antibiotics without causing hemolytic-uremic syndromes, and no inflammatory findings were observed by colonoscopy performed on day 28 after the onset.
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