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

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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. in 1958. Various complications have 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. Recently, Pavone et al. 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 described. 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°C, heart rate was 84 beats/min, and blood pressure was 140/85 mmHg. No lymph nodes were palpable.
尿閉を伴った幼児卵巢類皮膚囊腫摘出の1手術例

いった症例は今回がはじめての報告と思われる。手術は全例、片側卵巢摘出術が行われていた。可能な限り腫瘍摘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9は、卵巢腫瘍摘出の発症後36時間以上経過した例では腫瘍摘出が可能なものはなかったと報告しており、また組織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、低年齢児では腫瘍の診断に苦慮する場合が多く、卵巢摘出術も、むやみをえないと思われた。

類皮膚囊胞の診断は比較的容易で、小児においては、特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、囊胞内の骨、骨髄による高輝度イメージ、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9、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張し、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顕微する一助となった。

卵巢腫瘍による尿閉の機序として、腫瘍のデフラックス管への嵌頓による膀胱底部および尿道の機械的圧迫や、腫瘍が膀胱底部を拡張させることによる引き上げ現象で尿道が延長、偏位することなどが考えられている。本症例においては、右卵巣の左膀胱底部後方に存在しており、また茎持性により膀胱が強い炎症性腫脹を呈し、左膀胱底部は前方へ偏位していた。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、右卵巢は両側非腫大をきたし呈在しており、これが尿道間口部を圧迫していた可能性も否定できない。また、尿管後すみやかに腹痛が消失したことから、膀胱の緊張によって直腸が圧迫され、腹痛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消が遅れたという、一種の循環障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容易に説明を続けたことは反省すべきである。以上のことから、小児尿閉の原因のうち、特に腫瘍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。

まとめる

幼児期に発症した卵巢類皮膚囊腫摘出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。

文献

1) 有岡秀樹、小林健二、内藤広行、石川順一：CA 19-9、CA 125の異常高値を認めた小児卵巢成熟奇形腫の1例。小児科診療。56：477-480、1993。
4) 上芳敏子、中原優人、島田信宏：少女に多い卵巢瘜器、産科と婦人科。55：768-772、1988。
5) 西田敬、杉山徹、荒木篤宣、久保紀夫、三田村民夫、井出信、西村治夫、薬師寺道明：卵巢腫瘍の茎持出に関する臨床的および組織学的検討。産婦人科の実際。30：1463-1468、1981。
6) 片藤保、山崎俊彦：類皮膚囊胞の画像解析。産婦人科。55：805-807、1988。
7) 田畑雅章、松浦敏幸、橋本昌樹、平田輝夫：尿閉を主訴とした巨大卵巢囊腫の1例。臨床。36：1077-1079、1982。
8) 松本美代、渡辺雅幸、上村良成、大川順正：尿閉をきたした卵巢類皮膚囊胞の女児例。泌尿器科。39：85-87、1993。
Cardiac auscultation 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/mm³ with 89% neutrophils, and C-reactive protein concentration of 14.6 mg/dL. The total bilirubin (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/mm³, 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 intraparenchymal 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**

Carolii’s disease is classified into two types on the basis of the existence of congenital hepatic fibrosis. The first is the pure type of Carolii’s disease without congenital hepatic fibrosis, which is considered to be rare. The second type is associated with congenital hepatic fibrosis.
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. 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 hypertension. 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 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 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, hepatolithiasis, or cholangiocarcinoma. Magnetic resonance imaging (MRI) appears to be useful in the diagnosis of Caroli’s disease accompanied by cholangiocarcinoma.

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. The CT findings include strongly enhanced tiny dots in the dilated intrahepatic bile ducts, as reported by Choi et al. 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 medium administration. Furthermore, 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.

REFERENCES


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 diarrhea. 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 O157, resulting in the diagnosis of hemorrhagic colitis due to Escherichia coli O157 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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