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Case Report

Diagnosis of Caroli Disease with Gadoxetic Acid-Enhanced Magnetic Resonance Imaging

Ramazan Orkun Önder, Serdar Aslan, Tumay Bekci

Department of Radiology, Giresun University, Faculty of Medicine, Giresun, Turkey

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Corresponding author: Ramazan Orkun Önder, e-mail: orkunonder535@gmail.com

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Abstract

Caroli disease (CD) is a condition in which there are multiple cystic dilations of the bile ducts within the liver that are present from birth. In this case report, we present the imaging findings of a 33-year-old woman who was diagnosed noninvasively after gadoxetic acid (GA)-enhanced magnetic resonance imaging, which has been used in recent years, which is very rare in the literature. A woman, aged 33, was admitted to the hospital after experiencing recurring episodes of pain in the upper right area of her abdomen for a long period of time. Laboratory tests showed elevated liver enzyme levels and hyperbilirubinemia. Abdominal ultrasonography revealed cystic lesions in the liver and mild enlargement of the common bile duct. It was observed that cystic lesions in the liver filled with GA in the hepatobiliary phase and were associated with the intrahepatic bile ducts. Thus, the patient was diagnosed with CD. In the hepatobiliary phase, GA-enhanced magnetic resonance imaging allows the detection of other conditions in which a central dot sign (CDS) occurs (e.g., peribiliary cysts, periportal lymphedema, and jaundice due to biliary obstruction) and noninvasive diagnosis of CD in the absence of a CDS.

Keywords: Caroli disease, central dot sign, gadoxetic acid, hepatobiliary phase, MRI

INTRODUCTION

Caroli disease (CD) is a condition in which there are multiple cystic dilations of the bile ducts within the liver that are present from birth. Caroli disease can manifest as either diffuse or limited, and it can involve the formation of sac-like structures that connect with the biliary tree and form cystic formations. They are also classified as a type V choledochal cyst, according to the Todani classification.[1] Two categories of magnetic resonance imaging (MRI) contrast agents possess hepatocyte-selective characteristics: gadolinium-based and manganese-based agents. While gadolinium-based agents exhibit both hepatocyte and perfusion imaging properties, making them bimodal contrast agents, gadoxetic acid (GA) is a gadolinium-based hepatocyte-specific contrast agent.[2] In this case report, we present the imaging findings of a 33-year-old woman who was diagnosed with CD noninvasively after GA-enhanced MRI, which has been used in recent years, which is very rare in the literature.

CASE PRESENTATION

A woman, aged 33, was admitted to the hospital due to experiencing recurring episodes of pain in the upper right area of her abdomen for a long period of time. Her medical history was unremarkable. Upon physical examination, mild tenderness in the right upper quadrant was revealed. Laboratory results demonstrated elevated aspartate aminotransferase of 310 U/L (normal < 40 U/L), alanine aminotransferase of 207 units/L (normal < 41 U/L), direct bilirubin of 3.11 mg/dL (normal < 0.2 mg/dL), and total bilirubin of 4.67 mg/dL (normal < 1.25 mg/dL). Abdominal ultrasonography (US) revealed cystic lesions in the liver and mild enlargement of the common bile duct. The patient then underwent GA-enhanced MRI. Cystic lesions with high signal on T2-weighted MRI and low signal on T1-weighted MRI were observed in liver segments 4, 7, and 8 (the largest was 3 × 2 cm in segment 7) (Figure 1). Contrast enhancement was not observed in cystic lesions in venous-phase contrast MRI (Figure 2). Magnetic resonance imaging in the hepatobiliary phase of the liver 20 minutes after GA injection showed cystic lesions filled with GA (Figure 3). Thus, the patient was noninvasively diagnosed with CD and found to have an episode of cholangitis. The patient had no findings of hepatomegaly, splenomegaly, or portal hypertension. The patient was started on ursodeoxycholic acid 250 mg daily as supportive therapy, and conservative treatment for cholangitis and follow-up was recommended.

DISCUSSION

Caroli disease may be focal or diffuse and it is important to distinguish diffuse CD from autosomal dominant polycystic liver disease or peribiliary cysts in a cirrhotic liver. In terms of MR imaging findings, CD is characterized by multiple cysts within the liver, closely associated with the biliary system, often with a central dot sign (CDS).[3] While MRI may not show the communication between cystic lesions and draining bile ducts, this challenge can be overcome by utilizing GA-enhanced MRI. This advanced technique allows for the visualization of communications between cystic lesions and draining bile ducts, enabling the differentiation of CD from other conditions such as autosomal dominant polycystic liver disease or peribiliary cysts in a cirrhotic liver.[4,5] Various imaging techniques consistently reveal similar findings, with ultrasound being

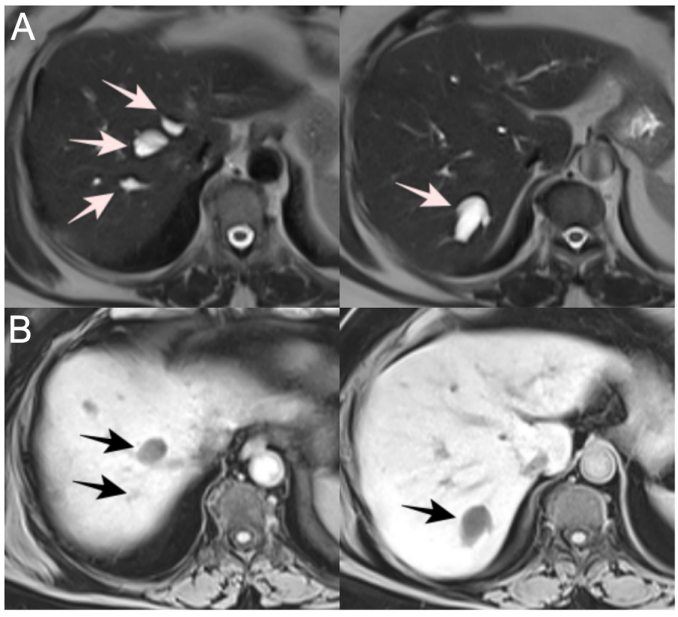

Figure 1. (A) High-signal cystic lesions (open arrows) in the liver observed on T2-weighted MRI. (B) Low-signal cystic lesions (filled arrows) in the liver observed on T1-weighted MRI. MRI, magnetic resonance imaging.

the initial test of choice, showing the presence of saccular dilatations in the intrahepatic ducts.[6,7] The CDS, which was not detected in our case, is a clue for the diagnosis of CD. The CDS represents the telltale appearance of intrahepatic portal vein branches surrounded by severely dilated bile ducts, which can be visualized by either computed tomography, MRI, or US. However, it may not be seen in every patient, and while initially considered specific for this disease, later results have shown that the CDS sporadically also occurs in other conditions (e.g., peribiliary cysts, periportal lymphedema, and jaundice due to biliary obstruction).[8-10] Cholangiography by endoscopic retrograde cholangiopancreatography (ERCP) shows nonocclusive cystic dilatations communicating with the biliary tree. However, invasive methods (e.g., ERCP and percutaneous transhepatic cholangiography) can have complications such as bleeding, infection, and pancreatitis. After venous injection of GA, up to 50% is taken up exclusively by organic anion transporter polypeptides in the sinusoidal membrane of normal hepatocytes and then excreted into the biliary ducts.[11] The increase in liver parenchyma is determined by the balance of this

MAIN POINTS

- Central dot sign may be a clue for Caroli disease (CD). However, it is not specific for this disease. Because it may also occur sporadically in conditions such as peribiliary cysts, periportal lymphedema, and jaundice due to biliary obstruction.
- Ultrasonography and computed tomography may not clearly distinguish dilatation of intrahepatic bile ducts from cystic lesions. However, on magnetic resonance imaging (MRI) with gadoxetic acid (GA) in the hepatobiliary phase, dilatation of the intrahepatic bile ducts can be easily detected as cystic lesions filled with the contrast medium.
- Gadoxetic acid-enhanced MRI in the hepatobiliary phase enables noninvasive diagnosis of CD, protecting patients from the complications of invasive procedures.

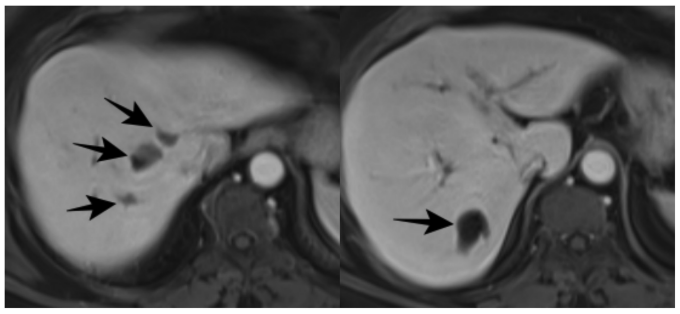

Figure 2. Nonenhancing cystic lesions (filled arrows) observed on venous-phase contrast-enhanced MRI. MRI, magnetic resonance imaging.

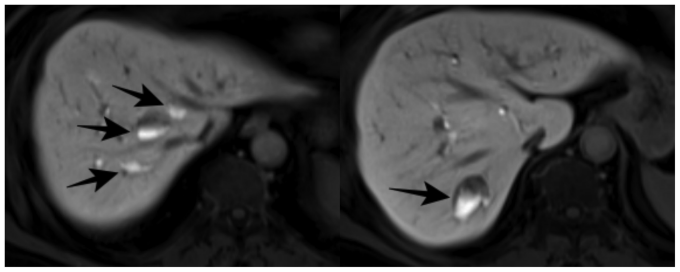

Figure 3. In the hepatobiliary phase, cystic lesions filled with GA (filled arrows) and associated with intrahepatic bile ducts. GA, gadoxetic acid.

transport mechanism and an optimal enhancement effect is observed for a period of 10-40 minutes after administration.[12] In recent years, it has been suggested that the use of contrast-enhanced MRI with GA allows a safe, noninvasive diagnosis of CD.[10,13-14] In our case, we diagnosed CD after GA-enhanced MRI without the use of invasive methods. In conclusion, in the hepatobiliary phase, GA-enhanced MRI allows the detection of other conditions in which CDS occurs (e.g., peribiliary cysts, periportal lymphedema, and jaundice due to biliary obstruction) and enables the noninvasive diagnosis of CD even in the absence of a CDS. This approach also protects patients from the complications of invasive procedures (e.g., ERCP and percutaneous transhepatic cholangiography) such as bleeding, infection, and pancreatitis.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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REFERENCES

1. Singham J, Yoshida EM, Scudamore CH. Choledochal cysts: part 2 of 3: Diagnosis. *Can J Surg*. 2009;52(6):506-511.
2. Ba-Ssalamah A, Uffmann M, Saini S, Bastati N, Herold C, Schima W. Clinical value of MRI liver-specific contrast agents: a tailored examination for a confident non-invasive diagnosis of focal liver lesions. *Eur Radiol*. 2009;19(2):342-357. [CrossRef]

3. Levy AD, Rohrmann CA Jr, Murakata LA, Lonergan GJ. Caroli's disease: radiologic spectrum withpathologic correlation. *AJR Am J Roentgenol*. 2002;179(4):1053-1057. [CrossRef]
4. Park MS, Kim BC, Kim T, Kim MJ, Kim KW. Double common bile duct: curved-planar reformatted computed tomography (CT) and gadobenate dimeglumine-enhanced MR cholangiography. *J Magn Reson Imaging*. 2008;27(1):209-211. [CrossRef]
5. Park MS, Yu JS, Lee JH, Kim KW. Value of manganese-enhanced T1- and T2-weighted MR cholangiography for differentiating cystic parenchymal lesions from cystic abnormalities which communicate with bile ducts. *Yonsei Med J*. 2007;48(6):1072-1074. [CrossRef]
6. Yamaguchi T, Cristaudi A, Kokudo T, Uldry E, Demartines N, Halkic N. Surgical treatment for monolobular Caroli's disease – report of a 30-year single center case series. *BioSci Trends*. 2018;12(4):426-431. [CrossRef]
7. Soares KC, Goldstein SD, Ghaseb MA, Kamel I, Hackam DJ, Pawlik TM. Pediatric choledochal cysts: diagnosis and current management. *Pediatr Surg Int*. 2017;33(6):637-650. [CrossRef]
8. Khalefa AA, Alrasheed M, Saeedan MB. Central dot sign. *Abdom Radiol*. 2016;41(11):2289-2290. [CrossRef]
9. Santiago I, Loureiro R, Curvo-Semedo L, et al. Congenital cystic lesions of the biliary tree. *AJR Am J Roentgenol*. 2012;198(4):825-835. [CrossRef]
10. Salvadori PS, Torres US, D'Ippolito G. Contrast-enhanced magnetic resonance cholangiography with gadoxetic-acid-disodium for the detection of biliary-cyst communication in Caroli disease. *Gastroenterol Hepatol*. 2016;39(10):669-670. [CrossRef]
11. Van Beers BE, Pastor CM, Hussain HK. Primovist, Eovist: what to expect? *J Hepatol*. 2012;57(2):421-429. [CrossRef]
12. Brismar TB, Dahlstrom N, Edsborg N, Persson A, Smedby O, Albiin N. Liver vessel enhancement by Gd-BOPTA and Gd-EOB-DTPA: a comparison in healthy volunteers. *Acta Radiol*. 2009;50(7):709-715. [CrossRef]
13. Choi BI, Yeon KM, Kim SH, Han MC. Caroli disease: central dot sign in CT. *Radiology*. 1990;174(1):161-163. [CrossRef]
14. Ahmadi T, Itai Y, Minami M. Central dot sign in entities other than Caroli disease. *Radiat Med*. 1997;15(6):381-384.