

A Case of Right Hepatic Artery Syndrome Diagnosed by Using SpyGlassDS™ System

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ABSTRACT

We report the case of a 68-year-old woman who had abdominal pain and slightly elevated biliary enzymes. Magnetic resonance cholangiopancreatography detected biliary duct stenosis, while contrast-enhanced magnetic resonance imaging showed that the right hepatic artery transversed the extrahepatic bile duct at the level of bifurcation of the bile duct. We performed endoscopic retrograde cholangiopancreatography and peroral cholangioscopy with the SpyGlass DS™ system. Then, mild extrinsic pulsatile compression of the bile duct was observed at stricture level with an intact bile duct epithelium. Therefore, she was diagnosed with right hepatic artery syndrome and underwent cholecystectomy. Six months later, her biliary enzyme level decreased, and the recurrence of pain gradually decreased.

Key words diagnostic imaging; extrahepatic cholestasis

Anatomical characteristics of the hepatic portal are the concentration of luminal structures such as the bile duct, portal vein, and hepatic artery. In other organs (e.g. esophagus, duodenum, ureter), vessels compress the adjacent organ and disturb the flow of their contents. However, it has rarely been reported that the bile ducts are compressed by the right hepatic artery (RHA).¹⁻⁷ In 1984, Tsuchiya reported the first case of jaundice and hepatolithiasis due to RHA compression on the common hepatic duct.¹ Since then, this condition has been called RHA syndrome.

PATIENT REPORT

The patient was a 68-year-old woman who visited a nearby hospital with a chief complaint of right upper quadrant pain. She was diagnosed with biliary dyskinesia, and the administration of flopropione improved symptoms temporarily. Three years later, her body weight was reduced by 3 kg after an aortic valve replacement surgery for aortic valve stenosis, and the same symptoms presented. A laboratory examination revealed the following: alkaline phosphatase, 787 IU/L; gamma-glutamyl transpeptidase, 93 IU/L; aspartate aminotransferase, 23 U/L; alanine aminotransferase, 18 U/L; CEA, 3.3 ng/mL; CA19-9, 35.1 U/mL; Span-1, 24 U/mL; and DUPAN-2, < 25 U/mL. Her anti-mitochondrial antibody test was negative. Thus, she was referred to our hospital for further examination and treatment.

The preferred diagnostic procedure included the use of a contrast agent, however, a past allergic reaction prevented us from using it. An abdominal plain computed tomography (CT) was conducted instead, which did not show any abnormal findings. However, magnetic resonance cholangiopancreatography (MRCP) revealed a stricture in both right and left hepatic ducts (Fig. 1) and wall thickening of the gallbladder. Endoscopic ultrasonography (EUS) could not detect wall thickening or tumors in the strictures of the bile duct, but was able to visualize aberrant RHA and gallbladder wall thickening due to Rokitansky-Aschoff sinuses (RAS). Contrast-enhanced magnetic resonance imaging (MRI) showed RHA transversed the extra hepatic bile duct at the level of bifurcation of the bile duct (Fig. 2). At this point, we suspected RHA syndrome and planned further examination. Endoscopic retrograde cholangiopancreatography (ERCP) showed the pressure excursion of the bile duct, and intraductal ultrasonography (IDUS) confirmed that the blood vessel was adjacent to the compressed site (Fig. 3). Then we performed a peroral cholangioscopy (POCS) with the SpyGlass DS™ system (Boston Scientific Corp, Natick, MA) and observed a mild extrinsic pulsatile compression of the bile ducts at the stricture level (Fig. 4). There was no epithelial neoplasm at the site of pulsatile compression. Owing to stenosis of

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Abbreviations: CT, computed tomography; ERCP, endoscopic retrograde cholangiopancreatography; EUS, endoscopic ultrasonography; IDUS, intraductal ultrasonography; MRCP, magnetic resonance cholangiopancreatography; MRI, magnetic resonance imaging; POCS, peroral cholangioscopy; RAS, Rokitansky-Aschoff sinuses; RHA, right hepatic artery

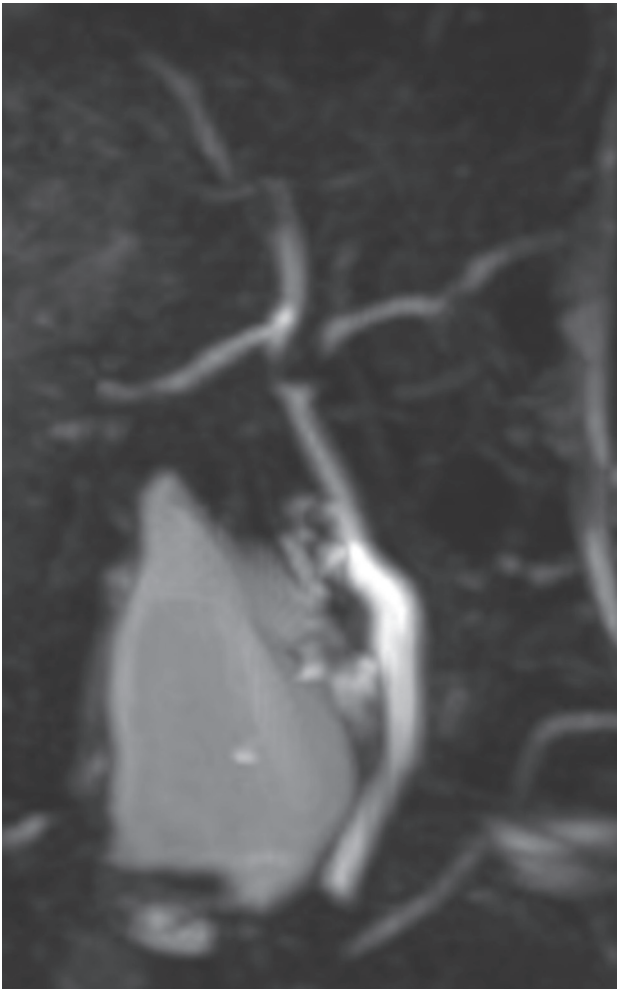


Fig. 1. MRCP; Both right and left hepatic ducts are constricted.

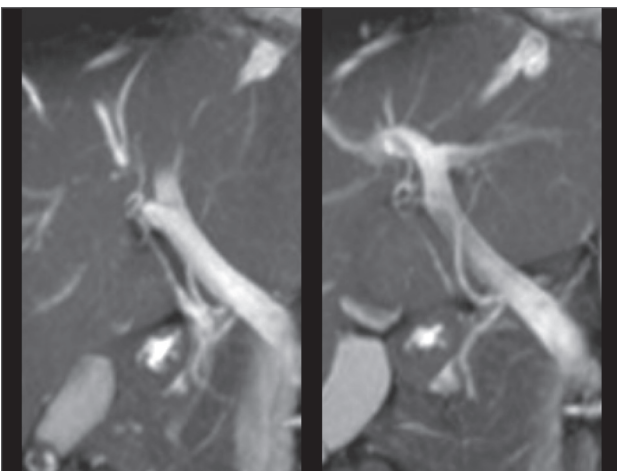


Fig. 2. Contrast-enhanced MRI showing RHA transversed the extra hepatic bile duct at the level of bifurcation of the bile duct.

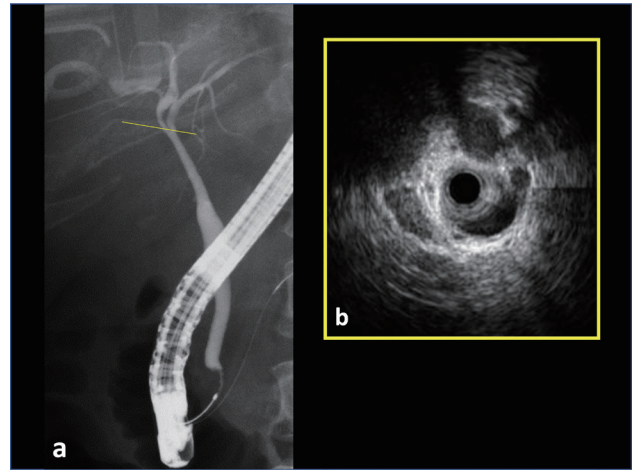


Fig. 3. a: ERCP showing the pressure excursion of the bile duct. b: IDUS showing that the blood vessel was adjacent to the same site.

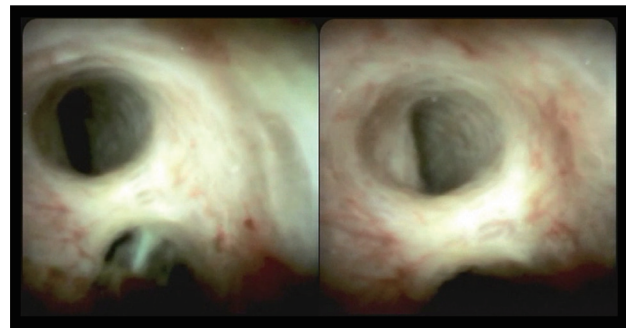


Fig. 4. POCS using SpyGlass DS™ system: Mild extrinsic pulsatile compression of the bile ducts at the stricture level with an intact bile duct epithelium.

the bile duct, we considered malignant tumor in differential diagnosis during the initial examination; however, POCS revealed pulsatility stenosis, and we confirmed that the diagnosis of RHA syndrome. However, there were abdominal symptoms and hepatic dysfunction, and we contemplated that follow-up observation was not valid. Meanwhile, adenomyomatosis of the gallbladder was observed, and the patient was scheduled to undergo cholecystectomy; thus, we detected follow-up RHA syndrome after cholecystectomy. The biliary tree enzyme level decreased (alkaline phosphatase, 317 IU/L; gamma-glutamyl transpeptidase, 65 IU/L), and the frequency of pain gradually decreased 6 months after cholecystectomy.

Table 1. Details of 8 reported cases of RHA syndrome

	Author	Age	Sex	Symptoms	Gall stone	Pprocedures	Observation Period	Clinical Course
1984	Tsuchiya et al.	59	F	Fever, Abdominal pain	Bile duct	Hepaticojejunostomy	–	Uneventful
1984	Tsuchiya et al.	57	F	Abdominal pain	Bile duct	Left partial hepatectomy + hepatico-jejunostomy	53 days	Dead
1994	Chung et al.	39	F	Fever, Jaudice	–	Cholecystectomy	6 months	Abdominal pain
2005	Izushi et al.	51	M	Abdominal pain	–	Cholecystectomy	3 years	Uneventful
2005	Miyashita et al.	55	M	Fever, Weight loss	Gallbladder	Hepaticojejunostomy	1 year	Uneventful
2012	Estiaghpour et al.	54	M	Abdominal pain, Jaudice	Bile duct	Endoscopic lithotripsy	–	Uneventful
2012	Tas et al.	74	F	Abdominal pain	–	–	–	Uneventful
2014	Mendes et al.	52	F	Abdominal pain, Jaudice	Bile duct	Hepaticojejunostomy	2 years	Uneventful

F, female; M, male.

DISCUSSION

Normally, the hepatic artery courses between the extrahepatic bile duct and the portal vein. Aberrant and replaced RHAs are well-known variants. Koops reported that normal anatomy of the hepatic artery was found in 79.1% patients and the anomalous arterial patterns in the remaining patients.⁸ Tsuchiya reported that 11 (14%) among 79 patients who underwent angiographies or operations due to intrahepatic stone showed the RHA anteriorly crossing to the bile duct.¹ He defined RHA syndrome as a variety of hepatobiliary symptoms caused by compression of the bile duct by the RHA. We searched PubMed with the keyword “right hepatic artery syndrome” and selected a report that was published in an English journal, met the definition of RHA syndrome, and could identify age, gender, and symptoms of the case. As a result, eight case reports were found in seven literature. (Table 1). Five cases were associated with gallstones, of which 3 cases were proximal to the compression site, 1 case was distal to the compression site, and 1 case was in the gallbladder. In RHA syndrome, the formation of gallstone is attributed to the bile stasis caused by compression of the common bile duct,¹ and later by the bacterial proliferation.⁹ In the other 3 cases, although gallstone was not present, patients suffered from some symptoms (2 cases, abdominal pain; 1 case, fever). It is not known exactly why the congenital anomalies cause symptoms only during adulthood in RHA syndrome. Chung et al. considered that the cystic

artery, which commonly branches from RHA, had tension and played some role, because the RHA became longer after cholecystectomy.² Some studies described that post-cholecystectomy patients have significantly wider common bile duct diameters.^{10, 11} Tanaka et al. investigated the coordination of the gallbladder and sphincter of Oddi and the effect of cholecystectomy on biliary pressure physiology in 7 patients using an indwelling micro transducer catheter placed in the bile duct.¹² They considered that the spasm of the sphincter of Oddi readily leads to a pressure rise if the gallbladder is absent, which may partly explain the change in common bile duct diameter and internal pressure after cholecystectomy. In our case, the expansion of the common bile duct diameter was unrecognized after cholecystectomy, but the symptoms and examination data improved.

There was no dilation of the upstream bile duct, but pulsatile stenosis that caused cholestasis was present. Making differential diagnosis from cholangiocarcinoma was difficult with static images such as CT and MR images; however, diagnosis was possible by observing dynamically. Both IDUS and POCS are capable of being observed dynamically. While IDUS dynamically observes a cross-sectional image of the bile duct, POCS has the advantage of being able to simultaneously be observed a specific area of the bile duct on the same screen. In addition, since IDUS sometimes cannot diagnose the intraepithelial extension of the bile duct,¹³

direct observation by POCS was performed to confirm the diagnosis.

In conclusion, arterial compression should be considered in the differential diagnosis of extrahepatic biliary obstruction. POCS with SpyGlass DS™ system is useful for the diagnosis of RHA syndrome.

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The authors declare no conflict of interest.

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