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Value of carbon dioxide wedged venography and transvenous liver biopsy in the definitive diagnosis of Abernethy malformation

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Abstract

We report a 25-year-old man who presented with congenital absence of the portal vein, or Abernethy malformation, a rare congenital disorder in which the mesenteric and splenic venous drainages bypass the liver and directly drain into the inferior vena cava through an extrahepatic portosystemic shunt. Magnetic resonance imaging, which showed multiple nodular lesions in both liver lobes that were associated with an absence of intrahepatic portal venous branches, strongly suggested the diagnosis of the Abernethy malformation. Carbon dioxide wedged venography and transvenous liver biopsy, which were performed in the same session by a right jugular approach, confirmed these findings. This technique can be considered a valuable alternative diagnostic tool to catheter arteriography and percutaneous transhepatic liver biopsy.

Key words: Congenital—Portosystemic shunt—Angiography—Biopsy—Carbon dioxide

Congenital absence of the portal vein (CAPV), or Abernethy malformation, is a very rare congenital anomaly consisting of direct drainage of the visceral venous blood through an aberrant shunt into the inferior vena cava, thereby bypassing the liver parenchyma. In most published cases, the diagnosis is suggested by noninvasive cross-sectional imaging [1, 2] and subsequently confirmed by catheter angiography [3, 4]. In a minority of cases, additional transhepatic liver biopsy [5], showing an absence of portal venules within the portal triads, confirmed the diagnosis. We report on the potentially additional value of carbon dioxide (CO_2) wedged venography and transvenous liver biopsy in the definitive diagnosis of Abernethy malformation.

Case report

A 25-year-old male patient was referred because of mild intermittent jaundice and pruritus. He did not use any medication or alcohol. Abdominal pain was absent. Clinical examination showed no significant abnormalities. Laboratory analysis revealed abnormal liver test results, with an aspartate aminotransferase level of 161 IU/L (normal < 38 IU/L), an alanine aminotransferase level of 85 IU/L (normal < 41 IU/L), and a gamma-glutamyltransferase (γ-GT) level of 242 IU/L (N < 53 IU/L); blood ammonia level was 73 μmol/L (normal < 32 μmol/L). Tests for viral hepatitis, autoimmune hepatitis, and hemochromatosis were negative.

Magnetic resonance imaging (MRI) of the liver showed multiple small nodular lesions in both liver lobes that were hyperintense on T2-weighted images (Fig. 1). Some were hypointense on T1-weighted images, and some were slightly hyperintense. The inferior vena cava was enlarged but the hepatic veins were normal. The extrahepatic portal vein also appeared enlarged, but the intrahepatic portal vein branches could not be depicted. The splenic and mesenteric veins had normal anatomic characteristics. The central intrahepatic and extrahepatic bile ducts were normal and there was no evidence of liver cirrhosis. Agenesis of the

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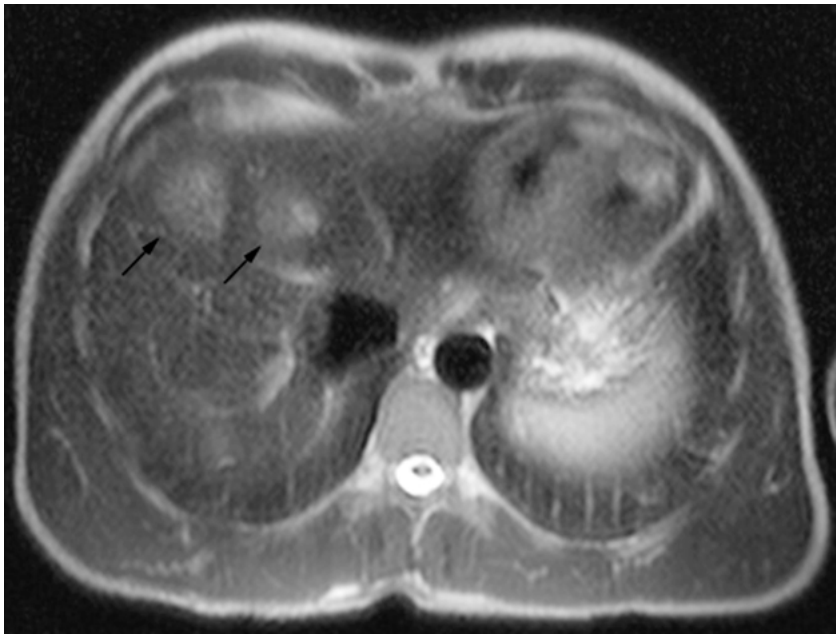

Fig. 1. MRI of the liver shows multiple small nodular lesions (*arrows*) that are hyperintense on T2-weighted images.

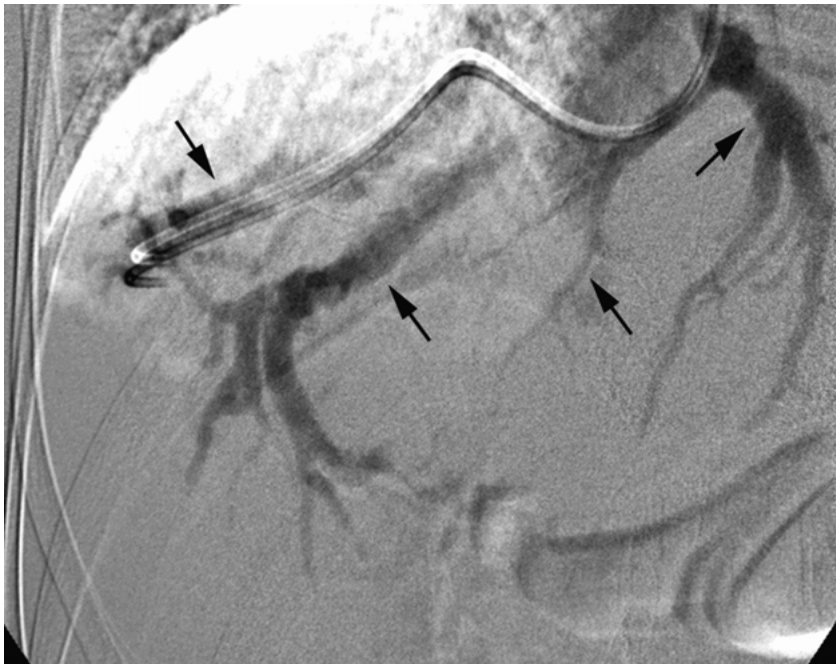

Fig. 2. Selective catheterization of the right hepatic vein and wedged CO_2 portography show no opacification of the portal vein. There is clear opacification of the hepatic veins (*arrows*) and venovenous communications.

intrahepatic portal vein branches associated with secondary multifocal nodular regenerative hyperplasia was suggested.

To confirm these vascular and parenchymal MRI findings, CO_2 wedged venography was performed through a right jugular approach. A 5-F Cobra catheter (Cobra C2, Cook Europe, Bjaeverskov, Denmark) was introduced under local anesthesia through the right internal jugular vein and placed in a wedged position in a right hepatic vein end branch. Subsequently, CO_2 was injected into the liver parenchyma by use of a manual injector (CO_2-Angioset, Optimed, Ettlingen, Germany). We injected 40 mL of CO_2 at a flow rate of 20 mL/s. Clear opacification of the right, middle, and left hepatic veins and communicating venules was demonstrated (Fig. 2). At the level of the catheter tip, clear penetration of CO_2 into the liver parenchyma was clearly visible, proving a good quality CO_2 wedged venography. However, opacification of portal vein branches could not be obtained (Fig. 2).

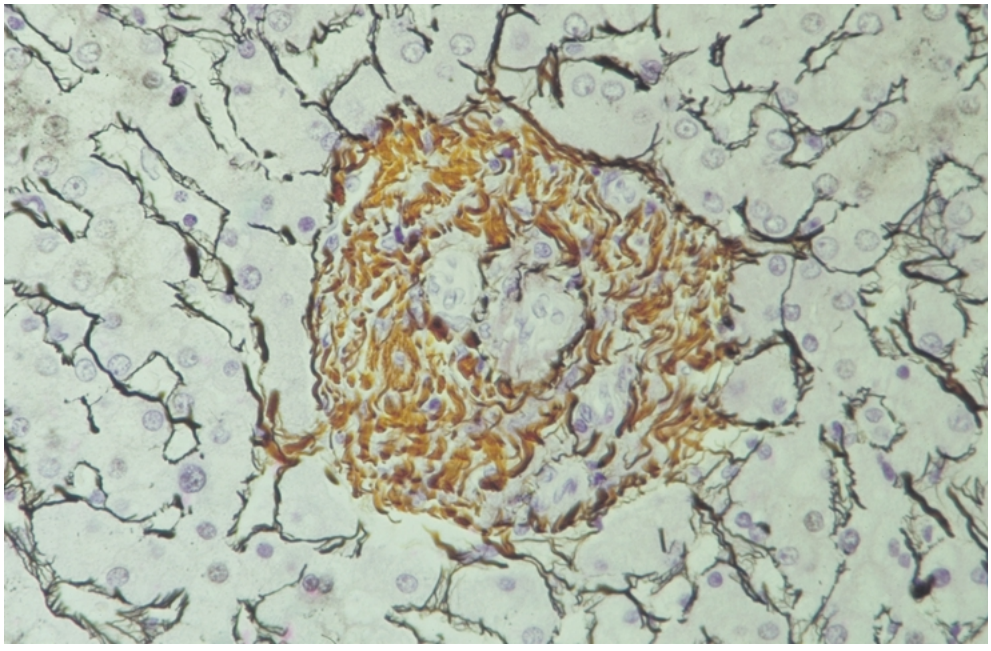

Fig. 3. A transjugular liver biopsy reveals nodular regenerative hyperplasia. Sinusoids are strongly dilated, which suggests shunting. In the liver acini there are bile ducts and hepatic arteries but no portal veins.

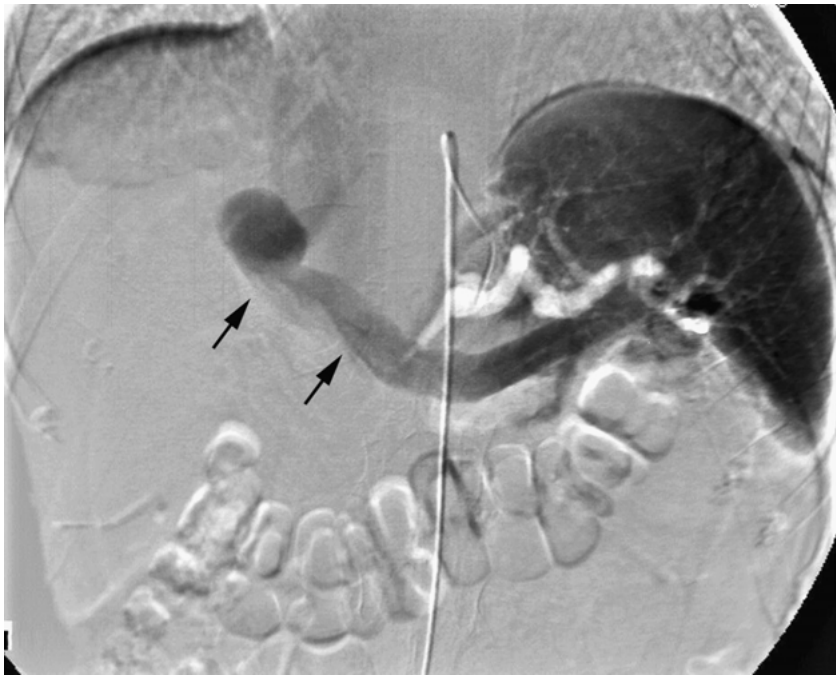

Fig. 4. Angiography through selective catheterization of the celiac trunk could not identify the presence of an intrahepatic portal vein. The splenic vein drains into the extrahepatic portal vein (*arrow*), which directly drains into the inferior vena cava.

Subsequently a 7-F long sheath was introduced over a stiff guidewire (Amplatz, Cook Europe) and four cylinders of liver tissue were biopsied (LABS-100, Cook Europe). Pathologic analysis of the collected biopsies revealed strongly dilated sinusoids, suggesting shunting, and liver acini presenting with bile ducts but without portal veins (Fig. 3). Based on these radiologic and pathologic findings, a definitive diagnosis of Abernethy malformation, type Ib according to the classification of Morgan and Superina [6], could be made. Additional celiac arteriography with venous phase confirmed the diagnosis (Fig. 4) but did not add additional findings to the diagnosis.

It was decided to follow the patient clinically, biochemically, and by hepatic ultrasound to exclude the potential development of liver tumors or potential progression into biliary cirrhosis.

Discussion

CAPV is an extremely rare anomaly that was first described by Abernethy in 1793 [7]. With the advances in

imaging modalities, there has been an increase in the number of cases and Murray et al. [8] recently published a literature review of 61 published cases.

The underlying embryonic mechanism of the development of an extrahepatic portosystemic shunt is unclear. However, its existence is not surprising because the portal vein and inferior vena cava are formed during the same time period, which explains the potential for congenital portosystemic anastomoses [9].

The commonly used classification of CAPV has been described by Morgan and Superina [6]. They classified the portosystemic shunt anomalies into two types. In type I, the liver is not perfused with portal blood, corresponding to a total end-to-end shunt. In type II, the liver is perfused with portal blood, corresponding to a partial end-to-side shunt. The type I lesion can further be subclassified into two types. In type Ia, the superior mesenteric vein and the splenic vein do not join and drain separately into the inferior vena cava or the iliac or renal vein. In type Ib, the superior mesenteric vein and splenic vein join to form a short extrahepatic portal confluence that drains into the inferior vena cava.

CAPV is frequently associated with a variety of other congenital anomalies including congenital heart disease and anomalies of the great vessels [10, 11], polysplenia [12], biliary atresia [12], renal tract and skeleton anomalies [8]. Affected patients can develop liver tumors, thus justifying a regular radiologic follow-up. These liver tumors can consist of benign lesions such as focal nodular hyperplasia, hepatocellular adenoma, or nodular regenerative hyperplasia [13, 14, 15] and of malignant lesions such as hepatocellular carcinoma or hepatoblastoma [3].

Currently a diagnosis of Abernethy malformation is mostly made by noninvasive cross-sectional imaging such as ultrasound, computed tomography, or MRI, which shows the shunt with or without additional intrahepatic portal vein branches [16]. These imaging techniques also can depict focal liver lesions, as in the present case. Although cross-sectional imaging in most cases can suggest the diagnosis, the definitive diagnosis can be made only with catheter angiography and by additional histologic analysis of the hepatic parenchyma that demonstrates the absence of hepatic portal venules within the portal triad. Histologic analysis is of diagnostic importance as Bellah et al. described a patient in whom intrahepatic portal branches were not revealed on ultrasound but clearly demonstrated on liver biopsy [17]. In our patient, the liver parenchyma was evaluated by MRI, which showed multifocal nodular hyperplasia and suggested the presence of a portocaval shunt. CO_2 wedged venography and histologic analysis of the liver parenchyma by transvenous liver biopsy, both obtained through the right transjugular approach, confirmed the tentative MR diagnosis of absence of the portal vein. CO_2 wedged venography is considered a good and safe technique for demonstrating the portal circulation [18, 19] and has other advantages over indirect portography that is obtained during visceral arteriography: only a venous puncture is required, free and wedged pressure measurements can be obtained, no iodinated contrast medium is injected, and transvenous liver biopsy can be made at the same time [20]. In the present case, indirect portography by an arterial approach did not add other significant information to our diagnosis and it can be considered redundant when performing CO_2 wedged venography and transvenous biopsy.

Treatment options strongly depend on the type of Abernethy malformation. In type II, occlusion of the shunt can be indicated in case of serious symptoms such as hepatic encephalopathy. The occlusion technique can be surgical [21] or, in selected cases, by percutaneous transcatheter coil occlusion [22]. In type I, close clinical, biochemical, and imaging follow-up is indicated. Only in case of severe symptoms of hepatic encephalopathy or with evidence of malignant liver nodules is liver transplantation the only option to cure the patient [3]. We proposed regular follow-up to our patient who presented with mild symptoms of type Ia Abernethy malformaton. In a case of malignant degeneration or exacerbation of symptoms, liver transplantation would be the only valuable treatment option.

In conclusion, this case demonstrates the potential value of CO_2 wedged venography associated with transvenous liver biopsy as a complementary diagnostic tool to MRI in the diagnosis of Abernethy malformation.

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