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Case Report

Usefulness of 320-row area detector computed tomography for the diagnosis of congenital absence of portal vein

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Abstract: Congenital absence of the portal vein (CAPV) is a rare malformation of the splanchnic venous system. It is easily misdiagnosed or never diagnosed due to lack of characteristic clinical manifestations. A case of CAPV complicating remodeling of the liver in a 24-year-old woman is here presented. Computed tomography and magnetic resonance imaging findings of the Type 2 portosystemic shunt are presented. Especially CT perfusion (CTP) imaging can reveal blood perfusion situation of the lesion, which is helpful for the diagnosis of CAPV. To our knowledge, this is the first report of application of CTP using 320-row CT in the diagnosis of CAPV.

Keywords: Portal vein, congenital absence, computed tomography, perfusion, magnetic resonance imaging

Introduction

Congenital absence of the portal vein (CAPV) is a rare malformation of the splanchnic venous system in which the intestinal and splenic venous drainage bypasses the liver and drains into the inferior vena cava (IVC), renal vein, or iliac vein through a congenital portosystemic shunt [1]. According to the literature [1, 2], most cases are accompanied by other abnormalities, such as cardiac and skeletal malformations and hepatic tumors. Focal nodular hyperplasia of the liver is a relatively common complication of CAPV. A patient with CAPV who had nodules in the liver and presented with remodeling of the liver is here reported. The case was accurately diagnosed using 4-dimensional CT angiography (4D-CTA) and CT perfusion (CTP) based on 320-row computed tomography (CT).

Case report

A 24-year old woman visited our hospital because of pain in the right kidney area. Abdominal ultrasonography (US) showed multiple hyperechoic lesions in the liver. She had no history of previous disease. Physical examination revealed no abnormal findings.

Non-contrast CT scan showed the following: the liver volume increased, there were multiple types of rounded nodules with slightly low density in the V and VIII segments of the right lobe of the liver, and their edges were clear. CT value of these shadows were about 27-39 HU (**Figure 1A**). Time-resolved 4-dimensional CT angiography (4D-CTA) was performed. Arterial phase: the hepatic inherent artery on the right side was more enlarged and distorted than that on the contralateral side. There were several visible distorted branches of right liver inherent artery distributed in the right hepatic lobe (**Figure 1B**). Portal venous phase: The anterior segment of the right branch of portal vein was interrupted. There were no obvious branches distributed in the corresponding area. The posterior segment of the right branch of portal vein was normal. (**Figure 1C** and **1D**). The serial images also showed that the right hepatic lobe was like a small liver because of the abnormal perfusion. The distorted hepatic inherent artery and its branches supplied this remodeling zone (**Figure 2A-D**).

After 4D-CTA, CT perfusion (CTP) imaging was performed and analyzed. Hepatic arterial and portal perfusion (HAP and HPP) and arterial

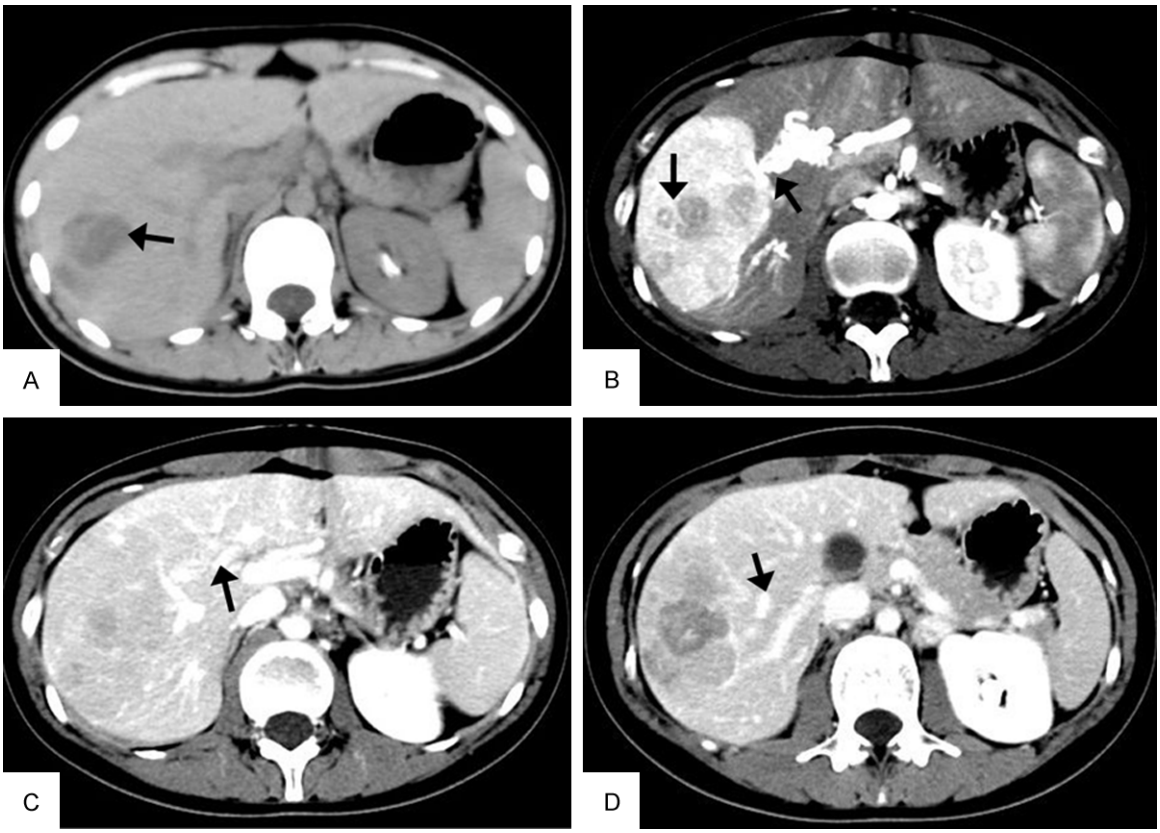

Figure 1. (A) Non-contrast CT image shows the increases in liver volume. Multiple hypodense lesions can be seen in segments V and VIII (arrow). (B) Arterial-phase CT scan shows hepatic inherent artery on the right side is more enlarged and distorted (arrow). Sections V and VIII of the right hepatic lobe is abnormally reinforced. The nodules are slightly reinforced, but relatively low perfusion. (C) Portal venous phase CT scan shows the interruption in the anterior segment of the right branch of the portal vein; no part of the portal vein is visible in the abnormal reinforced region in (B). (D) The normal posterior segment of right branch of portal vein can be seen.

perfusion fraction (APF) of normal and abnormal segments were calculated and compared (**Figure 3**). HAP, HPP, and APF for the abnormal area of the right hepatic lobe were 388.7 (ml/min/100 ml), 0 (ml/min/100 ml), and 99.9%. When HAP, HPP, and APF for the normal area of left hepatic lobe were 107.7 (ml/min/100 ml), 183.1 (ml/min/100 ml), and 37.3%. There was a significant difference between them. The perfusion parameters suggested that the abnormal area of the right hepatic lobe was almost fully supplied by the hepatic arteries.

Both 4D-CTA and CTP confirmed the diagnosis of congenital absence of portal vein (Type 2), complicating the remodeling of the liver. MRI images confirmed the presence of CAPV. Dynamic liver CT imaging revealed the nodules in the V and VIII segments were slightly enhanced, showing relatively low perfusion. Magnetic resonance imaging showed all of these lesions to be hyperintense masses on T1-weighted images (**Figure 4A**). Slight hyperintensity was visible on fat-suppressed T2-weighted images (**Figure 4B**). After gadolinium injection, slight enhancement was detected during the arterial phase and lesions became mildly hypointense on portal phase images (**Figure 4C**). A percutaneous fine-needle biopsy under ultrasound guidance was performed on the biggest liver nodule using a 20 gauge needle. It was confirmed FNH by pathology (**Figure 4D**).

Discussion

CAPV is a rare congenital malformation in which a portosystemic shunt prevents portal perfusion of the liver. Based on the hepatic parenchymal perfusion, two possible types of shunts have been proposed [3]. In the first type, there is complete diversion of the portal blood into the IVC through a complete portosystemic

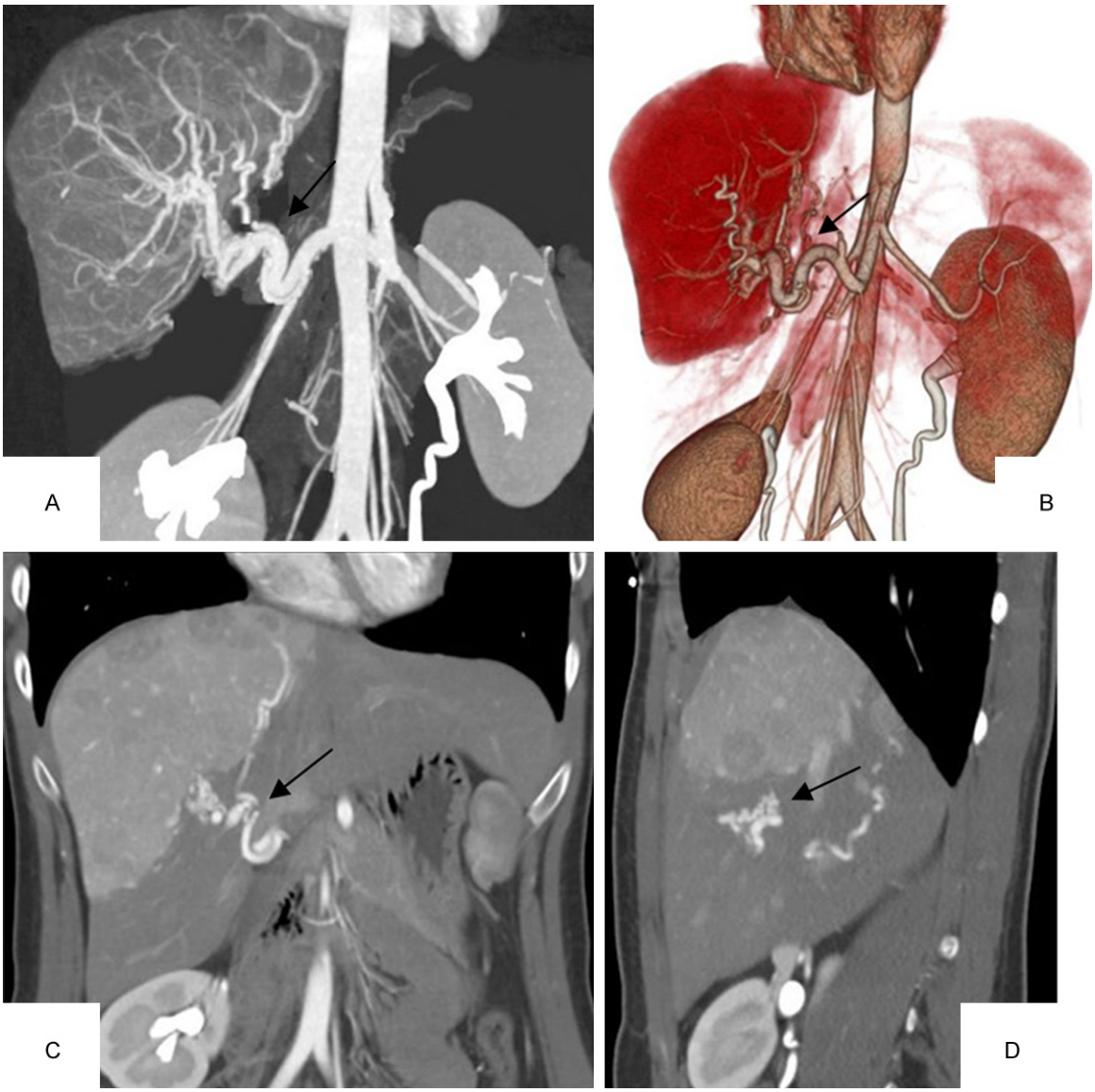

Figure 2. A. Coronal maximum intensity projection (MIP) image. B. VR image. C and D. Coronal and sagittal multiple planar reconstruction (MPR) images show the right hepatic lobe to resemble a small liver because of abnormal perfusion. The distorted hepatic inherent artery and its branches supply this remodeling zone (arrow).

shunt (side-to-end portosystemic anastomosis) bypassing the liver parenchyma. In the second type, as in the present case, only a portion of the portal blood drains into the systemic circulation through a partial shunt (side-to-side portosystemic anastomosis) partially conserving the PV supplies. The hepatic artery supplies the area lack of portal vein perfusion.

Time-resolved 4D-CTA and CTP using 320-row CT was performed in this case. During the arterial phase, the arterial blood perfusion was significantly higher than that of the surrounding normal liver tissue. During the venous phase, the anterior branch of portal vein was blocked. 4D-CTA data can be visualized from any angle during post-processing. The images can exhibit dynamic information. VR or MIP reconstructions can be used to assess the remolding of the liver in our case clearly. Perfusion imaging can detect regional and global changes in organ perfusion. It is an effective method of detecting hemodynamic characteristics [4, 5]. The 320-row CT system supports dynamic scanning over a range of approximately 16 cm and can provide more perfusion-related data

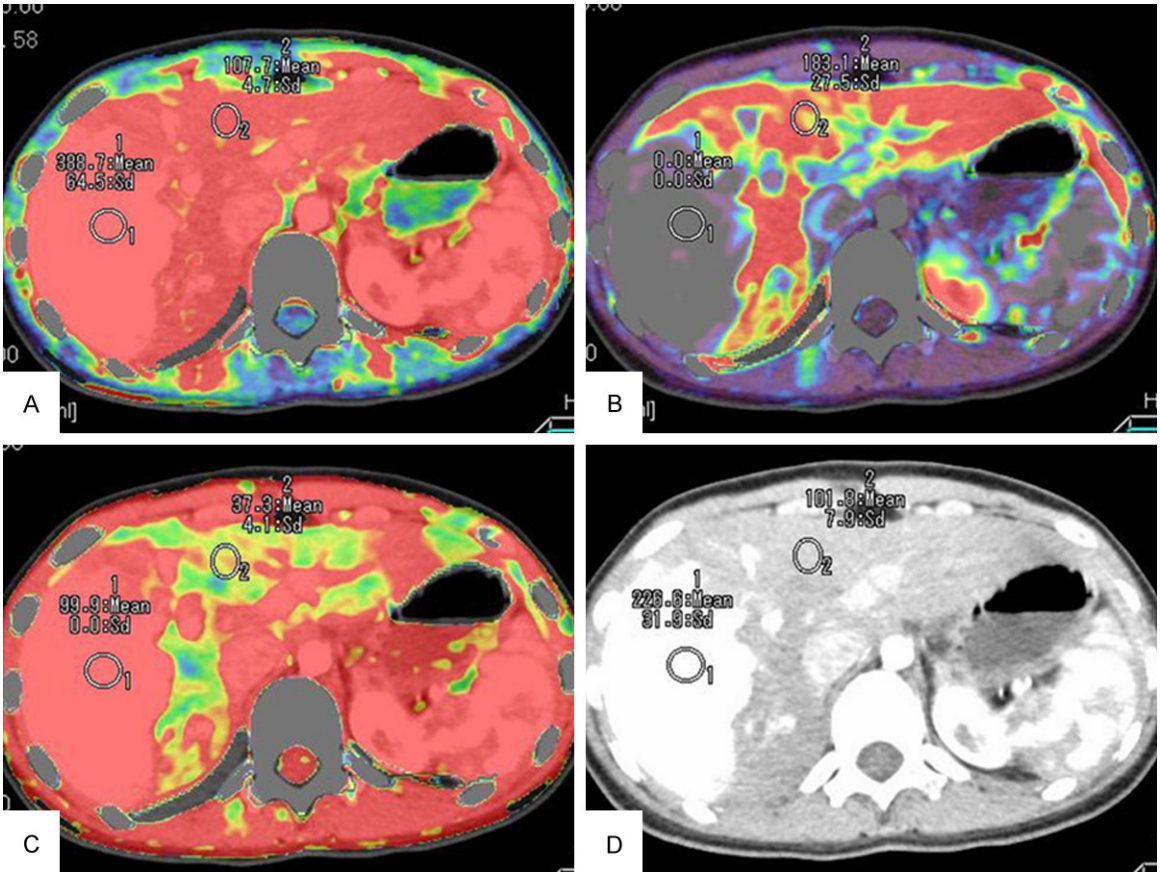

Figure 3. CTP images: CT hepatic arterial perfusion map shows an increase in arterial perfusion in the segments of V and VIII; the portal perfusion map shows a decrease in portal perfusion in the same area; arterial perfusion fraction map also shows an increase in arterial perfusion.

than conventional CT systems [6, 7]. With this system, the HAP, HPP, and APF of the abnormal area of the right hepatic lobe were found to be 388.7 (ml/min/100 ml), 0 (ml/min/100 ml), and 99.9%. The perfusion parameters suggested that the abnormal area of the right hepatic lobe lacked of portal vein perfusion, which was supplied by the branch of hepatic inherent artery, not by IVC.

No direct evidence of portosystemic shunts were found after studying CT images repeatedly. Neither was any evidence of other abdominal blood vessels found. This may be because the portosystemic shunt was too small to affect other related vascular hemodynamic vessels. The spatial resolution of CT images was much lower than that of digital subtraction angiography, so it failed to show the portosystemic shunts.

Multiple classes of relatively low perfusion lesions were visible in the right hepatic lobe. These may be related to the abnormal local blood supply or liver fault structure. This was confirmed by the fact that there was less perfusion of the right hepatic lobe than of the normal hepatic parenchyma around in portal venous phase and delayed phase CT scan. Several previous studies have investigated [8-10]. In patients with CAPV, some types of relatively low-perfusion lesions were confirmed by pathology as the regenerative nodules of the liver, hepatic focal nodular hyperplasia, or liver cyst and other benign lesions. In the current case, the biggest one of the nodules was confirmed as FNH by noodle biopsy.

Radiation exposure is a concern with 320-row CT. However, a low-dose radiation protocol, adaptive iterative dose reduction, has recently become available. The effective radiation dose for this 4D-CTA-CTP protocol amounted to 5.87 mSv. Finally, these new techniques of 4D-CTA and CTP based on 320-row CT scan can display the vascular malformation more clearly and

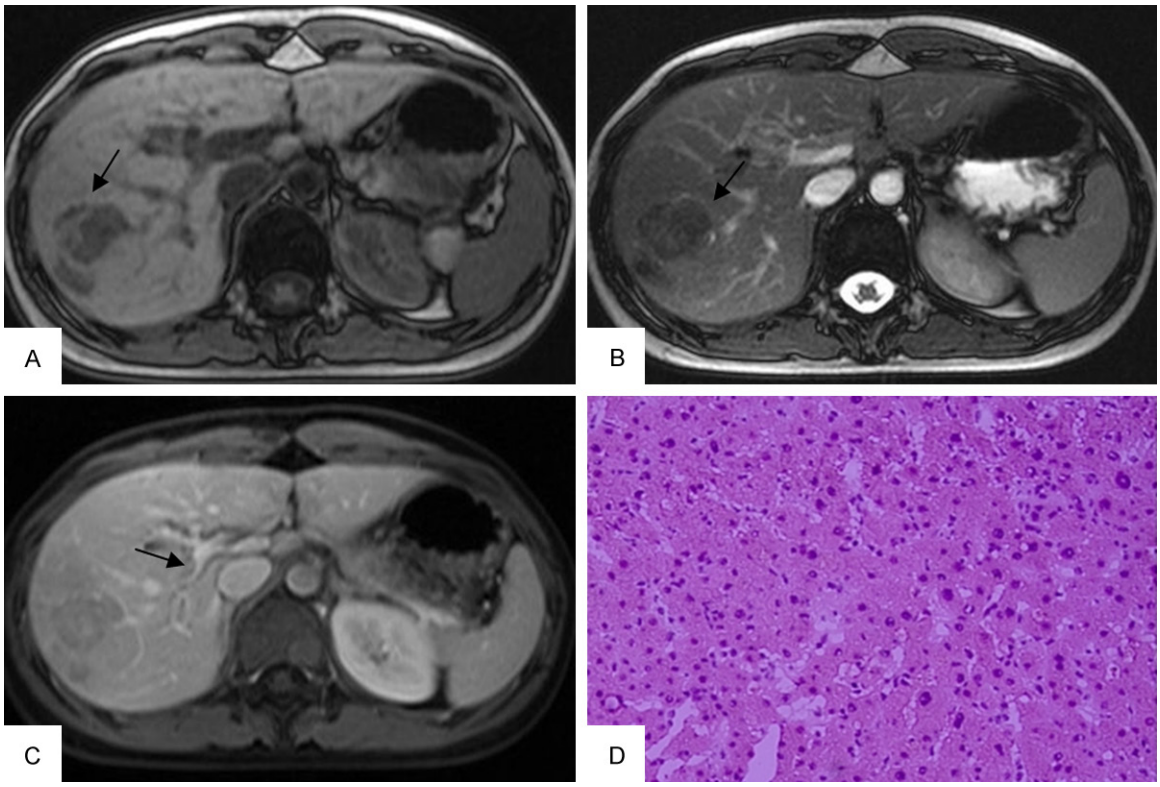

Figure 4. Lesions (arrows) are hypointense on (A) T1-weighted images and slightly hyperintense on (B) fat suppressed T2-weighted images. The absence of the anterior segment of right branch of portal vein (arrows) was visible on (C) contrast MRI images. (D) The nodule was confirmed to be FNH by pathology (hematoxylineosin, original magnification ×10).

accurately and the system can detect hemodynamic characteristics. These methods may play an important role in the evaluation of patients and development of treatment plans.

Disclosure of conflict of interest

None.

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