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Case Report

A case of incomplete duplication of the portal vein associated with multiple congenital anomalies ☆,☆☆

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ABSTRACT

Double portal veins are a duplication of the portal vein and normal portal vein with an accessory portal vein. We report a case of a 63-year-old asymptomatic female with double portal veins. There was fat accumulation observed in the area which was supplied by the first portal vein in normal position, and fatty sparing of the liver was observed in the area which was supplied by the second portal vein in the preduodenal position. The 2 portal veins were equal in size. Furthermore, the patient presented with multiple congenital anomalies, including double inferior vena cava, splenic lobulation, and accessory liver lobe. Therefore, double portal veins in our case were thought to be an incomplete duplication of the portal vein with multiple congenital anomalies.

Introduction

The portal vein (PV) is normally formed by the splenic and superior mesenteric vein junction, posterior to the neck of the pancreas. The main portal vein is divided into the right and left portal veins at the porta hepatis. There are 2 types of double PVs: a duplication of the portal vein (DPV) and normal portal vein with an accessory portal vein (APV) [1–9]. DPV is a very rare congenital anomaly, which has 2 separate portal veins course upward to the porta hepatis and divide into segmental branches [1–7]. Only a few cases with computed tomography (CT) evidence of DPV have been reported in the literature [1–5,7]. The APV has a small caliber with a caliber size which

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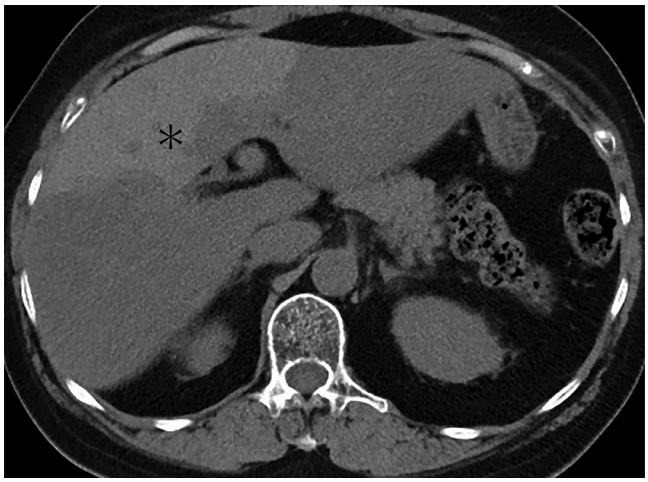

Fig. 1 – Unenhanced computed tomography (CT) abdomen shows fatty hepatic attenuation with focal sparing of the ventral segment of the right anterior sector (asterisk).

ranges from one-fifth to one-third of the main PV [9]. Criteria to distinguish DPV from APV remain unclear. We present a rare case of a 63-year-old female with double PVs suggesting incomplete DPV described by using a 0.4-mm slice thickness through photon counting CT.

Case report

A 63-year-old asymptomatic female was hospitalized as a result of a suspicion of a retroperitoneal cystic mass diagnosed using ultrasonography. Her serum liver enzyme levels (aspartate aminotransferase, 21 U/L; alanine aminotransferase, 25 U/L; cholesterol, 201 mg/dL; and total bilirubin, 0.3 mg/dL) and glucose level (91 mg/dL) were elevated. Unenhanced CT scans (Fig. 1) showed fatty liver (30 HU) with focal sparing (54 HU) at the anterior segment of the right lobe. Dynamic abdominal CT was performed in evaluating retroperitoneal cystic mass using photon-counting CT (NAEOTOM Alpha; Siemens Healthineers). We evaluated 2 dimensional images which include multi-planar projection of 0.4-mm slice thickness. Volume rendering and maximum intensity projection images of the vessels were reviewed as necessary.

One PV entered the normal liver hilum (PV1) and the other PV anterior to the pancreatic head (PV2) entered the liver inferiorly and supplied the ventral segment of the right anterior sector, on postcontrast enhanced CT scans (Fig. 2a-e). PV 1 flow was supplied from the splenic vein and superior mesenteric vein (SMV) (Fig. 3). Flow of the PV 2 was supplied from the SMV (Fig. 3). Both PVs connected the bridging part of the SMV, and the inferior mesenteric vein joined into the bridging part of the SMV (Fig. 3a, b). The ranges of PV1 and PV2 were almost equal and both PVs showed similar contrast- enhancement in all phases. The right gastric vein joined into the PV1, and the right gastroepiploic vein joined into the PV2. The splenic artery and the left gastric artery branched from the celiac artery, the left gastric artery branched from the left hepatic artery, and the common hepatic artery branched from the superior mesenteric artery. The left ligament of teres was normal. A small accessory lobe of the liver adjacent to the gallbladder was noted (Fig. 2b). There was a double inferior vena cava (Fig. 2b, Fig. 3), splenic lobulation (Fig. 1c), and discrete lobulations of the pancreatic head tissue as other congenital abdominal anomalies. Liver hemangiomas in the right lobe, left adrenal adenoma, and retroperitoneal lymphangioma coexisted. On the chest CT images, the right lung showed 2 lobes with bilateral incomplete inter-lobal fissure. Magnetic resonance cholangiopancreatography revealed a normal bile duct system. The common bile duct normally ran parallel to PV1.

Discussion

The portal venous system evolves through a complex process which includes the selective persistence of parts of the vitelline venous system and communication with the umbilical venous system during the development of the liver between the 4th and 12th weeks of gestation [10–12]. There are three cross-communications between the vitelline veins: cranial, middle, and caudal [1,11,12]. Cranial and caudal communications lie ventral to the gut, but the middle communication lies dorsal to the gut. There is selective involution of the venous network, and the caudal part of the right vitelline vein and the cranial part of the left vitelline vein progressively obliterate, which produces the PV [1,10–12]. The major anatomic variants of the portal venous system include the congenital absence of the PV, absent branching of the PV, and preduodenal PV [1–7,10,13,14]. The APV is a rare anomaly, wherein a small-caliber vein directly branched from the PV [8,9], and DPV is a very rare developmental anomaly, which has been described only in case reports [1–7]. DPV has separate portal veins on its upward course to the porta hepatis. There are 2 types of DPV reported [1–7]. One PV was derived from the splenic vein and the other PV was derived from the SMV in the first type [2]. The splenic vein and SMV joined to form a PV at the anterior portion of the pancreatic head and 2 PVs bifurcated outside the liver in the second type [1,4–6]. Two bifurcated PVs coursed to the porta hepatis in parallel in some cases [4,5]. In our case, the double portal veins could be an incomplete DPV since bridging of the SMV between the 2 PVs was present, and both PVs were of equal size. Additionally, PV2 was a preduodenal portal vein as well as previously reported DPVs [1–3,6]. However, a preduodenal portal vein was a large main portal vein, and a small tortuous portal vein mimicked a cavernous transformation which branched from the splenic vein in Yangs' case [3]. DPVs were in a preduodenal position in a case reported by Snavely et al. [6]. A preduodenal portal vein results from the abnormal obliteration of this dorsal connection and a persistence of flow through the caudal-ventral anastomosis [13]. Kitagawa et al. [7] reported a case of DPV with prepancreatic postduodenal portal vein (PPPV), and the PPPV only entered segment 2 of the liver. A majority of the DPV cases could present with a preduodenal location; however, other cases could not be evaluated if located between the PV and duodenum or pancreatic head [4,5]. The preduodenal PV is typically associated with other congenital

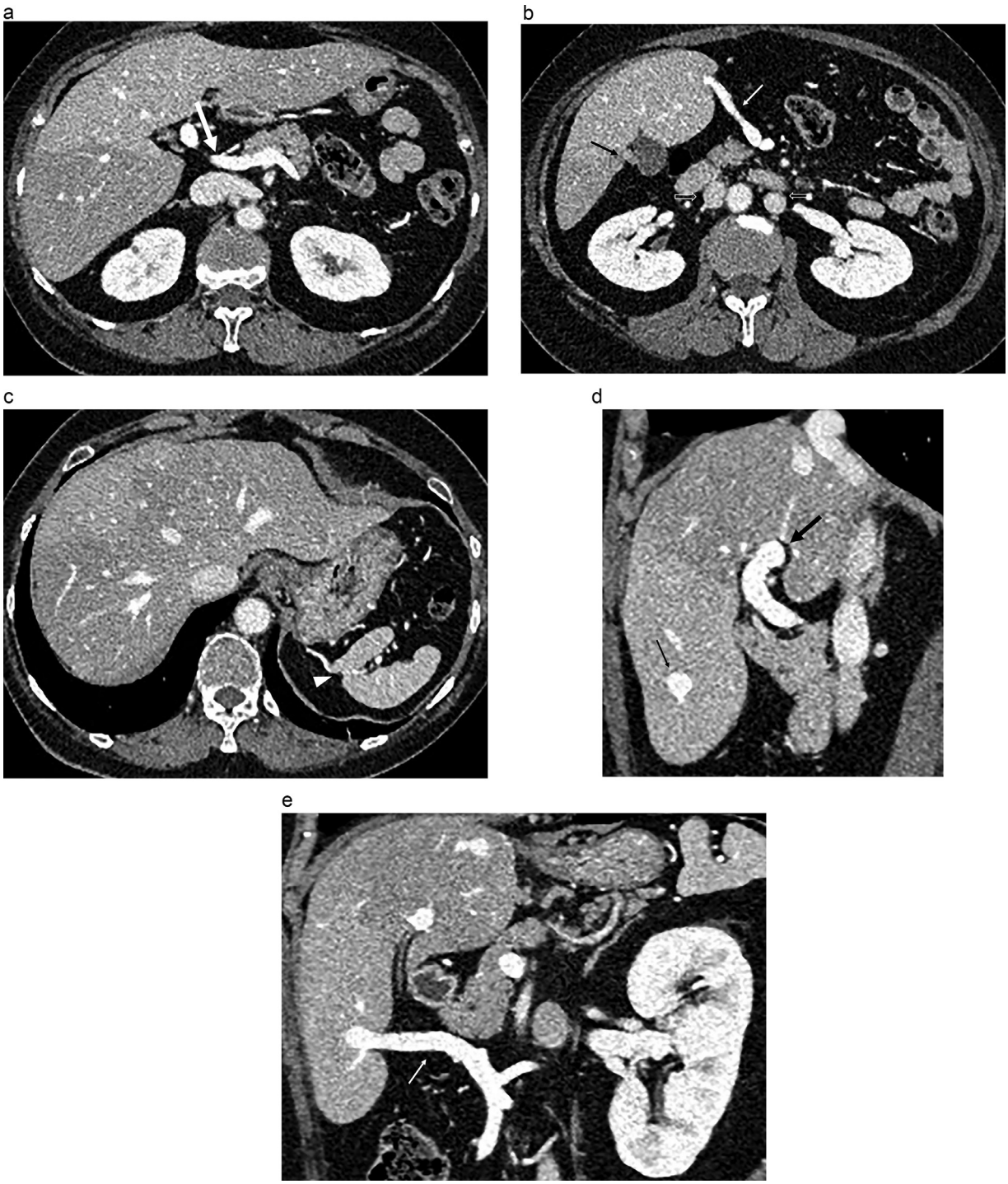

Fig. 2 – Contrast enhanced CT images. (a-c). Axial images show the first portal vein (PV) entered into the normal liver hilum (PV 1: large white arrow on a), the second PV anterior to the pancreatic head entered the liver inferiorly (PV 2: small white arrow on b), a small accessory liver is adjacent to the gallbladder (black arrow on b), and double inferior vena cava (white open arrows on b) is present. Note splenic lobulation (small arrowhead on c). (d) Sagittal image shows 2 portal veins (PV 1: large black arrow, PV 2: small black arrow). (e) Oblique coronal image shows the PV2 (small white arrow) connected to the SMV.

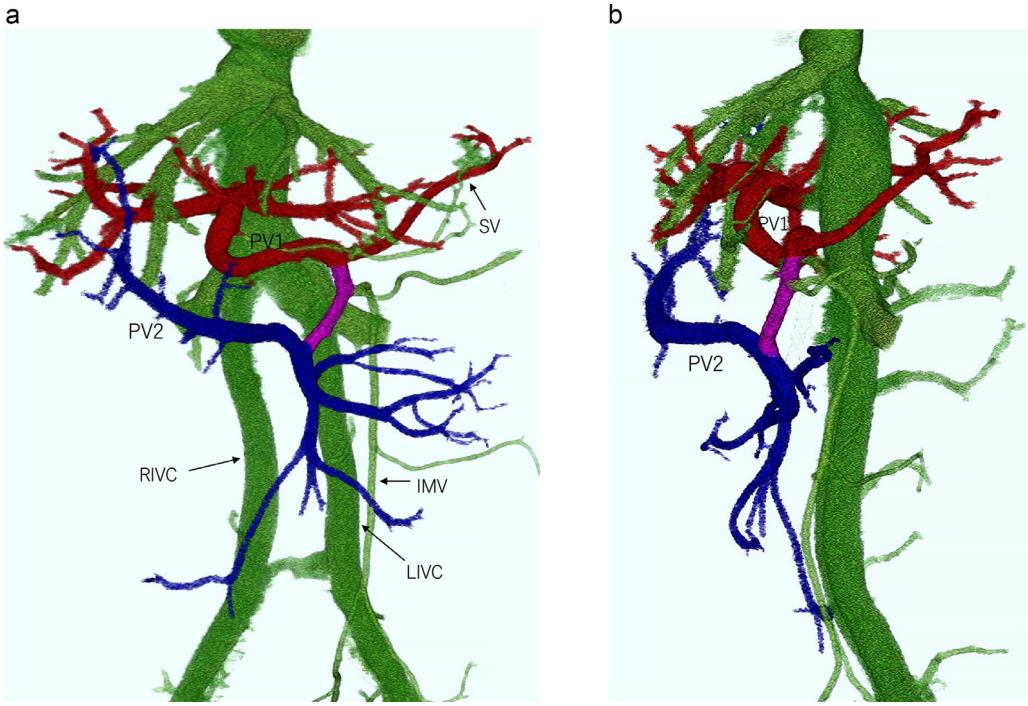

Fig. 3 – (a, b) Volume rendering image on frontal view (a) shows PV1 and PV2 were connected to a part of the superior mesenteric vein (violet vessel). Volume rendering image on lateral view (b) shows the PV2 ascending to the ventral right lobe. Green vessels: veins. Red vessels: PV1 and splenic vein. Blue vessels: PV2 and SMV. RIVC: right inferior vena cava. LIVC, left inferior vena cava; IMV, inferior mesenteric vein; SV, splenic vein.

anomalies, which include heterotaxia or polysplenia syndrome, situs anomalies, or biliary atresia although the preduodenal PV can occur as an isolated defect [13,14]. Our case was associated with splenic lobulation, and other coexistent congenital anomalies were double inferior vena cava, accessory liver lobe, hypoplasia of the pancreatic head, and abnormal interlobar pleura. However, in our case, the patient was asymptomatic, and the findings were incidentally found on CT examination. To our best knowledge, DPV associated with congenital anomalies has not yet been reported. An autopsy case of DPV presented normal arterial, venous, and bile duct systems [6]. Therefore, the cause of the coexistence of DPV and other congenital anomalies in our case remains unclear.

The blood supply of the DPV was different for each case [1,3,7]. The blood supply area was beyond the hepatic segment, and an intrahepatic communication of the DPV may exist [1,3]. PV2 supplied the ventral segment of the right anterior sector in our case.

There are 2 cases of DPV with fatty liver reported as well as in our case [1,3]. The difference in fat infiltration was observed, which were supplied by the 2 PVs, and the fat-spared area was supplied from the main SMV flow with decreased splenic venous flow in the three cases which include our case [1,3]. We hypothesize that fat sparing results from the main SMV flow with decreased pancreatic enzyme levels such as insulin.

DPV may cause abdominal pain and induce portal hypertension, which leads to the development of esophagogastric varices [2,6]. Portal hypertension results from the partial obstruction to the portal flow caused by the abnormal course [2].

DVPs are observed as nodular or massive lesions at the hepatic hilum; therefore, these should not be mistaken for the other disease entities, for example, lymphadenopathies [4]. Knowledge of the variations of the portal venous system which includes DPV helps in the proper planning of patient management, especially for interventional surgery and laparoscopic surgery [3]. An evaluation of DPV and its associated acquired and congenital anomalies is important, and recent high-resolution CT is helpful in determining the accurate anatomy.

Conclusion

We reported a case of double PVs with multiple congenital and acquired anomalies. There was fat accumulation observed in the area supplied by PV 1 with normal position, while sparing of fatty liver was observed in the area supplied by PV2 in the preduodenal position. The double PVs in our case was suggestive of an incomplete DPV because of the bridging of the SMV between the 2 PVs in the same size.

Patient consent

Informed consent for publication of their case was obtained from the patient.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2023.05.043.

REFERENCES

[1] Yang DM, Kim HC, Kim SW. Hepatic fat accumulation with sparing associated with portal vein duplication. Clin Imaging 2014;38(4):550–2. doi:10.1016/j.clinimag.2014.01.011.

[2] Dighe M, Vaiya S. Duplication of the portal vein: a rare congenital anomaly. Br J Radiol 2009;82(974):e32–4. doi:10.1259/bjr/81921288.

[3] Yang Q, Li J, Wang H, Wang S. A rare variation of duplicated portal vein: left branch derived from splenic vein mimicking cavernous transformation. BMC Gastroenrerol 2021;21(1):404. doi:10.1186/s12876-021-01970-8.

[4] Ito K, Matunaga N, Mitchell DG, Fujita T, Honjo K, Awaya H, et al. Imaging of congenital abnormalities of the portal veinous system. AJR Am J Roentgenol 1997;168(1):233–7. doi:10.2214/ajr.168.1.8976951.

[5] Ozbülbül NI. Congenital and acquired abnormalities of the portal venous system: multidetector CT appearances. Diagn Interv Radiol 2011;17(2):135–42. doi:10.4261/1305-3825.DIR.3124-09.1.

[6] Snavely JG, Breakell ES. Fatal hemorrhage from esophageal varices. Am J Med 1954;16(3):459–64. doi:10.1016/0002-9343(54)90361-7.

[7] Kitagawa S. Anomalous duplication of the portal vein with prepancreatic postduodenal portal vein. J Rural Med 2022;17(4):259–61. doi:10.2185/jrm.2022-009.

[8] Araki T, Hayakawa K, Hayashi S, Uchiyama G. Accessory lateral segmental branch of the portal vein. Radiology 1985;155(1):24. doi:10.1148/radiology.155.1.3975402.

[9] Zhang JS, Wang YP, Wang MQ, Yang L, Xing CC, Yu M, Cui Z. Diagnosis of an accessory portal vein and its clinical implications for portsystemic shunts. Cardiovasc Intervent Radiol 1996;19(4):239–41. doi:10.1007/BF02577642.

[10] Lee WK, Chang SD, Duddalwar VA, Comin JM, Perera W, Lau WF, Bekhit EK, et al. Imaging assessment of congential and acquired abnormalities of the portal venous system. RadioGraphics 2011;31(4):905–26. doi:10.1148/rg.314105104.

[11] Marks C. Developmental basis of the portal venous system. Am J Surg 1969;117(5):671–81. doi:10.1016/0002-9610(69)90404-8.

[12] Corness JA, McHugh K, Roebuck DJ, Taylor AM. The portal vein in children: radiological review of congenital anomalies and acquired abnormalities. Pediatr Radiol 2006;36(2):76–96. doi:10.1007/s00247-005-0010-4.

[13] Mimatsu K, Oida T, Kano H, Kawasaki A, Fukino N, Kida K, et al. Preduodenal portal vein, intestinal malrotation, polysplenia, and interruption of the inferior vena cava: a review of anatomical anomalies associated with gastric cancer. Surg Radiol Anat 2012;34(2):179–86. doi:10.1007/s00276-011-0894-7.

[14] Yi SQ, Tanaka S, Tanaka A, Shimokawa T, Ru F, Nakatani T. An extremely rare inversion of the preduodenal portal vein and common bile duct associated with multiple malformations. Report of an adult cadaver case with a brief review of the literature. Anat Embryol (Berl) 2004;208(2):87–96. doi:10.1007/s00429-003-0377-7.