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CASE REPORT

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An unusual developmental anomaly of duplicated portal vein

Genfa Du[1], Jie Li[2], Yueyong Qi[1]

[1]*Department of Radiology, Chongqing Songshan General Hospital, Chongqing, China*
[2]*Department of Paediatrics, Women and Children's Hospital of Chongqing Medical University/Chongqing Health Centre for Women and Children, Chongqing, China*

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Background: *Portal vein (PV) duplication is a rare developmental anomaly, but it plays an important role in the diagnosis and management of disease for radiologists and surgeons.*

Materials and methods: *A new variant of PV duplication with vein fenestration leading to choledochal stenosis and dilatation and thrombus was identified by computed tomography angiography (CTA) on a 59-year-old woman with a history of gallstones.*

Results: *A second PV originated from the superior mesenteric vein (SMV), which split into 2 branches encircling the common bile duct to form a vein fenestration, leading to choledochal stenosis and dilatation, with thrombus formation at the confluence.*

Conclusions: *This case report adds to the existing body of knowledge about the variation of the PV system. We present an embryological perspective for the case, which suggests the possibility of similar occurrences.* (Folia Morphol 2024; 83, 3: 737–739)

Keywords: portal vein duplication, variation, computed tomography angiography

INTRODUCTION

Portal vein (PV) duplication is a rare developmental anomaly described only in case reports [7]. Venous fenestration, which refers to bifurcations that reunite before drainage, has only been reported in cases [1]. Computed tomography angiography (CTA) allows the identification of the PV anatomy, and multiplanar reconstruction is ideal for displaying PV anatomy.

CASE REPORT

A 59-year-old female presented with a history of gallstones with episodic pruritus. Liver function tests were normal. CTA (Fig. 1a, b) showed the main branches of the duplicated PV system. PV1 was derived from the junction of the superior mesenteric vein (SMV) and the splenic vein (SV). PV1 entered the liver and supplied segments as normal. PV2 was derived from the SMV and coursed posterior to the descending duodenal and pancreatic neck. PV2 split into 2 branches that encircled the common bile duct, resulting in choledochal stenosis and dilatation (Fig. 2), before converging to form a vein fenestration with a thrombus at the confluence. It then coursed upward into porta hepatis and divided into the right portal vein (RPV) and the left portal vein (LPV). The LPV supplied segments II and IV of the liver; the RPV

Address for correspondence: Yueyong Qi, Chongqing Songshan General Hospital. No. 69, Xingguang Avenue, Yubei District, Chongqing, China; tel: 17783200118, e-mail: 1156455432@qq.com

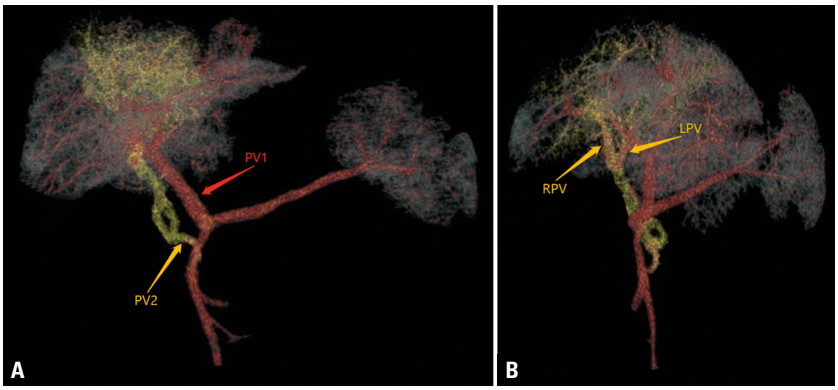

Figure 1. Volume-rendered images of CTA. **A.** PV1 derived from the junction of SMV and SV. PV2 derived from SMV forming the vein fenestration. **B.** PV2 dividing into RPV and LPV after entering the liver. CTA — computed tomography angiography; LPV — left portal vein; RPV — right portal vein; SMV — superior mesenteric vein; SV — splenic vein.

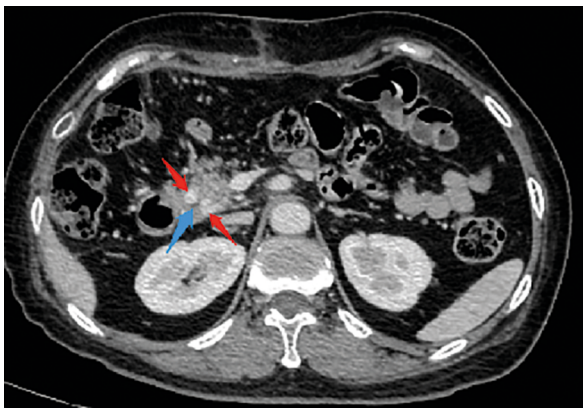

Figure 2. Axial thick slab reconstruction image of computed tomography angiography (CTA) showed the common bile duct (blue arrow) compressed by the bifurcated canals of vein fenestration (red arrow).

supplied segment V and possibly segments III, VI, and VII (poor visualisation due to the thinness of the branching vessels or inadequate resolution). The hepatic anatomy appeared normal.

The patient then underwent cholecystectomy for gallstones and received aspirin for thrombus. The thrombus disappeared after one month of follow-up.

DISCUSSION

PV develops from the bilateral vitelline and umbilical veins in 4–10 weeks. The vitelline veins form anastomotic network around the duodenum, consisting of caudal–ventral, dorsal, and cranial–ventral anastomoses. There is joining of the umbilical veins to the vitelline veins. Then, the right umbilical vein disappears while the left umbilical vein partly remains. The caudal and cranial parts of the anastomoses obliterate, and the dorsal anastomosis becomes the PV [4, 6].

There are several different ways in which the PV can be duplicated, depending on the position to the duodenum. However, none of the currently reported modalities apply to our case [4–7]. The PV2 could be a remnant of the right umbilical vein, and we might assume that the right umbilical vein continued to connect with both the ductus venosus and the SMV during the embryonic period (Fig. 3A–C). This is also conceivable because PV1 was located posterior to the duodenum and developed normally, proving that it was not the result of alteration in the anastomotic occlusion between vitelline veins. The aetiology of fenestration is unknown. According to the theories that have been proposed, it may be the result of obstructed growth by the common bile duct during development. Alternatively, vascular weakness associated with turbulent flow could result in endothelial reorganisation into separate vessels during development [1].

In the 6 cases of PV duplication reported to date, there were 2 cases with combined portal hypertension, one of which suffered fatal gastrointestinal haemorrhage [4, 5], 2 cases with fatty infiltration of the liver [6, 7], and 2 cases without complications [2, 3]. In our case, there was no evidence of portal hypertension or hepatic abnormalities, which proved that it was not the result of cavernous transformation, but thrombus was seen in the trunk of PV2. We suggested that it was related to the haemodynamics caused by the abnormal course, and vein fenestration has been reported to be a predisposing factor for deep vein thrombus. In addition, compression of the common

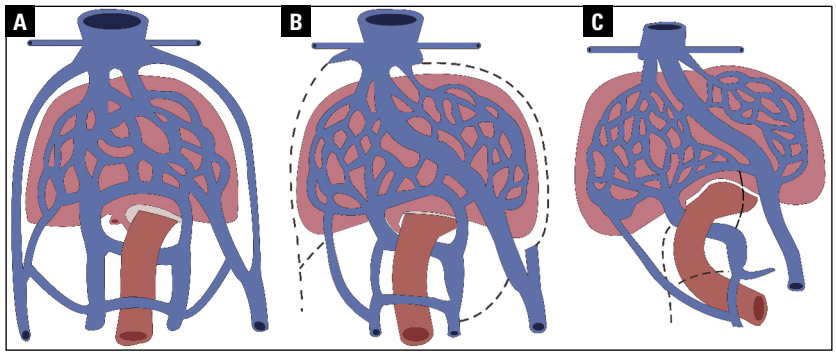

Figure 3. Schematic images of portal vein development. **A.** Foetal "symmetrical" hepatic circulation. If the right umbilical vein partly remained (**B**), it would lead to development of the portal vein (PV) anomaly in our patient (**C**).

bile duct by PV2 led to biliary stenosis and dilatation and triggered gallbladder stones.

CONCLUSIONS

Despite its rarity, PV duplication is of great surgical importance. Radiologists and clinicians need to be aware of this anomaly to help plan the patient's management properly.

ARTICLE INFORMATION AND DECLARATIONS

Ethics statement

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the medical Ethical Committee of the Chongqing Songshan General Hospital. Verbal consent was obtained from the patients, identifying data were removed, and all image data were obtained from routine imaging at our institution. Therefore, written informed consent was waived by our institutional review board.

Author contributions

GFD conceptualised and designed the study, drafted the initial manuscript, and reviewed and revised the manuscript. JL and YYQ supervised the project and critically revised the important intellectual content.

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Conflict of interest

The authors have no conflict of interest to declare.

REFERENCES

1. Contrera KJ, Aygun N, Ward BK, et al. Internal jugular vein duplication and fenestration: case series and literature review. Laryngoscope. 2016; 126(7): 1585–1588, doi: 10.1002/lary.25743, indexed in Pubmed: 26498831.
2. Dighe M, Vaidya S. Case report. Duplication of the portal vein: a rare congenital anomaly. Br J Radiol. 2009; 82(974): e32–e34, doi: 10.1259/bjr/81921288, indexed in Pubmed: 19168687.
3. Kitagawa S. Anomalous duplication of the portal vein with prepancreatic postduodenal portal vein. J Rural Med. 2022; 17(4): 259–261, doi: 10.2185/jrm.2022-009, indexed in Pubmed: 36397802.
4. Marks C. Surgical implications of portal venous system malformation. Ann R Coll Surg Engl. 1974; 55(6): 299–306, indexed in Pubmed: 4614690.
5. SNAVELY JG, BREAKELL ES. Fatal hemorrhage from esophageal varices, due to malformations and congenital stenoses in portal venous system. Am J Med. 1954; 16(3): 459–464, doi: 10.1016/0002-9343(54)90361-7, indexed in Pubmed: 13138614.
6. Yang DMo, Kim HC, Kim SW. Hepatic fat accumulation with sparing associated with portal vein duplication. Clin Imaging. 2014; 38(4): 550–552, doi: 10.1016/j.clinimag.2014.01.011, indexed in Pubmed: 24679652.
7. Yang Q, Li J, Wang H, et al. A rare variation of duplicated portal vein: left branch derived from splenic vein mimicking cavernous transformation. BMC Gastroenterol. 2021; 21(1): 404, doi: 10.1186/s12876-021-01970-8, indexed in Pubmed: 34702178.