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ABSTRACT

Background: Portal vein (PV) duplication is a rare developmental anomaly but has 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), split into two branches that 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.

Keywords: portal vein duplication, variation, computed tomography angiography

INTRODUCTION

Portal vein (PV) duplication is a rare developmental anomaly described only in case reports [1]. Venous fenestration, which refers to bifurcations that reunite before drainage, has only been reported in cases [2]. Computed tomography angiography (CTA) allows identification of the PV anatomy, 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 (Figure 1ab) 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 two branches that encircled the common bile duct, resulting in choledochal stenosis and dilatation (Figure 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 supplied the V segment and possible III, VI, VII segments (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

The PV develops from 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 [3][4].

There were a number of different ways in which the PV can be duplicated depending on the position to the duodenum. However, none of the currently reported modalities applied to our case [1] [3] [4] [5]. The PV2 could be a remnant of the right umbilical vein, we might assume that the right umbilical vein continued to connect with both the ductus venosus and the SMV during the embryonic period (Figure 3abc). 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 etiology of fenestration is unknown. According to the possible 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 reorganization into separate vessels during development [2].

In the six cases of PV duplication reported to date: two cases with combined portal hypertension, one of which suffered fatal gastrointestinal hemorrhage [4] [5]; Two cases with fatty infiltration of the liver [1] [3]; two cases without complications [6][7]. 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 hemodynamics 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 bile duct by PV2 lead 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.

REFERENCES

1. 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.

2. 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.

3. Yang DM, 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.

4. Marks C. Surgical implications of portal venous system malformation. Ann R Coll Surg Engl. 1974;55(6):299-306.

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.

6. 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.

7. 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.

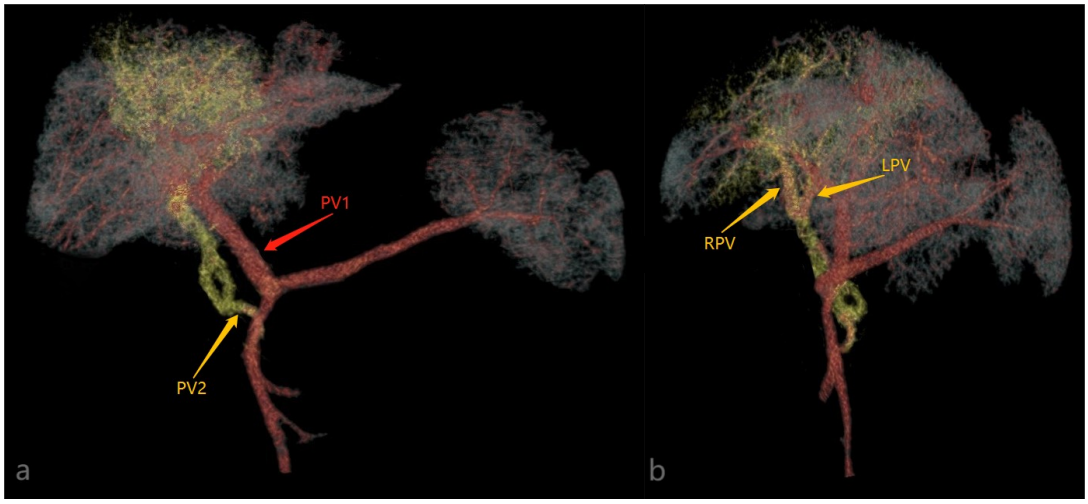

Figure 1. Volume-rendered images of CTA. (**a)** PV1 derived from the junction of SMV and SV. PV2 derived from SMV and formed the vein fenestration. (**b)** PV2 divided into RPV and LPV after entering the liver.

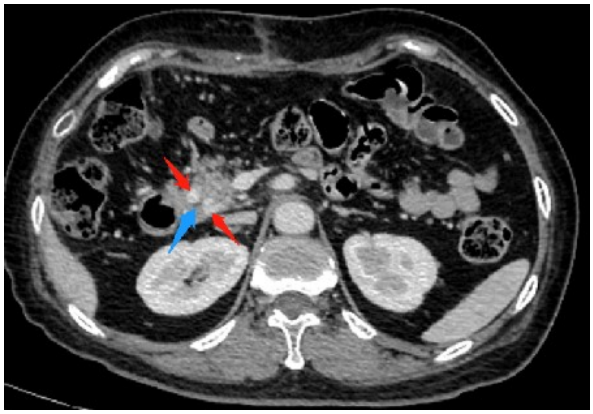

Figure 2. Axial thick slab reconstruction image of CTA showed the common bile duct (blue arrow) compressed by the bifurcated canals of vein fenestration (red arrow).

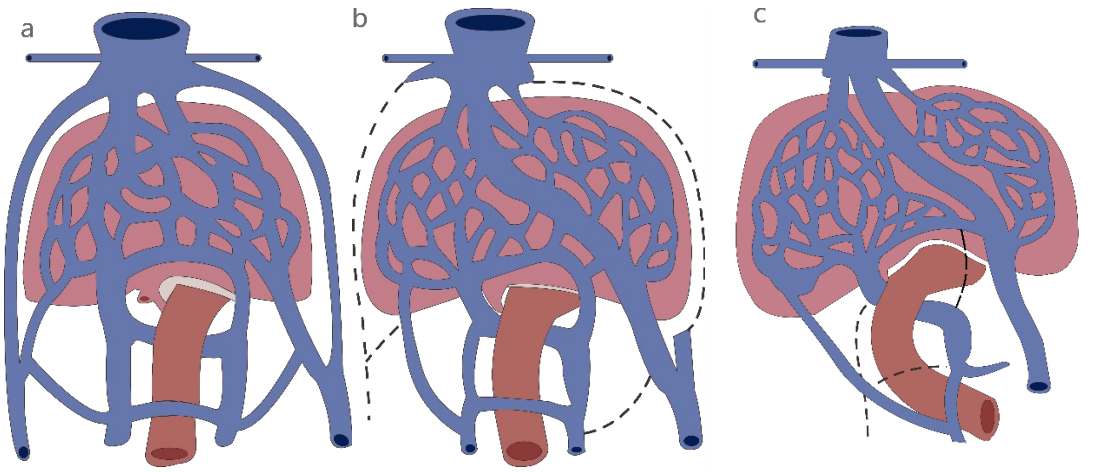

Figure 3. Schematic images of portal vein development. (**a)** Fetal “symmetrical” hepatic circulation. If the right umbilical vein partly remained (**b)**, it would lead to development of the PV anomaly in our patient (**c)**.