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Minireview and case report: Duplication of the portal vein and combinations

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ABSTRACT

This is a minireview and case report on a concurrent duplication of the portal vein and a duplication of the left renal vein. The portal vein system supplies about seventy five percent of the blood for the liver and is involved in the manifestation of several liver diseases such as portal hypertension and portosystemic shunts. The vitelline veins and their anastomoses are involved in this anatomical variation during development. Similar to that also variations or malformations of the renal veins resulted from early primitive structures and their obliteration or non obliteration, respectively. Two rather rare anomalies of two venous systems might have been a coincidence or a common cause.

1. The background

The portal venous system plays an important role in the correct functioning of abdominal organs as it drains the blood from the stomach, intestines, spleen and pancreas to lead it to the liver. Thereby 75% of the blood reaching the liver comes out of this portal venous system [1]. The rest is arterial blood reaching the organ through its vasa privata. The main blood vessel of this portal venous system is the vena portae which - in the majority of people - is formed by the confluence of the superior mesenteric vein and the splenic vein. The inferior mesenteric vein may drain into the splenic or superior mesenteric vein or - in rare cases - end directly in the portal vein [1]. The veins normally lie dorsal to the pancreas. Together with the proper hepatic artery and the bile duct the portal vein passes through the porta hepatis whereby the portal vein is dorsal to the other structures. Inside the organ it separates into a right and left branch which belong to different liver segments. In most individuals the left vein belongs to segment I-IV and the right one shows a division in an anterior (segments V and VIII) and a posterior component (segments VI and VII) [1]. In general the vascular branches of bile duct, hepatic artery and portal vein run parallel to each other forming Glisson's trias.

The portal venous system is involved in the manifestation of different diseases such as, for example, infracardial total anomalous pulmonary venous connection, portal vein thrombosis, portosystemic shunts or (cirrhotic or non-cirrhotic) portal hypertension [1]. Various anatomic anomalies have been described, including - amongst others - hypoplasia, atresia and duplication [1–5]. Smaller alternations and variants of the portal venous system are found in up to 35% of people [1].

Another venous system located in the abdomen includes the veins of the urogenital tract. Major parts of this system are the two renal veins merging into the inferior vena cava. Thereby, the left testicular/ovary vein and the left adrenal vein drain into the left renal vein, while the corresponding veins on the right side drain independently into the vena cava. To reach the inferior vena cava the left renal vein has to cross the aorta ventrally, resulting in the nutcracker position of the venal blood vessel between abdominal aorta and superior mesenteric artery. This position of the left renal vein can lead to the anterior nutcracker syndrome. A posterior nutcracker syndrome is possible if a second left renal vein is present and is located dorsal of the aorta, leading to similar symptoms as the anterior nutcracker syndrome [6]. The following study presents a description of a case with a duplication of two different venal blood vessels in one patient.

2. The case

The combined variation of the venal blood vessels was an incidental finding in a cadaver in our anatomical dissection course (male, 84 years, respiratory failure during uro-sepsis). We noticed a duplication of the left renal vein combined with a duplication of the portal vein, whereby the usual confluence between the splenic vein and the superior mesenterial vein was not found (see Figs. 1 and 2). In this patient the portal vein located on the left was formed by the confluence of splenic and

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A

B

Fig. 1. The portal vein (1) and the accessory portal vein (2) are presented uncoloured (A) and coloured turquoise (B). Further visible structures in A: common bile duct (A), gall bladder (B), right hepatic lobe (C), common hepatic artery (D).

inferior mesenteric veins (1). The other smaller one was the continuation of the superior mesenteric vein (2) with visible vascular branches draining the blood from pancreas and duodenum. All veins were located dorsally in relation to the pancreas and the duodenum. The superior mesenteric vein crossed the horizontal part of the duodenum in the usual way, together with the superior mesenteric artery. Four structures were

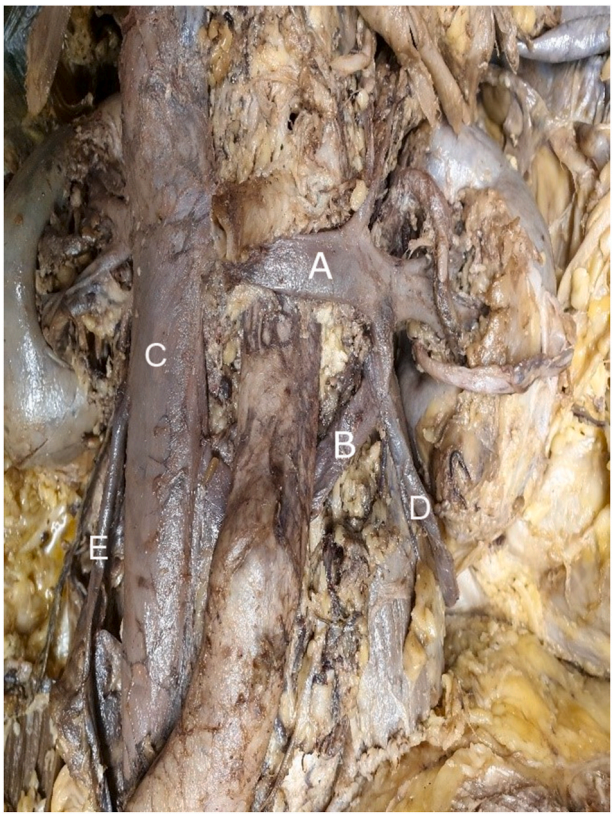

Fig. 2. Presentation of the anterior (pre-aortic) left renal vein (A) and the posterior (retro-aortic) left renal vein (B). Further visible structures: inferior vena cava (C), left testicular vein (D), right testicular vein (E). The human body which was used in this investigation came from the body donation programme of the Saarland University.

found in the hilus of the liver: a regularly-formed proper hepatic artery and a common bile duct as well as the two portal veins, whereby the continuation of the inferior mesenteric vein was located to the right of the continuation of the splenic vein, which had a larger lumen. Preparation of the blood vessels inside the liver parenchyma showed that both veins only had a short connection with each other. Both veins independently showed an intrahepatic separation into a left and right branch. While the portal vein 1 provided the blood for the I, II, III, and IVa, VII and VIII segment alone, the two portal veins shared segments VIb (left branch of portal vein II), V and VI (right branch of vena portae 2). Furthermore, it was observed that there was a comparatively large blood vessel containing blood from the gall bladder draining into the portal vein II.

Besides the normal renal vein located between the abdominal aorta and the superior mesenterial artery there was a second one located dorsal of the aorta and merging into the inferior vena cava caudal of the normal left renal vein. Furthermore, the left testicular vein drained its blood into the accessory left renal vein. No abnormality of the right renal vein was found.

3. Discussion

Both presented variations of the venous system were described previously. However, the combination of both variations has not been reported until now.

The duplication of the portal vein is a rare anatomic anomaly. However, multiple variations of the type of duplication are possible [2]. They differ in their location in relation to the duodenum (ventral or dorsal to this organ) and in their formation since the confluence of the three main veins that form the portal vein (Superior mesenteric vein,

splenic vein and inferior mesenteric vein) is variable [2,5]. Even a second entire hepatic system has been described [2]. In most cases there was no confluence of superior mesenteric vein and splenic vein, while the inferior mesenteric vein drained into either one of them, as in patients with only one regularly formed portal vein. It has been described that malformation can lead to portal hypertension through obstruction, but mostly these cases were incidental findings - as with our patient [3]. Remodelling of the vitelline veins during embryonic development has been presented as an explanation for the existence of two portal veins [4]. The liver and the portal venous system are formed between the fourth and twelfth week of pregnancy [1]. The portal venous system evolves from the vitelline veins. Three anastomoses are formed between these veins. The superior and inferior ones are located ventral to the intestinal tube, the medial one dorsal to it. In the course of normal development, parts of these embryonic structures, including major parts of the left vitelline vein, are obliterated, resulting in a single blood vessel - the portal vein. Thus, the main portal vein is formed by portions of the non-obliterating proximal part of the right vitelline vein, the middle/dorsal anastomosis and the distal non-obliterating part of the left vitelline vein. The superior anastomosis is incorporated into the developing liver parenchyma and is mainly responsible for the formation of the left portal vein. The superior mesenteric and splenic veins drain into the non-obliterating distal portion of the left vitelline vein caudal of the dorsal anastomosis.

Anastomoses play an important role in the correct positioning of the vein in relation to the intestinal organs. Thus, anomalies with deviant location of the veins in relation to the duodenum may be the result of malformation or obliteration of the anastomoses. This is, however, not so in the case of our patient as both portal veins were located dorsal to the duodenum. The duplication of the V. portae might, in this case, be the result of non-obliteration of parts of the left vitelline vein and separation from the anastomoses so that the V. lienalis evolved from the left vitelline vein and the V. mesenterica superior was formed by parts of the right vitelline vein and the medial and inferior anastomoses. However, this cannot be asserted with certainty.

The anterior nutcracker syndrome has been described by several authors [6,7]. Its symptoms are various and can range from microhaematuria to major flank pain. Similar symptoms can also be caused by the posterior nutcracker syndrome, in which the left renal vein is compressed between the abdominal aorta and the ventral body of the spinal vertebra [6]. However this anomaly can also be clinically silent. The existence of a second left renal vein can be explained through embryological development. Similar to the construction of the portal vein, primitive structures are formed and some of those obliterate again in the course of further development. During this procedure a renal collar is formed. Normally the intersupracardinal anastomosis (the one located dorsal to the primitive aorta abdominalis) obliterates so that only the ventral part, the intersubcardinal anastomosis persists [7]. In this case this obliteration probably did not take place so that both parts of the renal collar extended. Further malformations in this development are possible. Some may lead to a duplication of the left renal vein, whereby both veins are located dorsal to the abdominal aorta [7]. There is a classification in which six groups of congenital venous abnormalities are differentiated [8]. The reported body donor belongs to the third group which is the circumaortic left renal vein or venous collar. This anomaly (only cases with large left renal veins) is present in 1,5–8,7% of people [8]. This anomaly is often not diagnosed in patients due to the broad range of symptoms it causes or because patients experience no effects. Incidental findings are possible in MRI, CT or ultrasound. These methods of examination can help to detect the cause of pain experienced by patients with major symptoms. Furthermore, treatment will vary depending on the patient's symptoms. Observation is sufficient for some patients, others with severe pain and other symptoms might require open surgery [6]. The variations of renal vasculature is also of great importance for kidney transplantation [9]. The question of normality and variability and its significance in gross anatomy was nicely discussed recently [10].

4. Conclusion

The body donor presented here had two rather rare anomalies of two venous systems concurrently. It remains unknown whether this was merely a coincidence or if there might have been a common cause. This report is of interest for liver and kidney transplantation as well as other surgical interventions on these organs.

Ethical statement

The human body which was used in this investigation came from the body donation programme of the Saarland University.

Author contributions

All authors made the anatomical dissection and all authors wrote the manuscript.

Competing interests

The authors declare that they have no competing interests.

Funding

The study received no fundings.

Previous presentation

This case report was not presented before.

Consent

The body donor gave a written informed consent in the lifetime.

Declaration of competing interest

All authors declare that they have no conflicts of interest.

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References

[1] C. Carneiro, J. Brito, C. Bilreiro, M. Barros, C. Bahia, I. Santiago, F. Caseiro-Alves, All about portal vein: a pictorial display to anatomy, variants and physiopathology, Insights Imag. 10 (1) (2019) 38, https://doi.org/10.1186/s13244-019-0716-8.

[2] J.A. Corness, K. McHugh, D.J. Roebuck, A.M. Taylor, The portal vein in children: radiological review of congenital anomalies and acquired abnormalities, Pediatr. Radiol. 36 (2) (2006) 87–96, quiz 170-1.

[3] M. Dighe, S. Vaidya, Case report. Duplication of the portal vein: a rare congenital anomaly, Br. J. Radiol. 82 (974) (2009) e32–e34, https://doi.org/10.1259/bjr/81921288.

[4] G.E. Gondolesi, L. Bitetti, C. Rumbo, I. Sueiras, F.D. Lobos, G. Lieber, H. Oxilia, R. Villavicencio, A. Costaguta, P. Barros Schelotto, Pediatric living donor liver transplant in a recipient with biliary atresia and portal vein duplication-How did we manage it? Pediatr. Transplant. 23 (2) (2019), e13356 https://doi.org/10.1007/s00247-005-0010-4.

[5] R. Stauber, [A rare case of duplication of the portal vein], Zentralbl. Chir. 90 (34) (1965) 1896–1899.
[6] N. Skeik, P. Gloviczki, T.A. Macedo, Posterior nutcracker syndrome, Vasc. Endovasc. Surg. 45 (8) (2011) 749–755, https://doi.org/10.1177/1538574411419376.
[7] D.S. Kyung, J.H. Lee, D.Y. Shin, D.K. Kim, I.J. Choi, The double retro-aortic left renal vein, Anat. Cell Biol. 45 (4) (2012) 282–284, https://doi.org/10.5115/acb.2012.45.4.282.
[8] C.D. Karkos, I.A. Bruce, G.J. Thomson, M.E. Lambert, Retroaortic left renal vein and its implications in abdominal aortic surgery, Ann. Vasc. Surg. 15 (6) (2001) 703–708, https://doi.org/10.1007/s10016-001-0022-y.
[9] A. Gebremickael, M. Afework, H. Wondmagegn, M. Bekele, Renal vascular variations among kidney donors presented at the national kidney transplantation center, Addis Ababa, Ethiopia, Transl. Res. Anat. 25 (2001), 100145, https://doi.org/10.1016/j.tria.2021.100145.
[10] A. Żytkowski, R.S. Tubbs, J. Iwanaga, E. Clarke, M. Polguj, G. Wysiadecki, Anatomical normality and variability: historical perspective and methodological considerations, Transl. Res. Anat. 23 (2021), 100105, https://doi.org/10.1016/j.tria.2020.100105.