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Case Report

Congenital extrahepatic Portosystemic shunt with hypoplasia of the intrahepatic inferior vena cava: A rare case report ☆

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ABSTRACT

Congenital portosystemic shunt (CPS) is a rare vascular anomaly resulting in diversion of splanchnic or portal blood into the systemic circulation. Other vascular malformations associated with this entity are uncommon. A 4-year-old female child with a diagnosis of acute viral hepatitis had an incidental finding of extrahepatic CPS on a Doppler abdominal ultrasound. Contrast-enhanced computed tomography showed a dilated portal vein having H-type side-to-side communication with a hypoplastic intrahepatic portion of the inferior vena cava and a prominent dilated azygos vein. There was retroaortic left renal vein drained into the IVC which was visualized in its entirety. Echocardiography findings were normal and the patient was discharged after symptomatic treatment that achieved improvement. With the expansion of abdominal imaging, incidental cases of CPS are increasingly being diagnosed in children. Although vascular malformations associated with CPS are rare, early diagnosis of cases helps avoid complications during shunt closure.

Introduction

Congenital portosystemic shunts (CPS) are rare vascular malformations with a reported incidence of 1 in 30,000 to 1 in 50,000 live births which occur due to partial or complete diversion of portal blood into the systemic circulation [1–4]. A classification is made based on anatomy into intrahepatic (IH) and extrahepatic (EH) shunts, with physiologic, treatment, and prognostic differences existing between the 2 [3,5–7].

Abbreviations: CPS, Congenital Portosystemic Shunt; CECT, Contrast Enhanced Computed Tomography; MRI, Magnetic Resonance Imaging; EH, Extra Hepatic; IH, Intra Hepatic.

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EH shunts arise from the main portal vein (PV) while IH shunts originate from the portal venous branches [8]. CPS usually occur as isolated anomalies; however, they can be associated with cardiac, gastrointestinal, and other vascular malformations as well as genetic syndromes [1,4,9]. Vascular abnormalities associated with CPS are uncommon, and among those reported in the literature are splenic artery aneurysms, coronary artery fistulas, primitive hypoglossal artery, cutaneous and liver hemangiomas, azygos continuation of the inferior vena cava (IVC), and agenesis or double IVC [3,8,10].

Radiologic evaluation establishes the diagnosis and characterizes the CPS anatomy to guide treatment [3]. Doppler ultrasound (US) has been the primary imaging test, allowing estimation of the shunt and subsequent follow-up and treatment monitoring [8,11]. Cross-sectional imaging with contrast-enhanced CT (CECT) and magnetic resonance imaging (MRI) are used to delineate shunt anatomy and characterize potential focal liver lesions [1,3]. Angiography is recently a superior and gold-standard diagnostic technique for CPS [1].

Here, we present a case of an incidental diagnosis of EH CPS (type IIB) associated with other vascular malformations in a 4-year-old female patient who presented with a symptom constellation suggesting acute viral hepatitis. We have obtained informed consent of the patient's legal guardians to publish this case.

Case history

A 4-year-old female patient presented to our hospital - Tikur Anbessa Specialized Hospital, Addis Ababa, Ethiopia - with symptoms of fever, yellowing of the eyes, vomiting, and loss of appetite that had persisted for 2 weeks. She had no history of mental status change or dyspnea on exertion. She was investigated for the above complaints using laboratory tests and imaging modalities. The only pertinent laboratory findings were mild elevation of liver transaminases and direct hyperbilirubinemia; all other biochemical tests, including complete blood count, coagulation parameters, HBsAg, anti-HBc, and renal function tests were normal. With a clinical suspicion of acute viral hepatitis and obstructive jaundice, an abdominal US was ordered for further investigation, which showed a side-to-side shunt between a dilated main PV and the IVC (Fig. 1). The liver was of normal size and echogenicity, and no focal lesions were seen. The Doppler US examination showed a shunt with blood mixing at the communication site.

A CECT scan of the abdomen and pelvis was performed to better characterize the anatomy of the PV and IVC and to look for any concomitant abnormalities. It was remarkable for a fusiform dilation of the main PV having H-type side-to-side communication with the inferior vena cava. The right and left PVs had a reduced caliber, the intrahepatic portion of IVC was hypoplastic, and the 3 hepatic veins were seen separately (Fig. 2); otherwise, all segments of the IVC were clearly visualized. Additional vascular findings were also present including a tortuous retro-aortic left renal vein which drains to the IVC (not shown) and a prominent dilated azygous vein. Screening echocardiography was normal. With follow-up, the patient's symptoms subside and the laboratory parameters return to normal. A final diagnosis of EH CPS (type IIB) and resolved acute viral hepatitis was made based on interval clinical improvement and biochemical resolution . The patients' parents were informed of the diagnosis and the benefits of close follow-up. However, they disappeared from subsequent follow-ups.

Discussion

This case report presents an incidental diagnosis of CPS and concomitant anomalous systemic veins and a variant. A 4-year-old female child with a diagnosis of acute viral hepatitis had an incidental finding of extrahepatic CPS on a Doppler abdominal ultrasound. On contrast-enhanced computed tomography, there was a dilated portal vein with H-type side-to-side communication and a hypoplastic intrahepatic portion of the IVC. Retroaortic left renal vein was drained into the IVC as visualized in its entirety. Following symptomatic treatment that achieved improvement, the patient was discharged.

CPS are rare vascular malformations with uncertain prevalence and evolving classification and treatment schemes [2,4,5]. The pathogenesis involves aberrant development or involution of the fetal blood vessels, with different pathogenesis purported in IH and EH subtypes [2,4]. EH shunts are classified into type I CPS (Abernethy's malformation), involving an end-to-side shunt between the PV and IVC, with no intrahepatic portal flow and type II, side-to-side shunts, with some preservation of portal flow to the liver [3,12]. Type I malformation is divided into subtypes A and B: in type IA, the splenic vein and the superior mesenteric vein drain separately into the IVC while in type IB, these veins form a common trunk. Type II CPS is further classified based on the location of the shunt from the right or left PV (type IIA), the main PV (type IIB), or the mesenteric, splenic, or gastric veins (type IIC) [13].

Diagnosis is challenging given the rarity of this entity coupled with multisystem symptoms masquerading as other diseases, which leads to a significant delay in diagnosis [5]. On Doppler ultrasonography, the absence or no visibility of intrahepatic portal branches, as well as slow or absent portal flow and a compensatory dilatation of the hepatic artery, are suggestive findings [4]. Type IIB EH shunts may present with ectasia of the PV at the level of the shunt, as seen in our case [11]. CECT is typically performed next to further scrutinize the anatomy and location of the shunt [1]. Our patient had a dilated main PV connected to the IVC in a side-to-side fashion, and faintly visualized left and right PVs. Accordingly, the malformation we identified could be classified as type IIB CPS [13]. An MRI would have provided the same findings, although the information may be limited by motion artifacts in children [1,8]. Angiography has become an integral part of care performed either to detect a nonvisible PV and its intrahepatic branches or to close the shunt percutaneously [9]. When portal branches are not visible on the initial opacification of the shunt, IH portal system plasticity can be checked via a balloon occlusion of the shunt which often reveals distensible portal system [1]. Echocardiography is beneficial to identify associated cardiac abnormalities, reported to be present in one-third

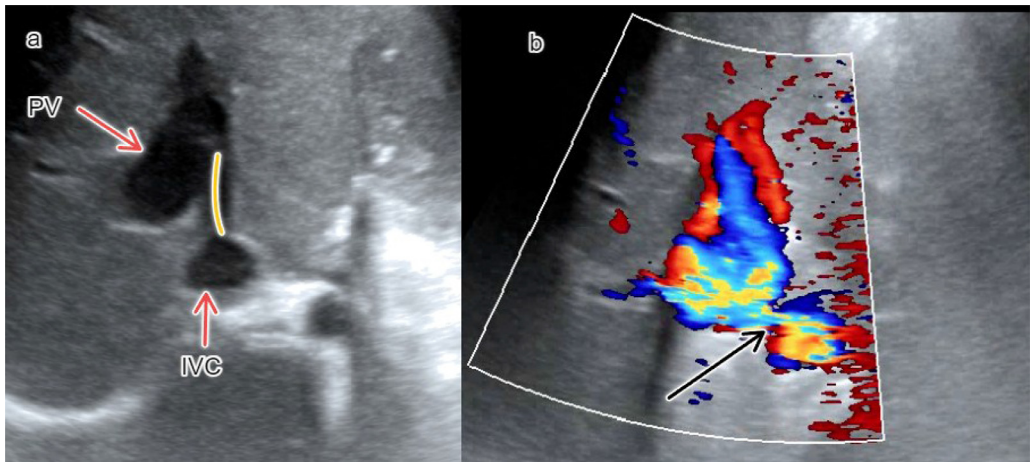

Fig. 1 – Type IIB CPS diagnosed in a 4-year-old girl. Gray-scale transverse US (A) demonstrates communication (path line) between a dilated PV and the IVC. Doppler US color image (B) demonstrates the vascular nature of the communication.

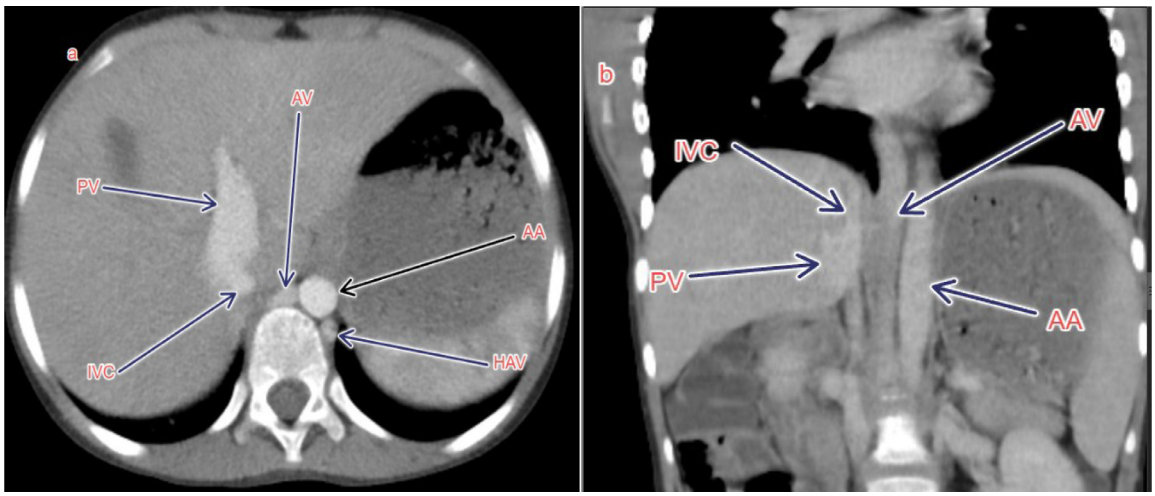

Fig. 2 – Axial (A) and Coronal (B) sections of a CT scan of the abdomen and pelvis done with IV contrast show a dilated trunk of the portal vein (PV) having a side-to-side communication with the inferior vena cava (IVC), which has a significantly reduced lumen caliber. The azygos vein (AV) is clearly prominent and dilated. AA: Abdominal aorta, AV: Azygos vein, HAV: Hemi azygos vein: IVC: Inferior vene cava, PV: Portal vein.

of cases, and screen for pulmonary hypertension before treatment [6–8].

Vascular malformations associated with CPS include splenic artery aneurysms, coronary artery fistulas, and cutaneous and hepatic hemangiomas [3,9]. Daniela et al. had recently reported EH CPS associated with arterial anomalies such as aberrant hepatic artery and supernumerary renal arteries [14]. In EH CPS associated with polysplenia syndrome, an azygos or hemiazygos continuation of the inferior vena cava can be seen [3]. In our patient, the intrahepatic portion of IVC was hypoplastic, yet all segments of the IVC were clearly present. The azygos vein was dilated which represents the presence of either systemic and/or splanchnic shunting towards it although none were identified. The left renal vein had a retroaortic and tortuous course which can be an incidental finding given its presence in 3% of the normal population [15]. Acknowledgment of all these anomalies is very important as there is a high risk of intraoperative catastrophic bleeding when encountering unknown vascular malformations [1,15].

Symptomatic CPS can present with cholestasis, mild elevation of liver enzymes, elevated serum ammonium levels, or rarely acute liver failure [1,4,7,16]. Although our patient had mild elevation of AST and ALT and direct hyperbilirubinemia, the acute onset of symptoms, the rapid recovery without treatment, and the absence of focal liver lesions argue against CPS being the culprit, although we cannot completely rule out this possibility [8].

Since shunts involving the EH PVs persist and carry risks of complications, it is suggested that these patients should receive treatment regardless of their symptoms. Early interven-

tion has been shown to preserve intellectual and psychosocial development and prevent permanent complications related to mortality such as pulmonary vascular diseases [6,7]. Thus, it is crucial to inform the patients and their caregivers of the possible long-term complications. However, controversy remains regarding the management of asymptomatic cases and the optimal follow-up for patients when closure is deferred.

Conclusion

We have found a case of CPS with hypoplasia of the intrahepatic portion of IVC and a prominent azygos vein possibly draining systemic and/or splanchnic collateral vessels, and a retroaortic left renal vein. With the expansion of abdominal imaging, incidental cases of CPS are increasingly being diagnosed in children. Associated vascular malformations should be sought carefully before invasive procedures to avoid catastrophic bleeding.

Authors' contributions

All authors contributed to the conduct of this research and read and approved the final version of the manuscript.

Ethics approval and consent to participate

Not applicable.

Patient consent

Written informed consent was obtained from the patient's parents for anonymized patient information to be published in this article.

Availability of data and materials

The data supporting the findings of the case are available upon request to the corresponding author.

REFERENCES

[1] Doi Y, Kim S, Ishigaki M, Sato K, Yoshimoto J, Mitsushita N, et al. Angiographic diagnosis for accurate assessment of congenital porto-systemic shunt and extrahepatic portal vein obstruction in children. Pediatr Int 2022;64(1):e14856.

[2] Carneiro DN, Rossi I, Oliveira NT, Oliveira LM, Rodrigues M, Araujo E, et al. Congenital intra-hepatic porto-systemic shunts diagnosed during intrauterine life: systematic review. J Clin Ultrasound 2022.

[3] Papamichail M, Pizanias M, Heaton N. Congenital portosystemic venous shunt. Eur J Pediatr 2018;177:285–94.

[4] Baiges A, Turon F, Simón-Talero M, Tasayco S, Bueno J, Zekrini K, et al. Congenital extrahepatic portosystemic shunts: abernethy malformation, in vascular disorders of the liver. In: VALDIG's guide to management and causes. Editors Cham. Springer International Publishing; 2022. p. 69–78.

[5] Korff S, Mostaguir K, Beghetti M, D'Antiga L, Debray D, Abella SF, et al. International registry of congenital porto-systemic shunts: a multi-centre, retrospective and prospective registry of neonates, children and adults with congenital porto-systemic shunts. Orphanet J Rare Dis 2022;17(1):1–9.

[6] McLin VA, Abella SF, Debray D, Guérin F, Beghetti M, Savale L, et al. Congenital portosystemic shunts: current diagnosis and management. J Pediatr Gastroenterol Nutr 2019;68(5):615–22.

[7] Chocarro G, Amesty MG, Encinas JL, Sánchez AV, Hernandez F, Andres AM, et al. Congenital portosystemic shunts: clinic heterogeneity requires an individual management of the patient. Eur J Pediatr Surg 2016;26:74–80.

[8] Kumar P, Bhatia M, Garg A, Jain S, Kumar K. Abernethy malformation: a comprehensive review. Diagn Interv Radiol 2022;28(1):21–8.

[9] Mohtashami A, Kiat A, Cross J, Simon R. Curtin A Catastrophic intraoperative bleeding due to congenital extrahepatic porto-systemic shunt anomaly: a surgical case report of two rare anomalies. Int J Surg Case Rep 2018;44:161–5.

[10] Vilgrain V, Paradis V, Wettere MV, Valla D, Ronot M, Rautou PE, et al. Benign and malignant hepatocellular lesions in patients with vascular liver diseases. Abdom Radiol 2018;43:1968–77.

[11] Bernard O, Abella SF, Branchereau S, Pariente D, Gauthier F, Jacquemin E, et al. Congenital portosystemic shunts in children: recognition, evaluation, and management. Seminars in liver disease. New York: Thieme Medical Publishers; 2012.

[12] Morgan G, Superina R. Congenital absence of the portal vein: two cases and a proposed classification system for portasystemic vascular anomalies. J Pediatr Surg 1994;29(9):1239–41.

[13] Lautz TB, Tantemsapya N, Rowell E, Riccardo A, Superina R. Management and classification of type II congenital portosystemic shunts. J Pediatr Surg 2011;46(2):308–14.

[14] Păcurar D, Dijmărescu I, Dijmărescu AD, Romaşcanu M, Adriana C, Becheanu CA, et al. A case report on an incidental discovery of congenital portosystemic shunt. Medicine 2019;98(31):e16679.

[15] Kim S, Hyeong, Shin C, Hwang J, Jou S, Lee W, Choi S, et al. Various congenital anomalies of the inferior vena cava: review of cross-sectional imaging findings and report of a new variant. Abdom Radiol 2018;43:2130–49.

[16] Abella SF, Lambert SBV, Fabre M, Steimberg C, Losay J, et al. Complications of congenital portosystemic shunts in children: therapeutic options and outcomes. J Pediatr Gastroenterol Nutr 2010;51(3):322–30.