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Congenital Absence of Portal Vein Combined with Retro-aortic Left Renal Vein: A Case Report

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Case Report

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ABSTRACT

In the past, Congenital Absence of Portal Vein (CAPV) was a rare occurrence. CAPV transports blood from the gastrointestinal system, gallbladder, pancreas and spleen to the liver. This blood is derived from digested substances and contains nutrients and poisons. The portal vein accounts for about 75% of all liver blood flow with the remaining 15% coming from the hepatic artery itself. In the hepatic veins, blood travels from the liver to the heart. A rare defect known as Congenital Absence of The Portal Vein (CAPV) causes the intestinal and splenic venous drainage leak into the systemic veins by leaving liver. However, because of advancements in imaging techniques, the number of CAPV cases detected has increased in recent years. Patients with CAPV may develop Portal Hypertension (PH) or Portosystemic Encephalopathy (PSE). However these symptoms do not usually appear until the patients are older. The cause of this disease is unknown, however most researchers believe it is linked to aberrant embryologic development of the portal vein. The left renal vein passes anterior to the aorta just below the origin of the superior mesenteric artery. A multislice CT scan revealed a variation in the portal vein and retro-aortic left renal vein. Variations in the left renal vein are critical when planning retroperitoneal surgery and vascular procedures. Abdominal procedures, transplantations and preoperative evaluation of endovascular interventions all require knowledge of a patient's portal vein and renovascular anatomy as well as detecting their variations and anomalies. We are reporting a rare case of congenital absence of portal vein along with retro-aortic left renal vein in an adult patient.

INTRODUCTION

John Abernethy reported the first case of Congenital Absence of The Portal Vein (CAPV) in 1793, Patients typically consult doctors for venous shunt problems, hepatic or cardiac abnormalities identified by Computed Tomography (CT), Magnetic Resonance Imaging (MRI) or ultrasound (MRI). Although the exact cause of this condition is unknown. Based on a postmortem examination of a 10-month-old girl that revealed the portal vein's termination in the inferior vena cava at the insertion level of the renal veins [1]. Complete portosystemic shunts that do not perfuse the liver *via* the portal vein are classified as type I while partial shunts with some portal perfusion to the liver are classified as type II [2]. The complexities of liver surgery and advances in operation techniques involving the liver have made variations in this region more important [3]. Just below the beginning of the superior mesenteric artery, the left renal vein flows anterior to the aorta. The left renal vein anomalies of a patient are significant to know, when considering retroperitoneal surgery and vascular treatments as well as when making a differential diagnosis of retroperitoneal lymphadenopathies [4]. It showed numerous congenital anomalies in addition to CAPV including the termination of the portal vein in the inferior vena cava near the insertion level of the renal veins. According to a report, type I portosystemic shunts are those that completely cut off the liver's access to the portal vein while type II shunts still allow some portal vein blood to flow to the liver. Type I is further divided into types Ia and Ib according to the morphology of the portal vein. The Superior Mesenteric Vein (SMV) and Splenic Vein (SV) drain separately in type Ia but they both do so in type Ib after joining to create a single trunk.

CASE PRESENTATION

A 58 year old male presented to surgery outpatient with history of abdominal bloating for 2 months. Clinical examination revealed no abnormality. Upper gastrointestinal endoscopy revealed grade 1 esophageal varices (Figure 1) with portal gastropathy (Figure 2). CT scan showed cavernous transformation of portal vein with no evidence of portal vein thrombosis (Figure 3). Main portal vein not visualized from its confluence. Scan also revealed retro aortic left renal vein (Figure 4) with normal renal parenchyma. Patient was conservatively managed and is closely followed up.

Figure 1. Endoscopy showing grade 1 esophageal varies.

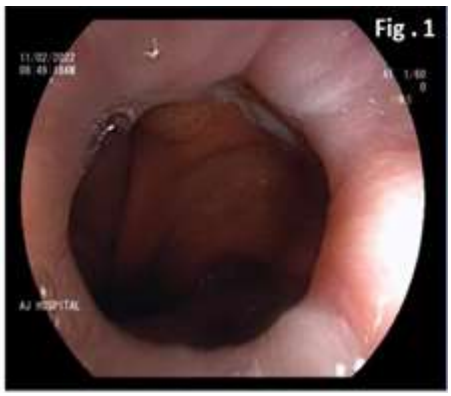


Figure 2. View of Portal gastropathy.

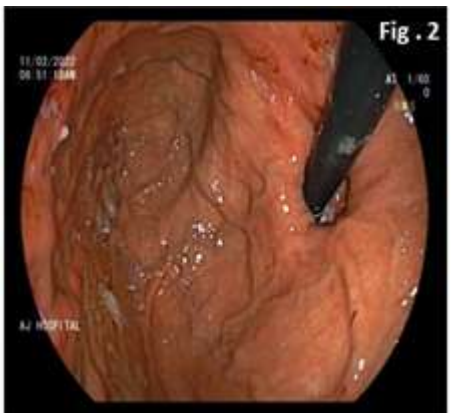


Figure 3. Axial CT Image showing non visualization of main portal vein.

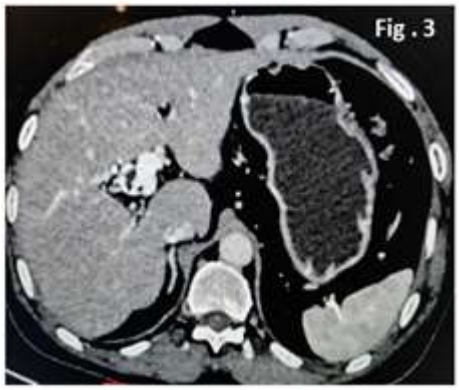
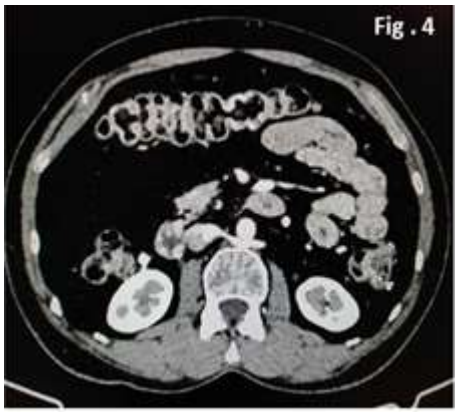


Figure 4. CT showing retro-aortic left renal vein.



RESULTS AND DISCUSSION

The two vitelline veins (including the intervitelline anastomosis) and the left umbilical vein give rise to the definitive portal vein. The interconnecting vitelline veins form the portal vein trunk at the culmination of a complex process that occurs between the third and sixth weeks of embryological development [5]. The Abernethy malformation, according to Howard and Davenport, is a congenital diversion of portal blood away from the liver *via* an end-to-side or side-to-side shunt [6]. As a result, CAPV with extrahepatic portocaval shunts is known as an Abernethy type I malformation. Patients with CAPV may develop Portal Hypertension (PH) or Portosystemic Encephalopathy (PSE). However these symptoms do not usually appear until the patients are older. The cause of this disease is unknown,

however most researchers believe it is linked to aberrant embryologic development of the portal vein. While SV and SMV return blood from the portal vein, the portal vein returns blood from the intraperitoneal portion of the gastrointestinal tract as well as the spleen, pancreas and biliary system. The portal vein splits into right and left branches at the portal hepatis (besides providing the quadrate lobe with an additional branch). Retro-aortic left renal vein is an atypical form of nutcracker syndrome, presenting with hematuria, flank pain or varicocele. They are divided into four types depending upon their drainage. A left retro-aortic renal vein may also be seen with other complex venous anomalies and may cause primary Budd-Chiari Syndrome [7].

CONCLUSION

In conclusion, the location of portosystemic shunts, congenital heart disease, and liver illness all affect the prognosis of CAPV patients. Patients with CAPV who have no additional anomalies have a varied outcome. For CAPV patients, a long-term follow-up with lab testing and imaging screening is advised. The small number of papers reporting the absence of the other complex portohepatic venous system, the rare occurrence of retro-aortic renal vein and the clinical symptoms that may accompany these venous anomalies emphasize the originality and importance of our case. We believe that understanding the variations in this area is critical for accurate diagnosis and therapy. The very rare variation presented here with congenital absence of portal vein and asymptomatic retro-aortic left renal vein may also contribute to the effectiveness of surgical interventions.

DISCLOSURES

Conflict of interest statement

The authors have no conflicts of interest to declare.

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