Case 17173

Eurorad

A middle-aged woman with a rare abdominal vascular malformation

Published on 22.03.2021

DOI: 10.35100/eurorad/case.17173
ISSN: 1563-4086
Section: Abdominal imaging
Area of Interest: Abdomen Mesentery Veins / Vena cava
Procedure: Shunts
Imaging Technique: CT
Special Focus: Congenital Case Type: Clinical Cases
Authors: Frederik Bosmans1,2, Ivan Pilate2, Filip M. Vanhoenacker1,2,3
Patient: 66 years, female

Clinical History:

A 66-year-old female presented to our hospital with a history of an unstable gait and increasing fatigue. Over the last few days, her symptoms had worsened and she was admitted for further work-up. On clinical examination asterixis was noted and laboratory findings showed high serum ammonia levels.

Imaging Findings:

3D virtual reconstructed image of a contrast-enhanced CT abdomen demonstrates multiple tortuous and dilated mesenteric veins in the upper abdomen (Fig. 1a). Additionally, a crossed fused renal ectopia is noted. This finding is better depicted on the parasagittal reformatted image (Fig. 1b). Coronal reformatted CT shows an engorged inferior vena cava (Fig. 2). Dilated splenic and mesenteric veins can be seen draining into the inferior vena cava. Axial reformatted CT through the porta hepatis reveals a complete absence of the portal vein (Fig. 3).

Discussion:

Background

Abernethy malformations are uncommon congenital extrahepatic portosystemic shunts (CEPS) redirecting the mesenteric venous blood away from the liver (Fig. 4). Abernethy malformations are classified into two types. In type I malformations, there is a complete absence of the portal vein. The splanchnic circulation drains into the inferior vena cava, renal or iliac veins. Type I is further divided into subtypes Ia and Ib. In subtype Ia, both the superior mesenteric and splenic vein drain separately into the systemic circulation (Fig. 4b). In subtype Ib, the superior mesenteric and splenic vein join together to form a portal vein equivalent lacking any intrahepatic branches directly draining into the systemic circulation (Fig. 4c). Type I malformations are predominantly found in females.
Type II malformations are partial portosystemic shunts with a remaining normal or hypoplastic portal vein and an extrahepatic end-to-end shunt between the portal vein and the systemic circulation (Fig. 4c) [1].

Clinical perspective

CEPS are often diagnosed as an incidental finding but can be symptomatic. Because portosystemic shunts bypass the hepatic circulation, normal metabolism of visceral venous return is disrupted. Accumulation of metabolites such as galactose or ammonia in the serum can lead to varying clinical symptoms. Galactosemia can lead to cataracts and hyperammonemia may be present with or without encephalopathy. Encephalopathy develops more often in longstanding cases [2]. CEPS are frequently associated with other congenital defects, particularly cardiovascular abnormalities. Other manifestations are hepatopulmonary syndrome, pulmonary arterial hypertension and an increased incidence of liver masses such as focal nodular hyperplasia, hepatic adenoma or hepatocellular

Imaging

US is the initial imaging modality of choice as it is fast, non-invasive and does not expose the patient to ionizing radiation. Although US may depict an absent portal vein, it may be insufficient to demonstrate the associated extrahepatic shunts. CT or MRI are the preferred imaging modalities to reliably confirm the portosystemic shunts and reveal any additional malformations [5].
Cavernous transformation of the portal vein (CTPV) is an important differential diagnosis. Long-standing thrombosis of the portal vein gives rise to numerous serpiginous venous collaterals. CTPV is frequently associated with signs of portal hypertension. Patients with CEPS usually do not have imaging features of portal hypertension such as ascites, varices, or splenomegaly. Another differential diagnosis is surgically created portosystemic shunts, in these cases, a thorough history is essential for the correct diagnosis [6].

Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Congenital extrahepatic portosystemic shunt , Portal cavernoma, Inferior vena cava aneurysm, Persistent ductus venosus, Cirrhotic liver with portal hypertension

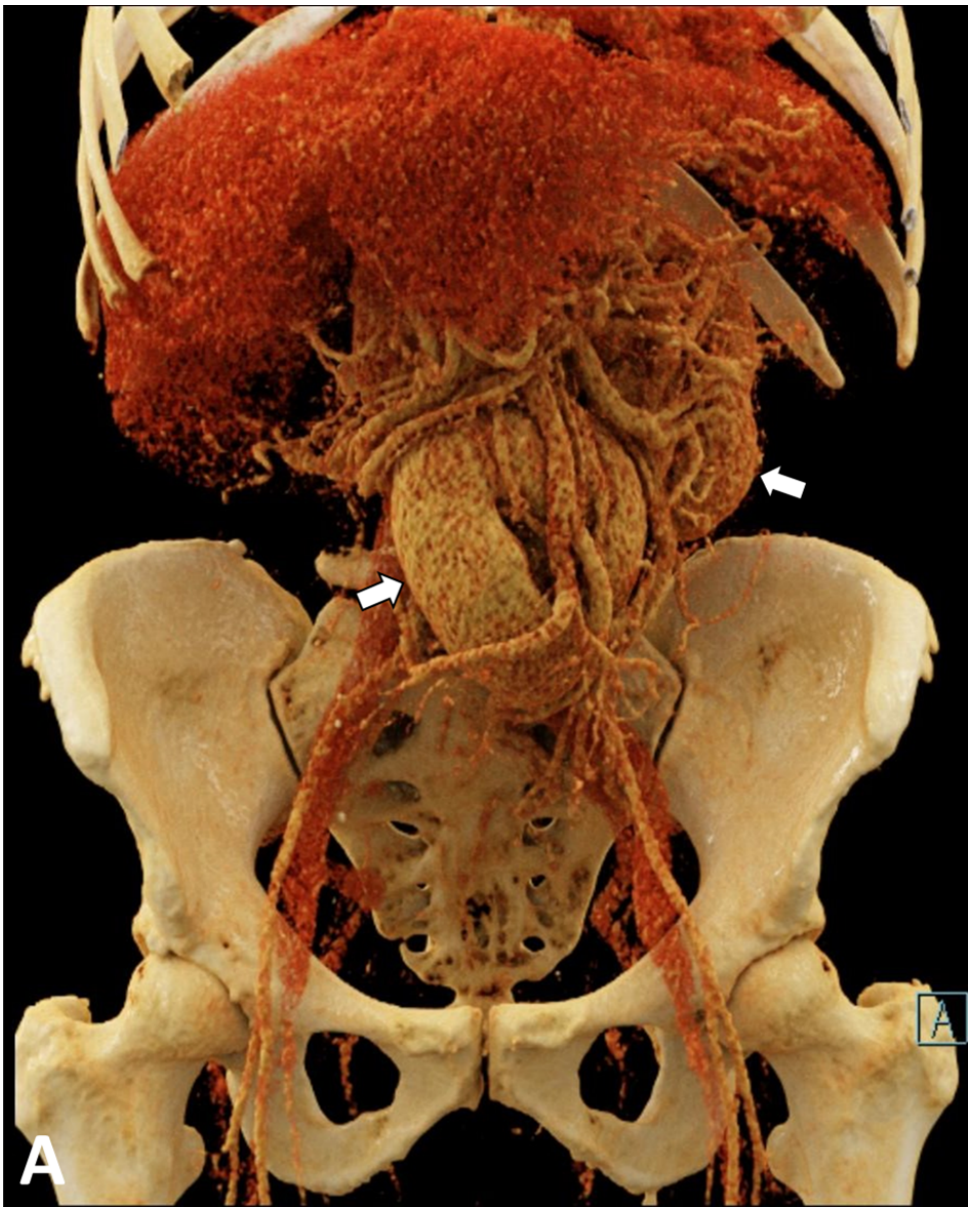
Final Diagnosis: Congenital extrahepatic portosystemic shunt

References:

Kwapisz L, Wells MM, AlJudaibi B (2014). Abernethy malformation: congenital absence of the portal vein. Can J Gastroenterol Hepatol. 28(11):587-8. (PMID: 25575105)
Berry GT, Segal S, Gitzelmann R (2006). Disorders of Galactose Metabolism. In: Fernandes J, Saudubray J-M, van den Berghe G, Walter JH, editors. Inborn Metabolic Diseases: Diagnosis and Treatment. Berlin, Heidelberg: Springer Berlin Heidelberg. p. 121-30.
Alonso-Gamarra E, Parrón M, Pérez A, Prieto C, Hierro L, López-Santamaría M (2011). Clinical and Radiologic Manifestations of Congenital Extrahepatic Portosystemic Shunts: A Comprehensive Review. RadioGraphics. 31(3):707-22. (PMID: 21571652)
De Vito C, Tyraskis A, Davenport M, Thompson R, Heaton N, Quaglia A (2019). Histopathology of livers in patients with congenital portosystemic shunts (Abernethy malformation): a case series of 22 patients. Virchows Archiv : an international journal of pathology. 474(1):47-57. (PMID: 30357455)
Gallego C, Miralles M, Marín C, Muyor P, González G, García-Hidalgo E (2004). Congenital Hepatic Shunts. RadioGraphics. 24(3):755-72. (PMID: 15143226)
Murray CP, Yoo SJ, Babyn PS (2003). Congenital extrahepatic portosystemic shunts. Pediatric radiology. 33(9):614-20. (PMID: 12879313)

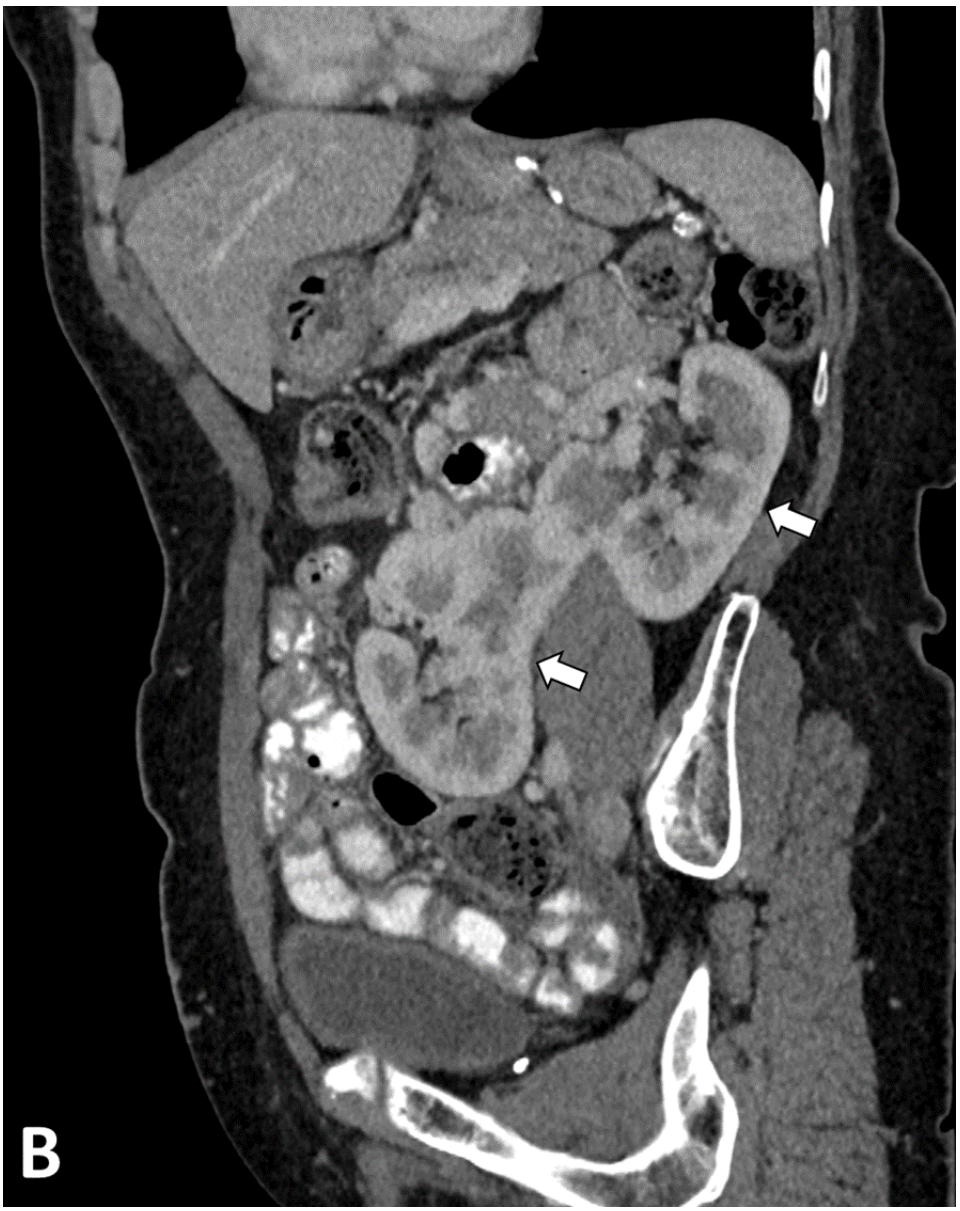
Figure 1

a



Description: 3D virtual reconstructed image of a CECT abdomen demonstrates multiple tortuous and dilated mesenteric veins in the upper abdomen. Note the presence of a crossed fused renal ectopia (arrows). **Origin:** © Department of Radiology, Algemeen Ziekenhuis Sint-Maarten, Mechelen, Belgium, 2020

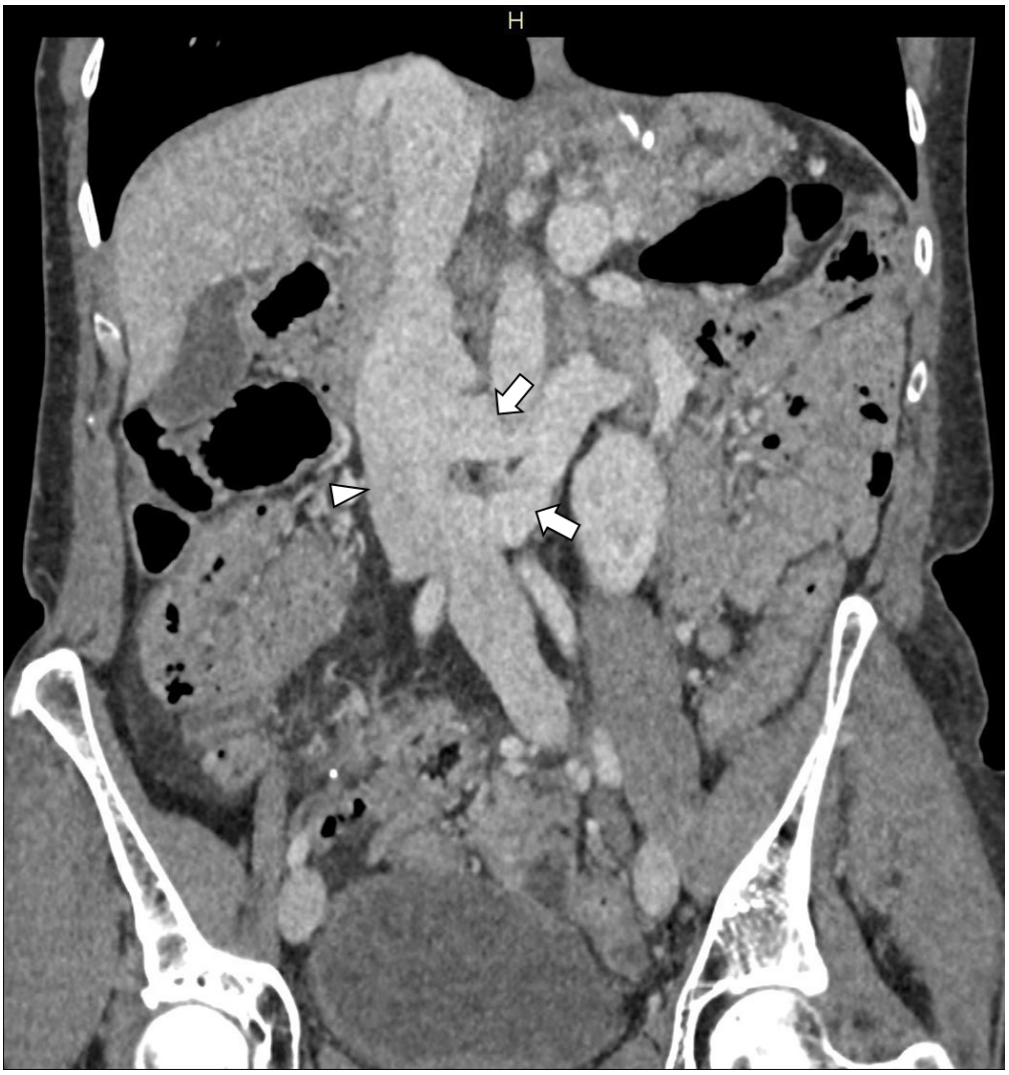
b



Description: Parasagittal reformatted CECT of the abdomen better depicts the crossed fused renal ectopia (arrows). **Origin:** © Department of Radiology, Algemeen Ziekenhuis Sint-Maarten, Mechelen, Belgium, 2020

Figure 2

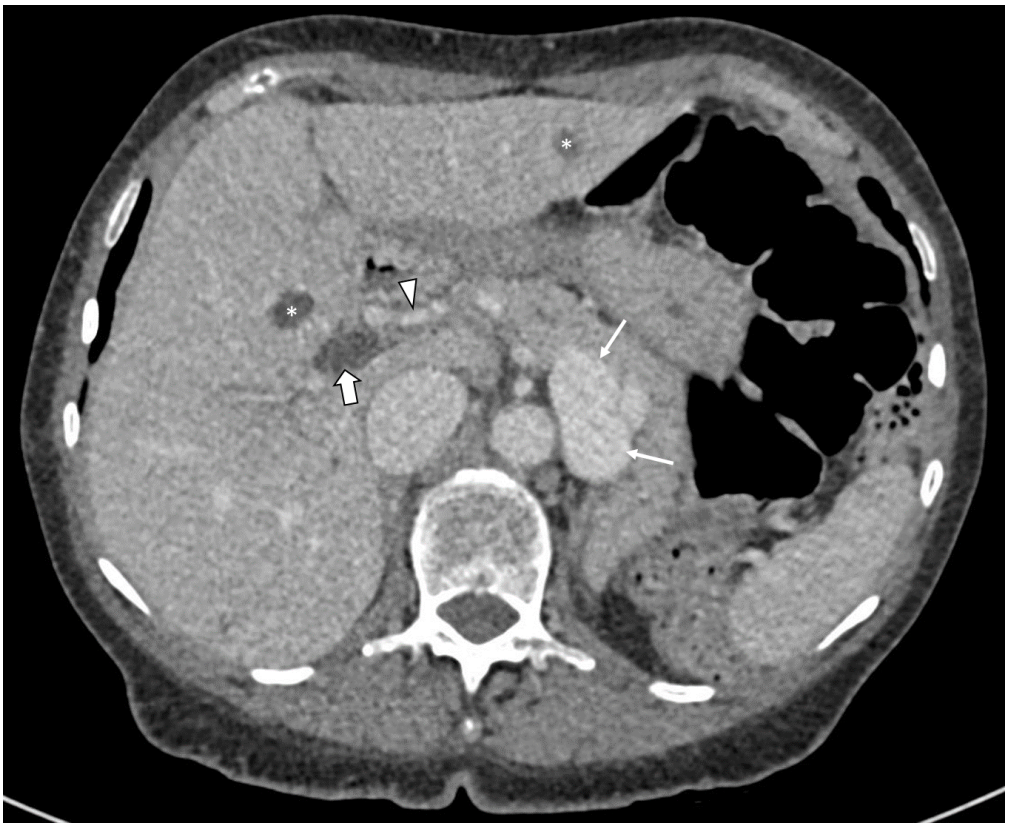
a



Description: Coronal reformatted CECT of the abdomen shows an engorged inferior vena cava (arrowhead). The dilated splenic and mesenteric veins can be seen draining into the vena cava (arrows). **Origin:** © Department of Radiology, Algemeen Ziekenhuis Sint-Maarten, Mechelen, Belgium, 2020

Figure 3

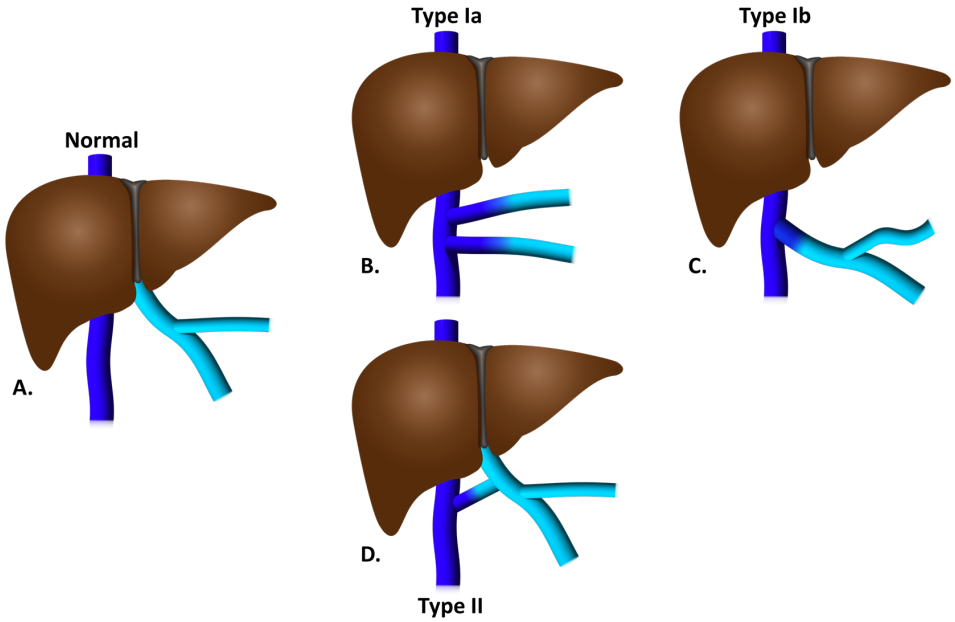
a



Description: Axial reformatted CECT through the porta hepatis reveals complete absence of the portal vein. There is dilatation of the common hepatic duct (arrow) and a prominence of the proper hepatic artery (arrowhead). Note the enlarged and tortuous splenic vein (thin arrows) and multiple biliary cysts (asterisk). **Origin:** © Department of Radiology, Algemeen Ziekenhuis Sint-Maarten, Mechelen, Belgium, 2020

Figure 4

a



Description: Types of Abernethy malformations. (A) Normal anatomy. (B) Type Ia: the splenic vein and superior mesenteric vein drain individually into inferior vena cava. (C) Type Ib: the splenic vein and superior mesenteric vein join before draining into the inferior vena cava. (D) Type II: normal or hypoplastic portal vein with an end-to-end shunt between the portal vein and inferior vena cava. **Origin:** © Frederik Bosmans, MD.