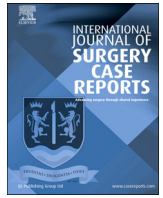




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Catastrophic intraoperative bleeding due to congenital extrahepatic porto-systemic shunt anomaly: A surgical case report of two rare anomalies

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ABSTRACT

INTRODUCTION: Abernethy malformations are extremely rare congenital anomalous portosystemic shunts. We report the case of a patient with a rare variant Abernethy malformation between the superior mesenteric vein and left renal vein, associated with a massive jejunal diverticulum.

PRESENTATION OF CASE: A 37-year-old Caucasian female presented to our emergency department with severe abdominal pain and proceeded to laparotomy for a presumed small bowel obstruction. At laparotomy she was found to have a massive diverticulum at the duodeno-jejunal junction, which was intimately associated with a venous malformation and the anomalous portosystemic shunt. Whilst mobilising the diverticulum, the patient developed catastrophic haemorrhage from the malformation. The patient underwent a complicated post-operative course however was eventually stabilised.

DISCUSSION: We discuss the anatomy and pathophysiology of anomalous portosystemic shunts and propose an embryological origin for our patients' anomalies.

CONCLUSION: Abernethy malformations are rare however may be associated with other intra-abdominal pathology and extreme caution is required when operating on these patients.

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1. Introduction

Congenital Extrahepatic Porto-Systemic Shunts (CEPS) were first described by Abernethy in 1793 discovered at autopsy of a female infant. The majority of reported cases have been discovered in childhood with a greater prevalence in females [1], although they can seemingly present in any age group.

Morgan and Superina [2] categorised CEPS into two anatomical classes based on the absence or persistence of hepatic portal venous flow. Type 1 is a complete shunt, with no portal venous blood able to enter the hepatic circulation. Type 2 is a partial shunt, in which an accessory venous pathway from the portal to the systemic venous system is present in addition to the portal vein. In Type 2 anomalies the portal vein is often hypoplastic. Kobayashi et al. [2] has further classified CEPS by the destination of their outflow, with the most common outflow being into the inferior vena cava (IVC) and designated Type A, being distinguished from those with outflow into the renal veins (Type B) and the iliac veins (Type C) (Fig. 1).

CEPS may be asymptomatic or may lead to a wide range of sequelae. They may be an isolated pathology but are more often associated with other anatomical anomalies [3,4].

We report the case of a patient with a rare Abernethy malformation between the superior mesenteric vein (SMV) and left renal vein (LRV) associated with a massive jejunal diverticulum.

Our case report work has been reported in line with the SCARE criteria [5].

2. Presentation of case

A 37-year-old woman presented to the emergency department with 12 h of severe epigastric abdominal pain. She had a background of un-investigated recurrent episodes of intermittent severe abdominal pain and had undergone a laparotomy as a 1-day old baby for an "abdominal cyst", the details of which were not available.

On examination, she was very distressed, she had a low-grade fever (38 °C), generalised abdominal tenderness with guarding in the epigastrium, and no features of chronic liver disease. She had a mild leucocytosis, and other blood tests including liver function tests were within normal limits.

A computed tomography (CT) of the abdomen and pelvis revealed what was presumed to be a dilated small bowel loop in the upper abdomen, 10 cm in diameter and containing faeculent mate-

Abbreviations: CEPS, anomalous extrahepatic porto-systemic shunt(s); CT, computed tomography; SMV, superior mesenteric vein; LRV, left renal vein; DJ, duodeno-jejunal.

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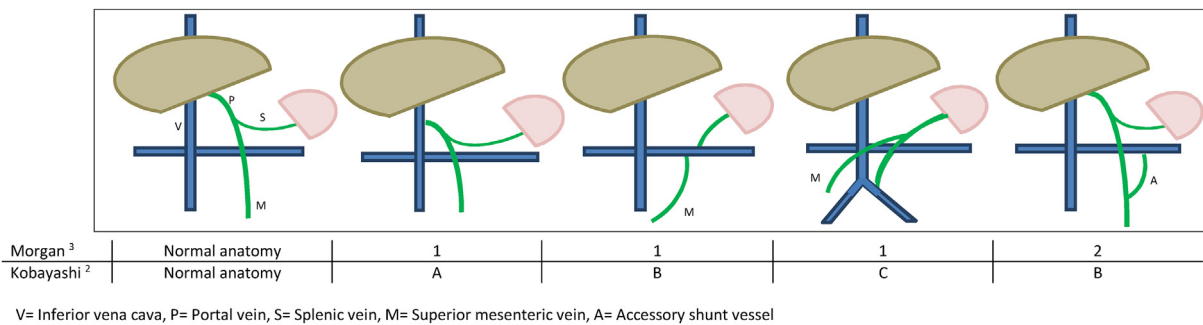


Fig. 1. Schematic examples of Abernethy malformation under the classification systems of Morgan et al. and Kobayashi et al.

Morgan ³	Normal anatomy	1	1	1	2
Kobayashi ²	Normal anatomy	A	B	C	B

V= Inferior vena cava, P= Portal vein, S= Splenic vein, M= Superior mesenteric vein, A= Accessory shunt vessel.

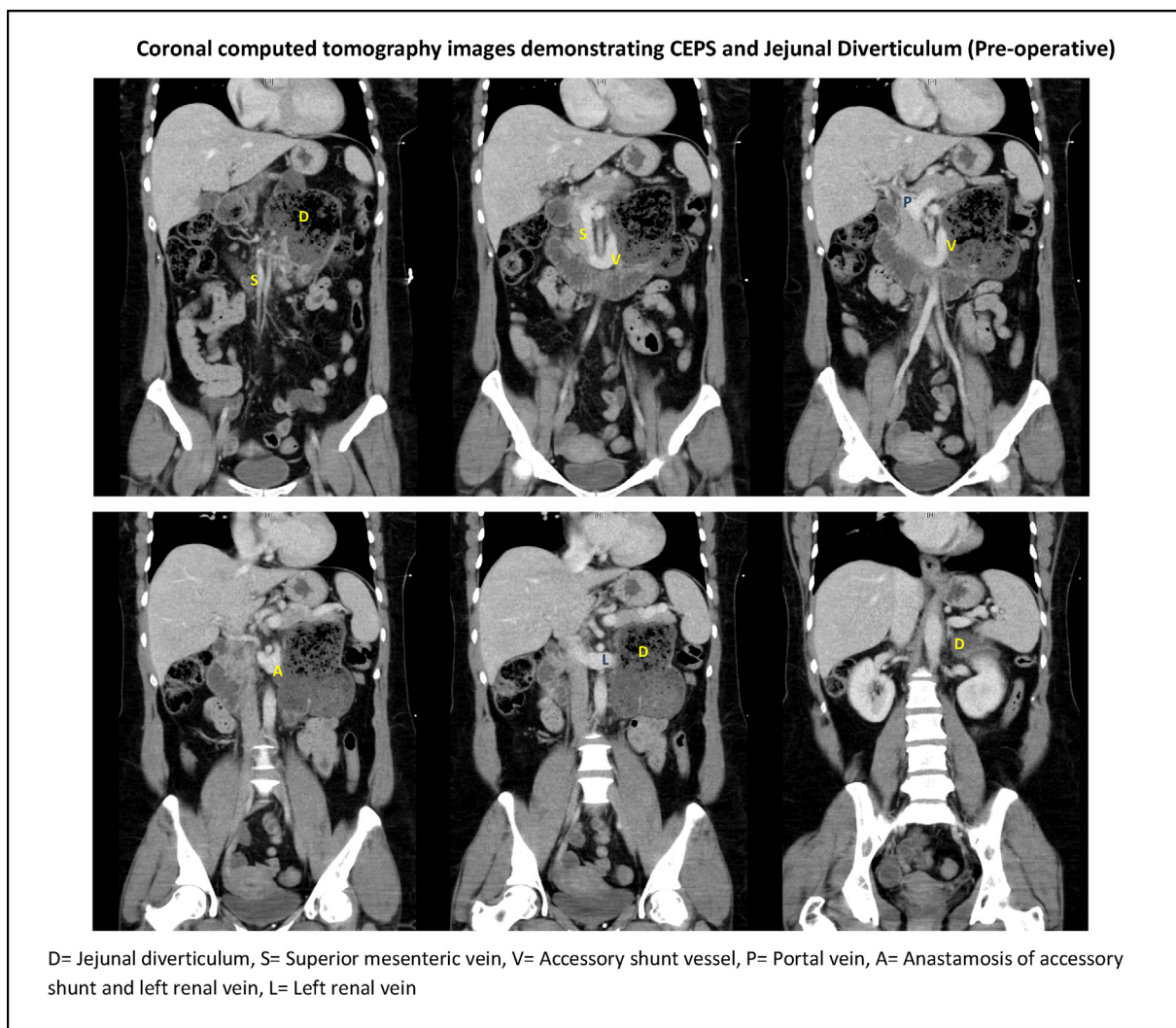


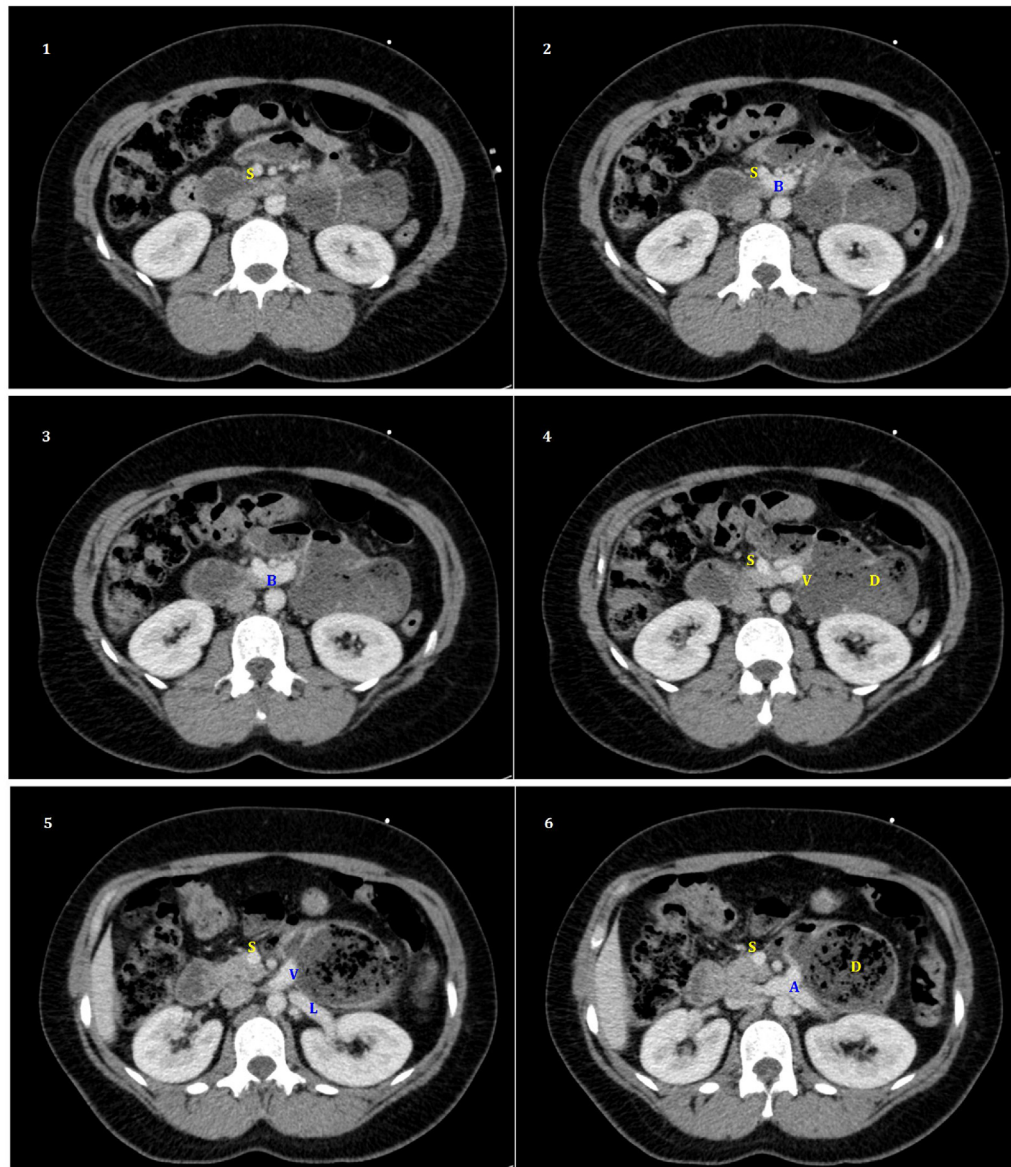
Fig. 2. Coronal computed tomography images demonstrating CEPS and Jejunal Diverticulum (Pre-operative). D=Jejunum diverticulum, S=Superior mesenteric vein, V= Accessory shunt vessel, P= Portal vein, A= Anastomosis of accessory shunt and left renal vein, L= Left renal vein

rial. Considered differentials included a closed loop small bowel obstruction or abnormally located caecal volvulus. Furthermore, an abnormal vein was noted joining the superior mesenteric vein (SMV) to the left renal vein (LRV). This large vein originated as a side branch of the SMV at spinal level L2 and made a small inferior loop before ascending to spinal level L1 where it turned posteriorly

to join the anterior aspect of the LRV in an end-to-side anastomosis (Figs. 2 and 3).

Despite large doses of opioid analgesia, the patient’s pain remained severe and it was decided to proceed to laparotomy. At laparotomy adhesive bands due to previous laparotomy were found and single massive thin walled diverticulum was found at

Axial computed tomography images demonstrating CEPS and Jejunal Diverticulum Ordered from inferior to superior



S= Superior mesenteric vein, B= “Branching” of SMV into accessory shunt vessel and superior mesenteric vein proper, V= Accessory shunt vessel, D= Jejunal diverticulum, L= Left renal vein, A= Anastomosis of accessory shunt and left renal vein

Fig. 3. Axial computed tomography images demonstrating CEPS and Jejunal Diverticulum Ordered from inferior to superior. S= Superior mesenteric vein, B= “Branching” of SMV into accessory shunt vessel and superior mesenteric vein proper, V= Accessory shunt vessel, D= Jejunal diverticulum, L= Left renal vein, A= Anastomosis of accessory shunt and left renal vein

the duodeno-jejunal (DJ) flexure extending up towards the splenic hilum; a nest of small veins was noted at the superior posterior aspect.

Diverticulum was the considered the cause of patient’s sign and symptoms that intra-operatively we detected.

Mobilisation of the diverticulum was problematic as it was difficult to establish a plane posteriorly. During this attempted mobilisation, we encountered sudden massive venous haemorrhage.

The haemorrhage was not controlled by standard haemostatic measures and it was unclear as to its exact source. A splenic tear was initially suspected and a splenectomy was undertaken. However, when this failed to control the haemorrhage, a medial visceral rotation was performed. Catastrophic bleeding was identified from the anomalous vein connecting the SMV with the LRV and from a nest of veins posterior to the diverticulum. These abnormal veins were extremely thin walled and friable and tore when oversewn. Eventually the haemorrhage was controlled but the

patient required massive transfusion, with intra-operative blood loss estimated at 7 L in total. The involved jejunum was resected and sent for histopathological examination which demonstrated chronic inflammatory changes with focal areas of necrosis.

3. Outcome and follow-up

Post operatively the patient was transferred to ICU and a relook laparotomy was planned for the following day. During re-look laparotomy she was found to have necrotic sections of the caecum, ascending, and transverse colon necessitating an extended right hemicolectomy. The patient underwent a protracted post-operative recovery requiring prolonged cardiorespiratory support and complicated by hypoxic hepatic, renal, pancreatic and brain injury.

She was discharged with full neurological recovery six months after her initial presentation.

4. Discussion

This case demonstrates a previously unreported association of CEPS with a massive jejunal diverticulum. It also highlights the potential for catastrophic bleeding when encountering such anomalous vascular malformations intra-operatively.

CEPS are extremely rare and less than 200 cases have been reported worldwide [1]. Lewis et al. reviewed the mesenteric vasculature of 323,222 imaging reports of non-cirrhotic patients and discovered only 4 cases of CEPS [6] (of which only one was Type 2). The anomaly may cause a range of complications, with presentations including liver failure, hyper-ammonaemia and hepatic encephalopathy, hepatocellular tumours, gastrointestinal bleeding, hepato-pulmonary syndrome and pulmonary hypertension [1,3,4]. More rarely, as in our case, they may be asymptomatic [1]. They are often associated with other congenital abnormalities such as biliary atresia, polysplenia, and gastrointestinal, genitourinary, vertebral and cardiac anomalies [1,3,4,7].

CEPS are thought to result from the persistence of embryonic vessels. In the embryo by the fifth week three main pairs of veins can be distinguished, the vitelline veins (carrying blood from the yolk sac to the sinus venosus), the umbilical veins (carrying oxygenated blood to the embryo) and the cardinal veins (draining the body of the embryo proper). Derivatives of the vitelline veins are the terminal part of the IVC, the hepatic veins and the portal vein. The paired cardinal veins initially subdivide creating the subcardinal veins draining the kidneys, before anastomosing to create the left renal vein; the right subcardinal vein develops into the renal segment of the IVC, which ultimately joins with the vitelline derived hepatic segment to create the complete IVC.

Congenital absence of the portal vein may be attributed to excessive involution of the *peri*-intestinal vitelline venous loop or to total failure of the vitelline veins to establish the critical anastomosis with the hepatic sinusoids or umbilical veins. Khoda et al. [8] propose that extrahepatic portosystemic shunts originate with the persistence of subcardino-hepatic anastomosis with the vitelline veins. The cause of these embryonic anomalies is poorly understood. Whilst a genetic basis has been associated with some vascular malformations, no current link has been established with human CEPS. However, CEPS are more prevalent in certain breeds of canine, suggesting an inherited predisposition.

Our case demonstrates an uncommon variant of a Type 2 communication between the SMV and the LRV with a normal sized portal vein. In most Type 2 CEPS the connection appears to lie between the portal vein and hepatic portion of the IVC, both originating from the vitelline vein. Our case was unusual in that the

shunt connected the SMV (vitelline origin) to the LRV (cardinal origin). The LRV was noted to have normal orientation.

Our case also demonstrated a massive diverticulum on the anti-mesenteric border at the DJ flexure. This was intimately related to the portosystemic shunt and surrounded by a “nest” of small veins. Single large proximal jejunal diverticula are rare, particularly in young patients. Although vascular malformations within a jejunal diverticulum have previously been reported [9], it seems unlikely that our patient would have two very rare unrelated pathologies in the same vicinity. Further, jejunal diverticula are usually associated with increased intraluminal pressure causing herniation of mucosa and submucosa through the muscularis mucosa, while the histology in our case showed normal bowel wall indicating that this may be a congenital diverticulum.

We hypothesise that this diverticulum is related to an index embryological event causing a local field change and developmental arrest allowing persistence of embryonic vessels and abnormal duodeno-jejunal development. In the foetus, the tributaries of the vitelline veins initially form a plexus, encircling the primitive duodenum. The index event may have arrested part of this development leading to a persistent venous plexus around the DJ flexure.

Furthermore, the possibility of a duodeno-jejunal duplication could also be considered for the pathogenesis of the diverticulum. Gastrointestinal duplications are rare congenital malformations and duodenal duplications account for up to 5% of gastrointestinal duplications. In the embryo, the alimentary canal goes through a transient solid state followed by recanalization to re-establish a lumen. However, some cavities do not fully fuse with the main lumen leading to a duplication of parts of the bowel. These may be completely separate from the main lumen with no connection, giving rise to a duplication cyst (the intra-abdominal cyst requiring excision as a neonate in our patient may have been of this nature). Alternatively, there may be some continuity with the main lumen; in time these can turn into diverticula. The pathogenesis of duplications is unclear, however intrauterine environmental factors such as trauma, hypoxia or a vascular accident have been proposed.

5. Conclusions

We present the case of a young woman with a CEPS associated with a massive small bowel diverticulum, these anomalies theoretically may share an associated embryological origin. The presence of a CEPS should alert the surgeon to the potential for other anomalous anatomy and the risk of catastrophic bleeding associated with unexpected vascular malformations.

Conflicts of interest

None to declare

Funding

None

Ethical approval

This study is exempt from ethical approval in our institution

Consent

Written informed consent was obtained from the patient.

Author contribution

A. Mohtashami and A. Kiat completed the literature review and prepared the manuscript.

J. Cross provided review and amendment of the manuscript.

J. Cross, R. Simon and A. Curtin were present at the initial surgery and provided insight into the case.

Guarantor

Ali Mohtashami will act as guarantor of this work.

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