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Case Report

Congenital Extrahepatic Portosystemic Shunt in a Teenager: a Case Report

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Abstract

The congenital extrahepatic portosystemic shunt is a rare anomaly characterized by aplasia or hypoplasia of intrahepatic portal venous branches that generate complete or partial extrahepatic shunting of portal venous intra-systemic veins. The clinical presentation is variable, including nausea, abdominal pain, anorexia, and jaundice. This entity has been associated with other congenital anomalies, and the most common are the cardiovascular anomalies. Imaging studies are essential to confirm the diagnosis, establish pre-operative planning, and determine the surgical procedure to be performed. We presented the case of a 15-year-old adolescent who had abdominal symptoms and was found to have a portal venous vascular abnormality.

Keywords

gastrointestinal hemorrhage, liver circulation, portal system

INTRODUCTION

The first report about the congenital extrahepatic portosystemic shunt (CEPS) was in 1793 by John Abernethy, hence the name Abernethy's malformation. CEPS is a rare anomaly characterized by aplasia or hypoplasia of intrahepatic portal venous branches that generate complete or partial extrahepatic shunting of portal venous intra-systemic veins.[1] It can be classified into numerous categories based on the vascular relationships.[1] The clinical presentation is variable, ranging from asymptomatic to severe forms of hepatic encephalopathy, pulmonary arterial hypertension, and hepatopulmonary syndrome.[2,3] Additionally, up to 70% of cases are detected before the age of 18.[1] Imaging tests are necessary for diagnosis and to establish pre-operative planning.[3,4] The treatment depends on symptoms, type of shunt, and shunt ratio.[3] We described a case of a teenager who had abdominal symptoms and was found to have a portal venous vascular abnormality.

CASE REPORT

A 15-year-old adolescent with a prior appendectomy and upper gastrointestinal bleeding sought medical attention in the emergency room due to 20 days of abdominal pain in the right upper abdomen, accompanied by a weight loss of around five kilograms. Furthermore, within the past 48 hours, the patient experienced two episodes of vomiting and a fever reaching a maximum of 39°C. The patient's physical examination revealed normal vital signs, including a blood pressure of 114/58 mmHg, a heart rate of 67 beats per minute, a breath rate of 18 breaths per minute, and a temperature of 36.3°C. The abdomen was soft, but there was tenderness

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in the right hypochondrium and no symptoms of peritoneal irritation.

Initial laboratories were reported without alterations, as well as a normal total abdominal ultrasound. Abdominal computed tomography (CT) with contrast showed a portal venous vascular anomaly suggestive of an Abernethys malformation ***(Fig. 1)****. Additionally, a portal Doppler reported scarcer collateral vessels in the splenic hilum* ***(Fig. 2)****.*

The patient's abdominal pain worsened during hospitalization, prompting a new abdominal CT scan that suggested ileum and the previously reported perisplenic and mesenteric collateral circulation ***(Fig. 3)****. An esophagogastroduodenoscopy (EGD) was done, which revealed a hiatal hernia Hill I, LA grade B esophagitis, a gastric ulcer Forrest III, and two duodenal ulcers Forrest IIC and III. The patient received treatment with an anti-ulcer drug (lansoprazole 30 mg per day and sucralfate 1 g every 8 hours), which resulted in an improvement of his symptomatology. He is currently undergoing outpatient vascular surgery.*

DISCUSSION

Abernethy malformation is a rare congenital vascular anomaly with an estimated prevalence of one per 30000–50000 live births.[5] Morgan and Superina categorize it into

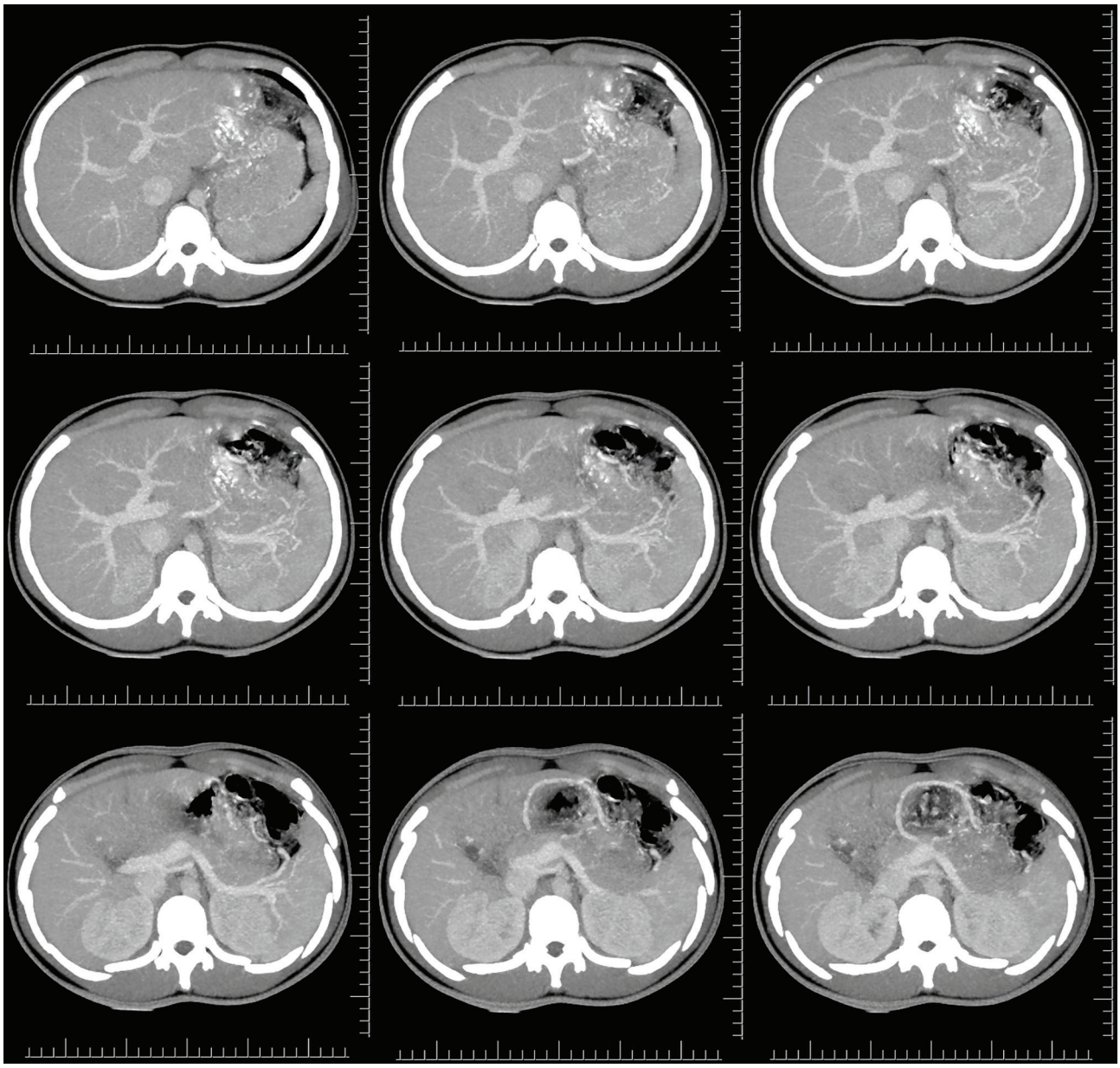

Figure 1. An abdominal CT scan with contrast revealed a portal venous vascular anomaly along with a variant of the superior mesenteric anatomy. The portal and splenic veins were slightly dilated, and the spleen had a maximum diameter of 14 cm, which could be a sign of an Abernethy-type malformation.

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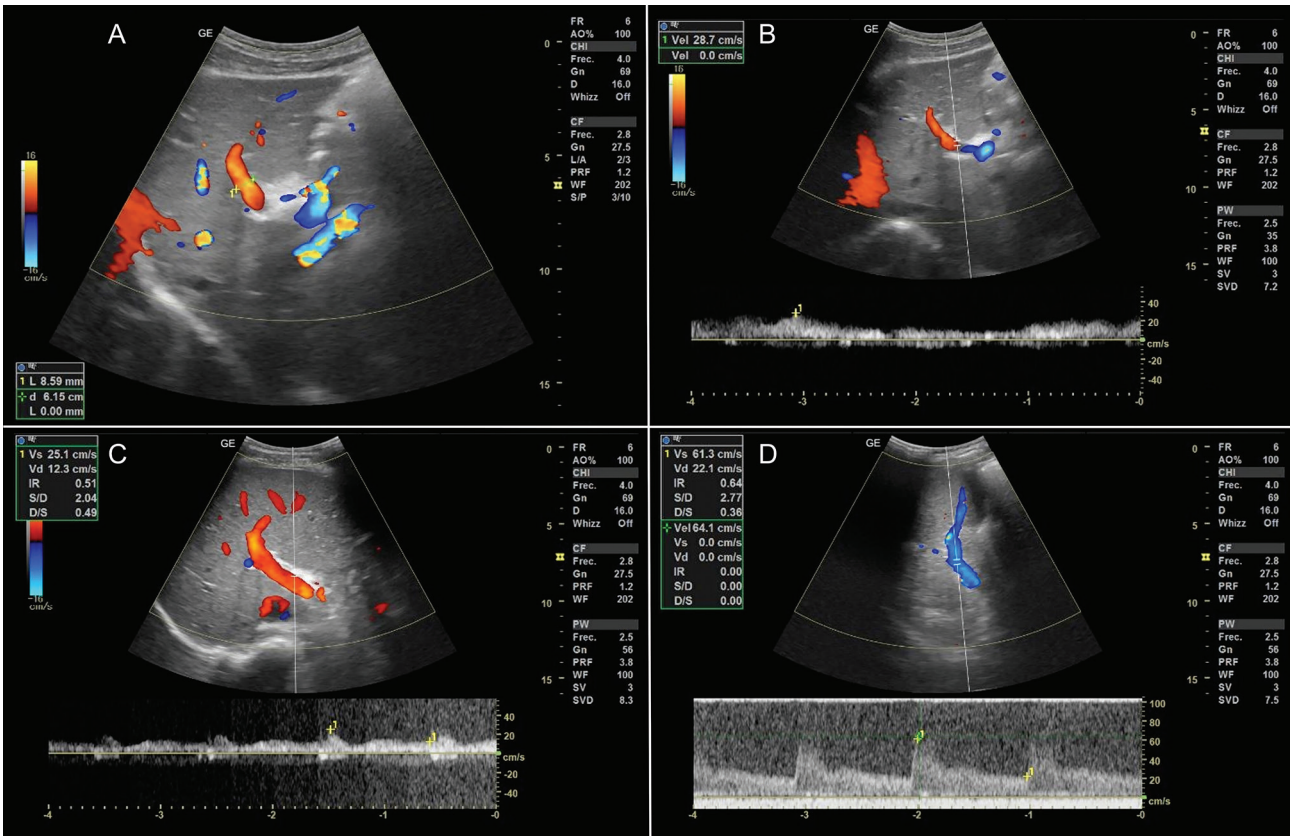

Figure 2. A portal Doppler showed an 8.5-mm-diameter portal vein with a hepatopetal flow velocity of 28 cm/s and a hepatic artery with an IR of 0.51 and a velocity of 30 cm/s. The spleen was 106 mm in size and had a splenic vein with a velocity of 17 cm/s and an artery with an IR of 0.6 and a velocity of 64 cm/s. There were fewer collateral vessels in the splenic hilum.

two types based on whether it is a complete (type I) or partial (type II) portosystemic shunt.[6] Simultaneously, type I can be further categorized into subtypes Ia and Ib based on whether the superior mesenteric vein and splenic vein converge with the portal vein or not.[4] This entity has a wide spectrum of symptoms, including nausea, abdominal pain, anorexia, and jaundice. Other symptoms include respiratory effects like dyspnea, cyanosis, and pulmonary hypertension.[4] The majority of the patients have preserved liver function. Other laboratory findings, including hyperammonemia and high serum galactose, can be developed during childhood.[7]

Imaging studies are required to confirm the diagnosis and subclassify among the options. Multi-slice computed tomography (MSCT) is a rapid test with high spatial resolution that, in contrast with other studies, can detect small vessels.[4] When other studies yield inconclusive results, angiography becomes the preferred method. Finally, a liver biopsy is decisive in differentiating between total or partial aplasia.[1,3] This entity has been associated with other congenital anomalies, and the most common are cardiovascular.[1,8] Other complications associated with that entity include neonatal cholestasis, encephalopathy, liver masses, and, in unusual cases, osteoporosis, upper gastrointestinal bleeding, and nephrotic syndrome.[5,9,10]

In symptomatic patients, or those who have a shunt ratio >60%, in cases of type I shunt, a liver transplantation is required, and type II requires closure by either endovascular or surgical means.[3] Additionally, surgical hepatectomy can be considered for adenomas or hepatocellular carcinomas, which are associated with these vascular anomalies with a prevalence of 15% and 12%, respectively.[7] In the presented case, our patient had nonspecific clinical manifestations. It is important to mention that he experienced a previous episode of upper gastrointestinal bleeding with no identifiable reason, which could be linked to the vascular abnormality. The imaging studies were crucial for diagnosing the condition. However, additional studies are necessary because this pathology is linked to other congenital abnormalities. Similarly, a specialist should conduct follow-up to assess the need for surgical intervention and minimize the risk of complications.

CONCLUSIONS

CEPS is a rare anomaly with a wide spectrum of symptoms, from asymptomatic to life-threatening events. The imaging studies are the cornerstone of diagnosis. Treatment is based on the pattern of the shunt, including liver transplantation or shunt closure via endovascular or surgical means, to reduce the risk of complications.

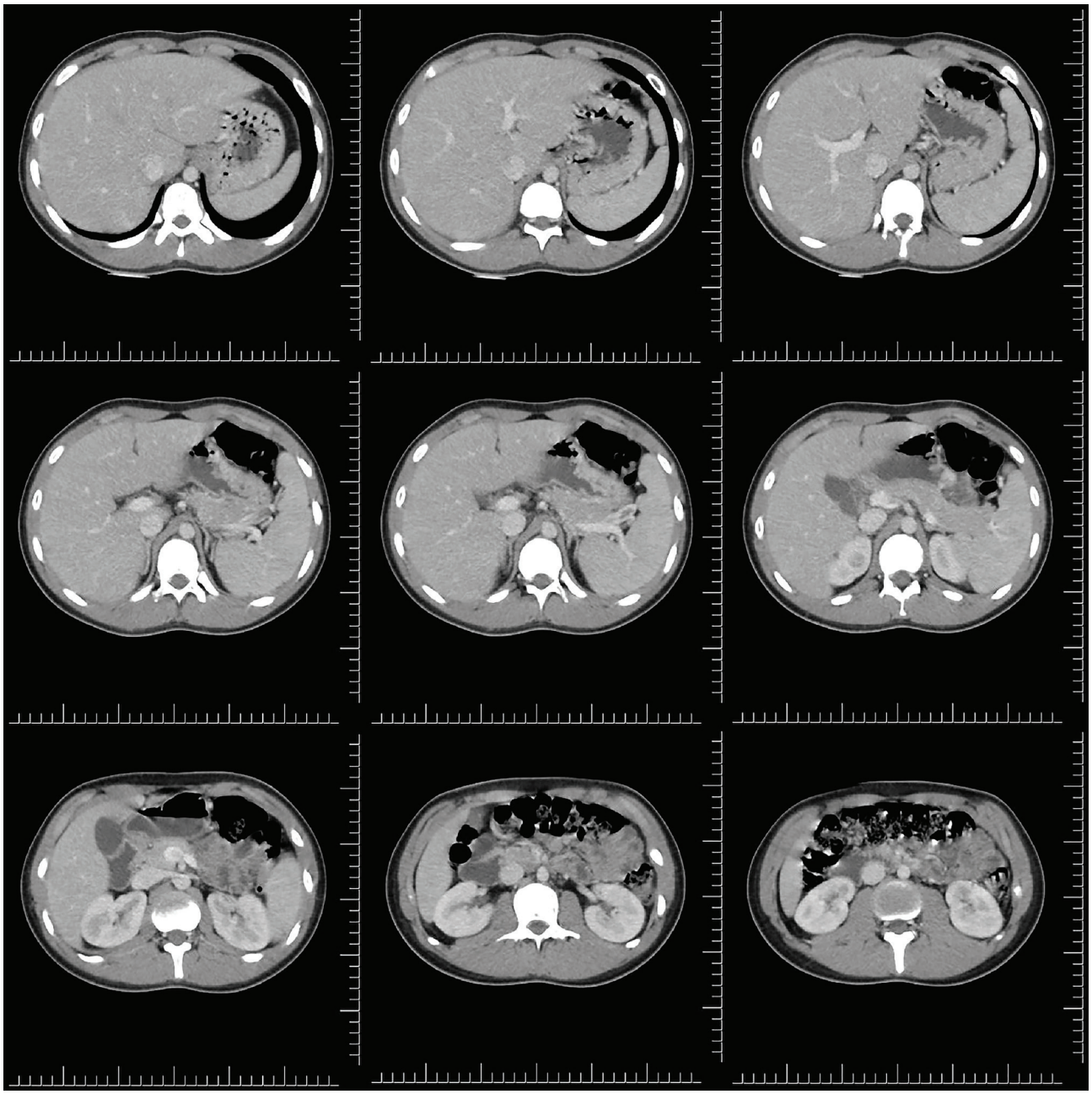

Figure 3. An abdominal CT scan with contrast revealed an unspecific, moderate distention of the duodenum, a limited evaluation of the gastric chamber, and a moderate distention of the small intestine, primarily in the mesogastrium, indicating ileum, as well as perisplenic and mesenteric collateral circulation.

Authors contribution

D.A.M. and A.R.U.: investigation and writing – original draft; D.F.V., M.A.O.M., and A.M.B.D: conceptualization and investigation; L.G.A: supervision, and writing – review & editing.

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Врождённый внепечёночный портосистемный шунт у подростка: описание случая

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Резюме

Врождённый внепечёночный портосистемный шунт – редкая аномалия, характеризующаяся аплазией или гипоплазией внутрипечёночных ветвей воротной вены, которые создают полное или частичное внепечёночное шунтирование внутрисистемных воротных вен. Клинические проявления разнообразны и включают тошноту, боли в животе, анорексию и желтуху. Эта патология связана с другими врождёнными аномалиями, наиболее распространёнными из которых являются сердечно-сосудистые аномалии. Исследования с помощью визуализации необходимы для подтверждения диагноза, установления предоперационного планирования и определения хирургической процедуры, которая должна быть выполнена. Мы представили случай 15-летнего подростка, у которого были абдоминальные симптомы и была обнаружена аномалия воротной вены.

Ключевые слова

желудочно-кишечное кровотечение, печёночное кровообращение, портальная система