Atypical Manifestation of Patent Ductus Venosus in a Child: Intervening against a Paradoxical Presentation

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A 6-year-old boy with massive gastrointestinal bleeding was found to have a large patent ductus venosus (PDV). Systemic symptoms of PDV (eg, hypergalactosemia and hepatic, pulmonary, and cardiac dysfunction) are frequent. However, gastrointestinal bleeding with the presence of a large portosystemic shunt is not a known complication of this anomaly. The shunt was successfully treated with embolization by using the Amplatzer vascular plug, with immediate cessation of bleeding. The authors propose that relative ischemia of the bowel, rather than portal hypertension, was the cause of the gastrointestinal bleeding in this child.

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Abbreviation: PDV = patent ductus venosus

THE ductus venosus is a fetal communication between the portal and systemic venous systems, shunting blood directly from the umbilical vein into the inferior vena cava. No longer required in neonates, the ductus venosus closes after birth. Patent ductus venosus (PDV) is a rare congenital anomaly in which portal blood maintains a direct communication with the systemic veins beyond the neonatal period. According to published reports, while PDV can be found incidentally without any associated symptoms, systemic manifestations such as hypergalactosemia, hepatic encephalopathy, and hepatopulmonary syndrome with heart and liver dysfunction are commonly encountered. Treatment approaches for PDV include conservative therapy and surgical or interventional occlusion.

Herein, we report a case of a large and unusual PDV in a child who presented with massive gastrointestinal bleeding, which was successfully treated with transvenous embolization by using the Amplatzer vascular plug.

CASE REPORT

Our institution does not require institutional review board approval for the publication of retrospective case reports. However, in keeping with the ethical conduct of studies, the principles of the Declaration of Helsinki were followed.

A 6-year-old boy was admitted to the intensive care unit at our institution with severe acute gastrointestinal bleeding. Multiple episodes of melena and bright red blood per rectum were encountered before and during resuscitation. On admission, the patient was severely anemic (hematocrit level, 14%–18% [0.14–0.18]), had an elevated total/direct bilirubin level (up to 4.9/4.2 mg/dL [83.8/71.8 μmol/L]; normal range, 0.3–1.2/0–0.4 mg/dL [5–21/0–6.8 μmol]), a borderline mildly elevated blood urea nitrogen level (up to 24 [8.6 mmol/L]; normal value, 5–18 mg/dL [1.8–6.4 mmol/L]), and mildly elevated liver enzymes (aspartate aminotransferase, 86 U/L; alanine aminotransferase, 113 U/L). The coagulation profile was unremarkable. The patient received multiple transfusions with blood and blood products and was started on continuous octreotide infusion.

The patient's medical history was significant for an "arteriovenous malformation of the liver," which was detected at prenatal and postnatal ultrasonography (US) at an outside institution. These studies were not available for immediate review. In addition, the patient had agenesis of the corpus callosum and mild developmental delay.

Upper and lower gastrointestinal endoscopy and biopsies failed to reveal the cause of the bleeding. Exploratory laparotomy and intraoperative small bowel enterostomy were performed. Multiple small bowel segments demonstrated focal erosions and superficial ulceration, with small points of bleeding; these segments

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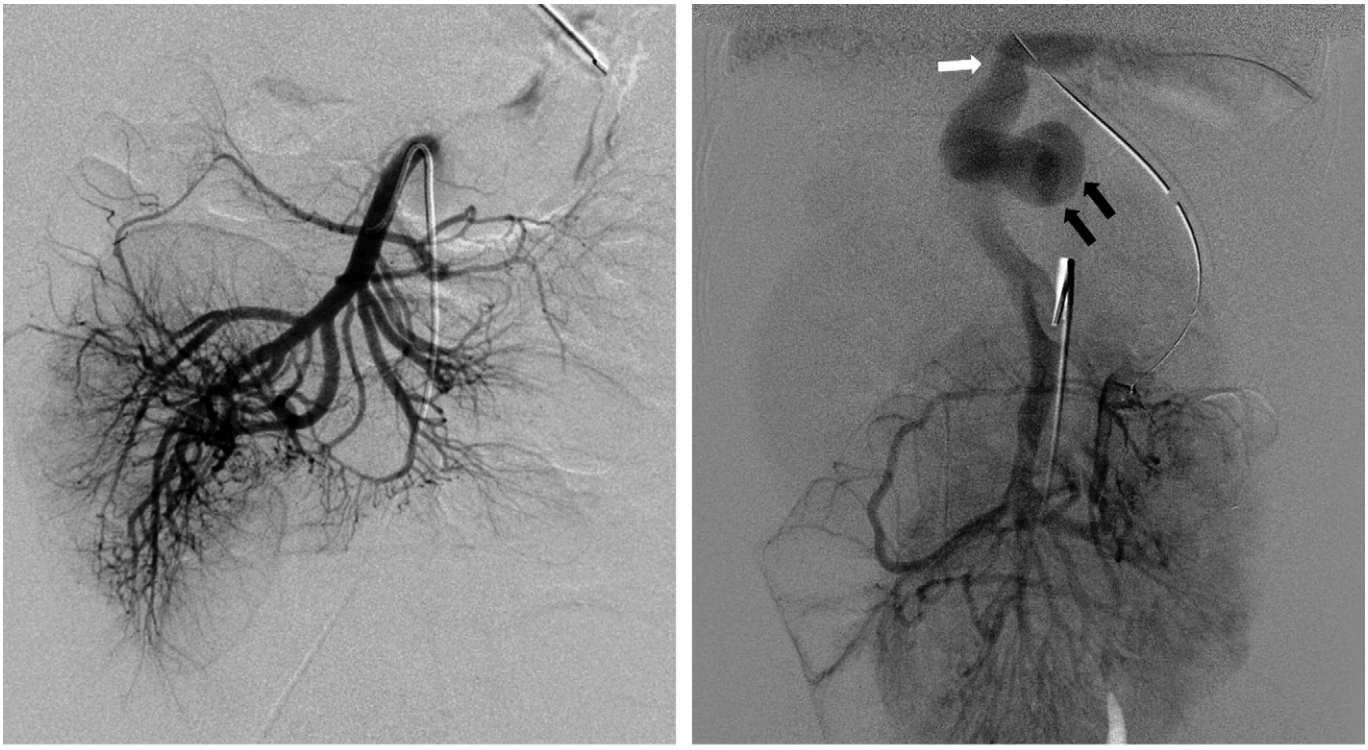

a. b.

Figure 1. Images from selective angiography of the superior mesenteric artery. **(a)** Angiogram obtained in the arterial phase demonstrates the prominent size of this vessel, without evidence of shunting or bleeding. **(b)** Image in the venous phase shows a large and tortuous intrahepatic portal vein (black arrows) with direct communication with the hepatic venous confluence. The white arrow indicates the ductus venosus.

were subsequently resected. An incidental Meckel diverticulum was noted and laparoscopically resected. During the same operation, a left partial colon resection was performed as earlier dynamic abdominal scintigraphy with technetium 99m–labeled red blood cells suggested accumulation of labeled red blood cells in the splenic flexure.

However, on the 2nd postoperative day, the child experienced another significant gastrointestinal bleed. Computed tomography (CT), angiography, and liver biopsy were performed. At angiography, the superior mesenteric artery was enlarged without angiographic evidence of active bleeding or arteriovenous shunting (**Fig 1a**). On the portal phase of mesenteric arteriography, a large tortuous PDV was demonstrated between the portal and hepatic vein confluence (**Fig 1b**). No opacification of the intrahepatic portal branches was noted. The CT findings helped confirm the diagnosis (**Fig 2**) and showed normal-appearing liver parenchyma with no evidence of a mass lesion.

Histopathologically, the liver specimen exhibited focal mild centrilobular sinusoidal dilatation and perisinusoidal fibrosis. The bowel specimens showed mucosal and submucosal vascular congestion. There was no evidence of vascular malformation, inflammation, or cirrhosis.

Due to persistent bleeding not responding to conservative measures and after extensive multidisciplinary discussion at our vascular anomalies conference, the decision was made to proceed with percutaneous embolization of the PDV.

Written informed consent was obtained. The procedure was performed under general anesthesia. Bilateral internal jugular accesses were established with a 10-F vascular sheath into the right internal jugular vein and a 5-F sheath into the left. A 5-F Berenstein catheter (Cordis, Miami, Florida) was advanced into the right hepatic vein through the left internal jugular sheath. Biplane digital subtraction was used for right hepatic venography, which was used to mark the confluence of the hepatic veins.

Through the right internal jugular sheath, the PDV was catheterized directly in a retrograde fashion. Portal venography was performed. A 13-mm Berenstein occlusion balloon (Boston Scientific, Natick, Massachusetts) was advanced into the PDV, and portal venography was repeated with the balloon inflated. This demonstrated two "underdeveloped" branches of the intrahepatic portal vein (**Fig 3**). The diameter and length of the PDV were measured (14 and 25 mm, respectively). Pressure measurements within the shunt were 13 mm Hg without balloon occlusion and 21 mm Hg with occlusion. At this juncture, it was decided to proceed with shunt occlusion by using a slightly oversized Amplatzer vascular plug. An 8-F long Cook straight vascular sheath was advanced through the right jugular sheath and the tip placed into the shunt. Venography