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Case Report

Ruptured idiopathic hepatic artery pseudoaneurysm causing portal vein thrombosis with portal hypertension and variceal bleeding ☆,☆☆

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ABSTRACT

Portal vein thrombosis (PVT) is an important cause of noncirrhotic portal hypertension. Non-cancerous extrinsic compression of portal vein to drive PVT formation is rare, but important to identify. A 64-year-old female with idiopathic hepatic artery pseudoaneurysm (HAPA) rupture 7 months prior presented with acute-onset hematemesis and melena and was found to have prehepatic portal hypertensive variceal bleeding. Her HAPA-related retroperitoneal hematoma had resulted in portal vein compression, thrombosis, and cavernous transformation despite prompt stent graft placement across the ruptured HAPA, and required definitive treatment by transjugular intrahepatic portosystemic shunt creation with portal vein reconstruction utilizing a trans-splenic access. This case highlights the importance of interval abdominal imaging and hypercoagulability screening for noncirrhotic patients at-risk for PVT, which identified the patient as a heterozygous carrier of Factor V Leiden.

Introduction

The prehepatic occlusion of the portal vein by thrombus is an important cause of portal hypertension (PHTN) [1]. Portal vein thrombosis (PVT) occurs in diverse clinical contexts that drive thrombus formation by potentially enabling multiple elements of Virchow's triad (stasis, endothelial injury, and hypercoagulability) [2]. Not surprisingly, local factors acting in the vicinity of portal vein including inflammation, infection,

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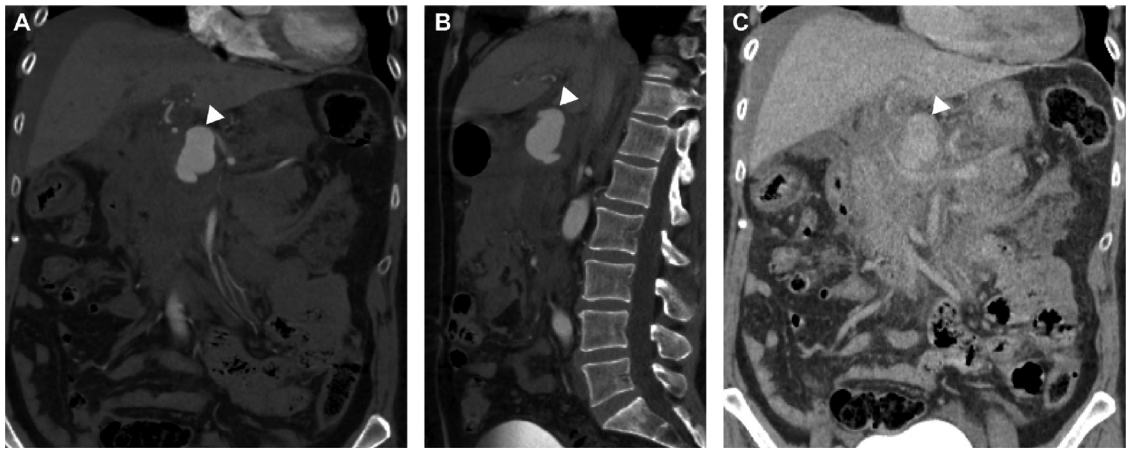

Fig. 1 – Initial presentation. Slices of contrast-enhanced CT in arterial (A-B) and delayed (C) phase showing ruptured common HAPA (arrowhead, A-C) prior to stent grafting. Ruptured HAPA measuring 4.0 cm x 2.4 cm x 2.3 cm (A-B) impinged on portal venous structures in the porta hepatis (C).

malignancy, and iatrogenic injury are frequently identified, as well systemic factors such as inherited or acquired hypercoagulability.

Hepatic artery pseudoaneurysm (HAPA), which lacks a complete 3-layer arterial wall, tends to rupture or fistulize rather than cause mass effect on neighboring structures [3,4]. HAPA most commonly occurs in the right or common hepatic artery and often has an identifiable prior injurious event such as abdominal trauma, iatrogenic hepatobiliary procedure, or local inflammation [3]. We describe a case of ruptured idiopathic common HAPA with unique sequelae of PVT, cavernous transformation, and PHTN-related variceal bleeding.

Case report

A 64-year-old Caucasian female with no past medical history presented to an outside institution with new-onset, nonbilious, nonbloody vomiting and severe epigastric abdominal pain. There was no history of alcohol use, liver disease, trauma, hepatobiliary procedures, pancreatitis, or hypercoagulable state. Computed tomography (CT) scan of her abdomen showed a ruptured common HAPA causing a large retroperitoneal hematoma (Fig. 1). She underwent emergent hepatic artery stent graft placement (6 mm × 5 cm covered Viabahn, Gore Medical, Flagstaff, AZ) and was discharged on aspirin and clopidogrel with resolution of her symptoms.

Seven months later, she represented with an episode of sudden-onset hematemesis. She reported several months of mild diffuse abdominal pain and distension, as well as 3 recent episodes of self-limited melena. Her blood pressure was 162/97 mm Hg and heart rate was 83 beats/min. Physical exam was significant for soft, slightly distended abdomen without any organomegaly. Labs showed a hemoglobin of 6.7 g/dL (reference range 12-16), platelets 342 k/mm^3 (reference range 150-450), international normalized ratio 1.06 (reference range 1-2), normal leukocyte differential, and normal liver chemistries. CT abdomen and pelvis with contrast showed a resolving hematoma related to the ruptured HAPA, however with interval development of cavernous transformation of the portal vein. The liver appeared normal in size and echotexture, but there was trace ascites (Fig. 2). She was transferred to our institution for further management.

Upon arrival, she was hemodynamically stable. Aspirin and clopidogrel were held. Esophagogastroduodenoscopy identified large bleeding esophageal varices with red wale signs, which were banded. She developed hematochezia and a hemoglobin drop to 6.3 g/dL despite receiving 4 units of packed red blood cells over the next 3 days. Repeat endoscopy showed small esophageal varices and portal hypertensive gastropathy, but no active bleeding. She continued to have PHTN-related hematochezia requiring serial blood transfusions. During the course of her stay, she developed abdominal distension and ascites requiring paracentesis. Ascitic fluid analysis was consistent with spontaneous bacterial peritonitis (ascitic fluid: 800 WBC, 60% neutrophils), which was treated with ceftriaxone and intravenous albumin.

Interventional radiology was consulted and she underwent a transjugular intrahepatic portosystemic shunt (TIPS) creation with portal vein reconstruction utilizing trans-splenic access (Fig. 3). A 6-Fr sheath was inserted into the splenic vein *via* trans-splenic approach. A 0.035 inch (0.89 mm) Glide Advantage wire (Terumo Medical Corporation, Somerset, NJ) and 5-Fr Berenstein catheter were used to navigate through the native, now-stenotic main portal vein channel. A 10 mm snare was positioned in the mid-right intrahepatic portal vein. Via a 10-Fr sheath access in the right internal jugular vein, the snare was targeted and a Rosch-Uchida needle (Cook Medical, Bloomington, IN) was used to cross from the middle hepatic vein into the right portal vein. A 10 mm × 8 cm + 2 cm Viatorr (Gore Medical) was deployed and postdilated to 10 mm. Prolonged balloon angioplasty to 10 mm was also performed along the unstented, stenotic portion of the native portal vein. After TIPS, the mean portosystemic gradient decreased from 13 to 4 mm Hg. Her hematochezia and ascites resolved and she was discharged.

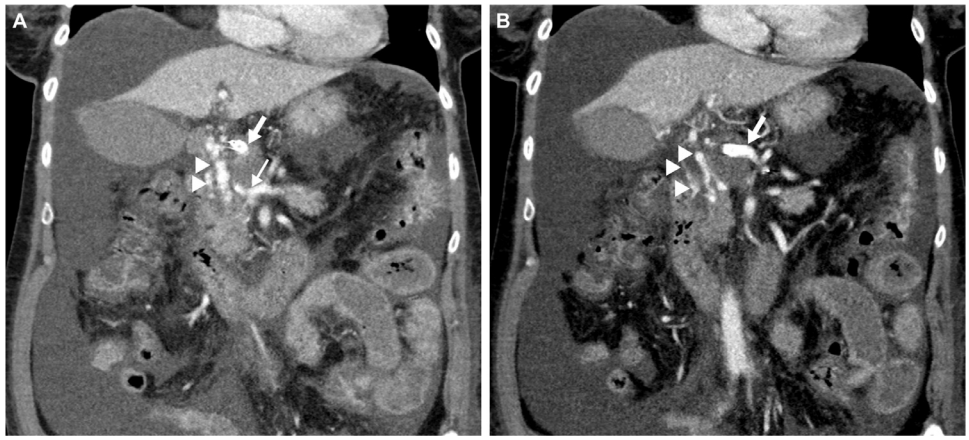

Fig. 2 – Second presentation at 7 months. Two slices of contrast-enhanced CT in venous phase showing previously ruptured common HAPA with stent graft (thick arrow, A-B) causing portal cavernous transformation (arrowhead, A-B). Superior mesenteric and splenic veins merging to form the stenotic native portal vein (thin arrow) giving rise to collaterals.

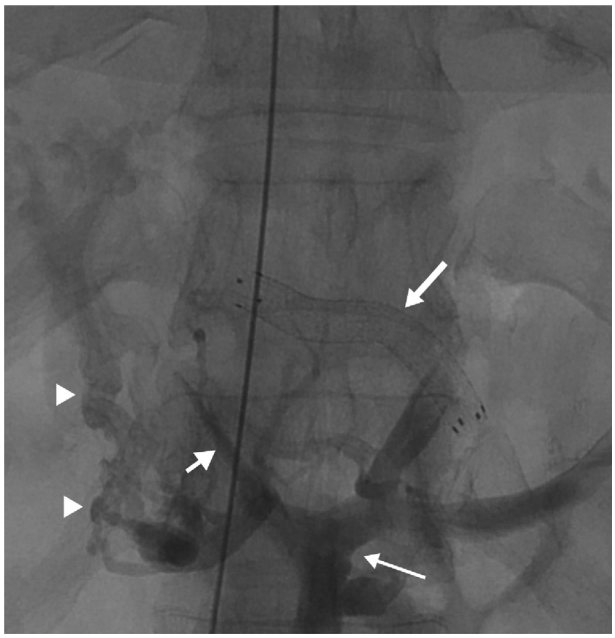

Fig. 3 – Second presentation at 7 months. Trans-splenic venogram during TIPS showing superior mesenteric and splenic veins merging to form the prestenotic portal vein (thin long arrow) with collaterals typical of cavernous transformation (arrowheads). The stenotic thread-like native main portal vein channel (short arrow), adjacent to common HAPA with stent graft (thick long arrow), was successfully recanalized as the TIPS was created from middle hepatic vein to right portal vein.

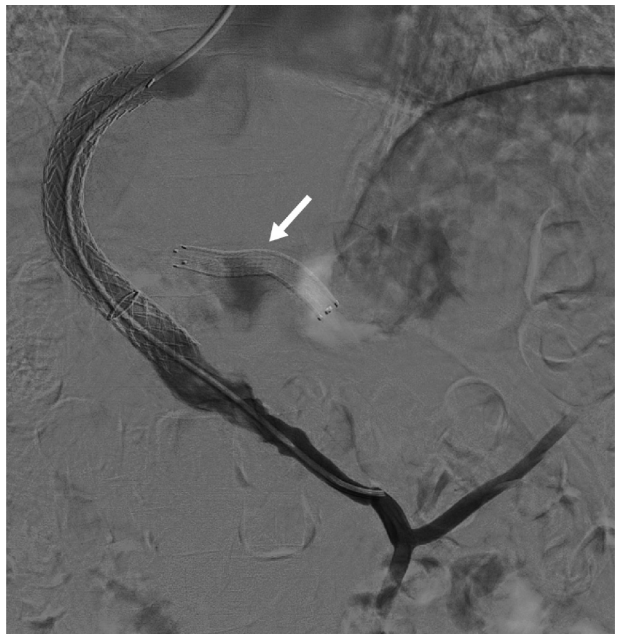

Fig. 4 – Follow-up after second presentation. Conventional venogram 1-month post-TIPS showing brisk flow through TIPS without filling defect or anomaly. Nearby common HAPA with stent graft (thick long arrow).

At 1-month follow-up, she was asymptomatic, and her TIPS was patent both by color Doppler ultrasound and a conventional venogram (Fig. 4). Subsequently at 3-month clinic follow-up, color Doppler ultrasound continued to show patent TIPS. Screening for hypercoagulability revealed Factor V Leiden (FVL) heterozygosity, but was negative for elevated homocysteine, antiphospholipid antibodies, and prothrombin G20210A.

Discussion

We present a patient with noncirrhotic, prehepatic PHTN-related variceal bleeding due to idiopathic ruptured HAPA and resultant portal vein compression and thrombosis. This was a unique clinical course for HAPA, which has a high risk

for progression, fistulization, or rupture, and often presents as right upper quadrant pain, gastrointestinal bleeding, or hemoperitoneum [4].

Her clinical course with the development of PVT likely resulted from an interplay of HAPA-related extrinsic compression slowing portal flow combined with her inherited FVL hypercoagulability. Inherited or acquired hypercoagulability is frequently identified in patients with PVT: one study noted 26 of 36 noncirrhotic PVT patients had an identifiable hypercoagulable state [5]. FVL is the most common inherited hypercoagulability in those of European descent with 5% frequency. Interestingly, studies of both noncirrhotics and cirrhotics have noted no statistically different FVL frequency in PVT patients vs controls, with a trend toward increased FVL in one study [5-8]. This is in sharp contrast to statistically higher prothrombin G20210A frequency in PVT, which suggests that FVL alone may not play as potent of a role in stimulating PVT development.

Noncirrhotic PVT is rare compared to cirrhosis-related PVT [9]. Not surprisingly, there is a lack of guidelines for PVT screening in at-risk noncirrhotic patients in contrast to cirrhotic patients, where guidelines exist for ultrasound screening due to higher risk for hepatocellular carcinoma during which PVT may be found [10]. The key benefit to identifying PVT early is the possibility of early anticoagulation initiation with low molecular weight heparin or vitamin K antagonists, which can help dissolve clots and halt thrombus extension [9,11]. Though rare, PVT expanding proximally into superior mesenteric vein can cause life-threatening intestinal infarction.

In our patient, HAPA rupture resulted in significant extrinsic compression of the portal vein at the porta hepatis. Given the slow-resolving and persistent nature of this hematoma's mass effect on the portal vein, we posit that our patient could have benefited from regular PVT screening, hypercoagulability testing, and consideration of pre-emptive TIPS before she developed refractory PHTN-related bleeding. This raises an important management consideration, because hematoma is not limited to rare pseudoaneursymal ruptures and can arise after trauma, biopsy, or surgery. Of note, our patient did undergo CT abdominal imaging to follow-up HAPA stent graft placement 1 month after her procedure, but did not have any subsequent follow-up due to the COVID-19 pandemic.

The development of complete PVT with cavernous transformation has historically posed technical challenges for performing TIPS to treat refractory PHTN-related bleeding [12]. Various strategies to facilitate locating and navigating through the intrahepatic portal vein have included ultrasound guidance and percutaneous transhepatic access [13]. The strategy we chose, portal vein reconstruction with trans-splenic access, has emerged as a very safe and effective strategy for treating complete PVT with cavernous transformation in both cirrhotics [14,15] and noncirrhotics [16].

Patients with PVT without other comorbidities including ours have a good prognosis [17]. This report highlights PVT and PHTN development despite timely HAPA stenting and resolving extrinsic compression, which underscores the need for follow-up, hypercoagulability screening, and definitive intervention in this unique patient population.

Authors' contributions

K. Ni provided care from endoscopic service and wrote the manuscript. C. Jansson-Knodell provided care from endoscopic service, helped interpret clinical findings, and revised the manuscript. M.E. Krosin and P.M. Haste performed TIPS creation with portal vein reconstruction, interpreted radiological findings, and revised the manuscript. I. Obaitan and L.E. Nephew led the care from hepatology service, helped interpret clinical findings, and revised the manuscript. S.V. Sagi performed endoscopic interventions, helped interpret clinical findings, and revised the manuscript.

Patient consent

Informed patient consent was obtained for publishing this case report.

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