A rare case of gastric varices and splenic artery aneurysm secondary to splenic arteriovenous fistula

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ABSTRACT
A 33-year-old male with no relevant medical history presented with a few months of fatigue and reduced exercise tolerance and was found to have iron-deficiency anemia. An esophagogastroduodenoscopy revealed a cluster of isolated gastric fundal varices with high-risk stigmata. Serologic workup for cirrhosis was negative, and a FibroScan measured liver stiffness at 4.2 kilopascals. Computed tomography (CT) of his abdomen and pelvis showed non-cirrhotic portal hypertension, as well as the presence of a splenic arteriovenous (AV) fistula and splenic artery aneurysm (SAA). Resection of the fistula, SAA, and spleen completely resolved the gastric varices and anemia.

KEYWORDS: arteriovenous fistula; gastric varices; splenic artery aneurysm

CASE PRESENTATION
A 33-year-old male with no relevant medical history presented to his primary care provider with a few months’ history of fatigue, reduced exercise tolerance, and mild upper abdominal discomfort, with no history of overt gastrointestinal bleeding. Laboratory tests revealed a low hemoglobin level of 4.6 mmol/L and ferritin of 7.6 pmol/L with normal liver function tests. He was referred to gastroenterology for further workup of his iron deficiency anemia. An esophagogastroduodenoscopy (EGD) revealed a cluster of isolated gastric fundal varices with high-risk stigmata as indicated by adherent clots (Figure 1), which was confirmed on endoscopic ultrasound (Figure 2). Chronic liver disease workup was performed, including serologies for hepatitis A, B, C; immunoglobulins profile; autoimmune antibodies; ceruloplasmin; and alpha-1-antitrypsin, all of which were normal. The patient denied any history of significant alcohol use or other risk factors for cirrhosis. A FibroScan measured liver stiffness at 4.2 kPa, suggesting a low likelihood of fibrosis.

He underwent computed tomography (CT) of his abdomen and pelvis, which revealed a dilated
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Figure 1: Esophagastroduodenoscopy revealing a cluster of isolated gastric fundal varices with stigmata of adherent clots.

Figure 2: Radial endoscopic ultrasound with doppler confirms the presence of large fundal varices.

Figure 3: CT scan revealing a fistula between the splenic artery and vein (yellow arrow) with early enhancement of the splenic vein from the shunt (blue arrow) and a large saccular splenic artery aneurysm (SAA) measuring approximately 2 × 3 × 3 cm (red arrow).

The patient had a history of portal and splenic vein suggestive of portal hypertension, with no thrombosis. There was hypodensification of the spleen posteriorly, concerning for infarct. Workup for hypercoagulable disorders including lupus anticoagulant ratio, anti-cardiolipin IgG, protein C and S levels, anti-thrombin activity, and Factor V Leiden mutation were normal. To better assess the portal system vasculature, he underwent a triple-phase CT of his abdomen (Figure 3–5).

The CT scan revealed a fistula between the splenic artery and vein (Figure 3, yellow arrow; Figure 4) with early enhancement of the splenic vein from the shunt (Figure 3, blue arrow) and a large saccular splenic artery aneurysm (SAA) measuring approximately 2 × 3 × 3 cm (Figure 3, red arrow; Figure 5). There was no thrombus within the portal venous system. Given the size of the aneurysm and presence of splenic infarct, the patient...
underwent laparoscopic splenectomy and resection of the SAA. An EGD 8 months after showed resolution of the gastric varices, and follow-up blood tests showed that hemoglobin and iron indices had normalized.

**DISCUSSION**

While cirrhosis is the most common cause of portal hypertension, non-cirrhotic portal hypertension is an important clinical entity for clinicians to recognize. The various etiologies of non-cirrhotic portal hypertension can be divided into pre-hepatic, intrahepatic, and post-hepatic causes. In a patient with no pre- or post-hepatic thrombosis and no risk factors or symptoms of schistosomiasis or other intrahepatic pathology, clinicians should consider splanchnic AV fistulas in the differential for non-cirrhotic portal hypertension (1). Dedicated vascular imaging should be obtained to assess for these conditions to avoid delays in diagnosis and management.

Splenic arteriovenous (AV) fistulas are rare, having first been described in the nineteenth century by Wiegert (2). The majority occur after rupture of an SAA but can also be traumatic, congenital, or infectious in origin. In our case, increased blood flow through the splenic AV fistula likely led to the development of the SAA, non-cirrhotic portal hypertension and gastric varices. This hypothesis is supported by the resolution of gastric varices following the resection of the splenic AV fistula and SAA by laparoscopic splenectomy. The splenic infarct seen on imaging may have been caused by a smaller AV fistula shunting blood away from the spleen and directly to the splenic vein. Unfortunately, this cannot be confirmed with the imaging that had been done previously.

Sinistral, or left-sided portal hypertension, is a rare phenomenon whereby a pathologic process increases backflow from the splenic vein into the short gastric and gastroepiploic veins, resulting in portal hypertension, which is confined to the left
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This can lead to the development of the isolated gastric varices without esophageal varices, distinguishing this from generalized portal hypertension (although isolated gastric varices may also occur with generalized portal hypertension) (4). As the splenic vein runs closely along the superior pancreatic surface, it is susceptible to pathology in the pancreas. The most common causes of sinistral portal hypertension involve obstruction or thrombosis of the splenic vein, which may occur from chronic pancreatitis, neoplasms, pancreatic pseudocysts, and other pathology (3).

Portal hypertension occurs in more than half of patients with splanchnic AV fistulas (5,6). The pathogenesis is thought to be due to increased portal blood flow and adaptive portal fibrosis (7). Development of SAA due to splenic AV fistula has been observed in two previous case reports (8,9). Previous case reports have described the development of esophageal varices from splenic AV fistulas (10,11). There has been one reported case of gastric variceal bleed secondary to splenic AV fistulas, in a 49-year-old-man who also had a SAA (12). Our case report is the first documented occurrence of gastric varices secondary to splenic AV fistula in a young male with no past medical history or alcohol use. In previous cases, the splenic AV fistulas were treated with transcatheter arterial embolization in one case (10) and with laparotomy and resection of the fistula in the others (11,12).

Given the rarity of splenic AV fistulas and SAA, there are no published guidelines on the management of these clinical entities. While visceral artery aneurysms are uncommon, SAA is the most common (60%), followed by hepatic (20%) (13). Prevalence of SAA was 0.1% to 0.2% in an autopsy study (14), while a study of 3,600 arteriograms found an incidence of 0.78% (15). Rupture of SAA is associated with a 25% mortality, and management of these entities should be done in consultation with general surgery and interventional radiology (16). Different approaches include resection with or without splenectomy, surgical ligation, and endovascular techniques, including transcatheter arterial embolization and covered stent exclusion. The optimal approach will depend on various factors, including patient comorbidities, size of the SAA, and location of the AV fistulas. While there is no specific size criterion for the treatment of SAA supported by the literature, 2 cm is a commonly cited cut-off (17–19).

CONTRIBUTIONS: Conceptualization, JH Zhu, F Zhou; Investigation, F Zhou, CB Lightfoot, G Williams, JH Zhu; Writing – Original Draft, F Zhou; Writing – Review & Editing, F Zhou, CB Lightfoot, G Williams, JH Zhu; Supervision, JH Zhu.

ETHICS APPROVAL: N/A

INFORMED CONSENT: We confirm that informed patient consent has been secured from all patients whose personal information is included in the manuscript or the parents or guardians of minors.

REGISTRY AND THE REGISTRATION NO. OF THE STUDY/TRIAL: N/A

FUNDING: No funding was received for this work.

DISCLOSURES: The authors have nothing to disclose.

PEER REVIEW: This article has been peer reviewed.

ANIMAL STUDIES: N/A

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