Isolated Gastric Fundal Varices Caused by Diffuse Large B-Cell Lymphoma

Miles Shen, AB¹, Sarangdev Vaidya, MBS¹, Nneoma Okoronkwo, MD², Edward Alder, MD³, and Sushil Ahlawat, MD²

¹Rutgers New Jersey Medical School, Newark, NJ
²Department of Gastroenterology, Rutgers New Jersey Medical School, Newark, NJ
³Greystone Park Psychiatric Hospital, Morris Plains, NJ

ABSTRACT

A 59-year-old man with diffuse large B-cell lymphoma, recently diagnosed from a renal biopsy, presented to the emergency department with melena, dizziness, and epigastric pain. He was tachycardic and had a hemoglobin level of 6.4 g/dL. Esophagogastroduodenoscopy revealed isolated gastric fundal varices with stigmata and no esophageal varices. Abdominal ultrasound with Doppler showed a normal-appearing liver, patent splenic vein and hepatic vasculature, and no splenic vein thrombosis. He was managed supportively and discharged. A positron emission tomography-computed tomography scan for staging later revealed extensive neoplastic involvement of the pancreas, gastrohepatic ligament, celiac trunk, and perigastric and splenic hilar regions.

INTRODUCTION

Portal hypertension is a well-understood sequela of liver cirrhosis. Approximately 20% of patients with portal hypertension present with gastric varices, of which gastroesophageal type varices comprise the majority.¹ Portal hypertension may also arise in non-cirrhotic individuals because of isolated obstruction of the splenic vein, a syndrome known as left-sided portal hypertension (LSPH). LSPH most commonly arises from splenic vein thrombosis and less commonly from splenic vein obstruction due to neighboring mass effect.² LSPH can result in the formation of isolated gastric varices (IGV) due to retrograde drainage through the short gastric and gastroepiploic veins—these varices can potentially result in life-threatening upper gastrointestinal bleeding (UGIB).²,³ We present a case of diffuse large B-cell lymphoma (DLBCL) causing UGIB through the formation of isolated gastric fundal varices (Sarin classification: IGV-1).¹

CASE REPORT

A 59-year-old man with a history of schizophrenia, substance use disorder (alcohol, cocaine, marijuana, and tobacco), hypertension, hypothyroidism, gout, and hepatitis C presented from a long-term inpatient psychiatric hospital with sudden-onset right-sided flank pain. Computed tomography (CT) scan of the abdomen and pelvis revealed a low-attenuating lesion with mild enlargement in the upper pole of the right kidney, 2 low-attenuating lesions in the spleen, and retroperitoneal lymphadenopathy. CT-guided biopsy of the kidney lesion revealed a high-grade DLBCL. Treatment planning was initiated with medical oncology, and the patient agreed to undergo chemotherapy; a whole-body positron emission tomography-CT (PET-CT) scan for staging was scheduled. Abdominal ultrasound with Doppler at this time showed a normal-appearing liver, patent main hepatic arteries, patent hepatic veins, and patent main, right, and left portal veins with hepatopetal flow. There were no ascites, but 2 hypoechoic splenic masses were detected.

One month later, the patient presented to the emergency department after experiencing 3–4 weeks of melena, generalized abdominal pain, nausea, lightheadedness, and malaise that acutely worsened over the past day. Vital signs were significant for tachycardia with a heart rate of 101 beats/min and relative hypotension with a blood pressure of 110/70 from a baseline of 140/90s. Physical examination was significant for skin pallor and epigastric tenderness. Laboratory results revealed a hemoglobin level of 6.4 g/dL (baseline of 13.6 g/dL). The patient was resuscitated with 2 units of packed red blood cells and underwent an emergent
esophagogastroduodenoscopy, which showed IGV-1 with stigmata of recent bleeding and no esophageal varices (Figure 1). The patient was managed conservatively with intravenous octreotide, carvedilol, and pantoprazole. Repeat liver ultrasound with Doppler was performed, which showed a normal-appearing liver with patent splenic vein and hepatic vasculature; no thrombosis was noted. He was discharged 2 days later in a stable condition on propranolol for variceal bleeding prophylaxis. One week later, he underwent the staging PET-CT, which revealed neoplastic involvement of the entire pancreas, gastrohepatic ligament, celiac trunk, perigastric, and splenic hilar regions with highly avid fluorodeoxyglucose uptake and was started on rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) chemotherapy 2 weeks later (Figure 2). After completing 6 cycles of R-CHOP, a postchemotherapy PET-CT (8 months after the initial PET-CT) demonstrated complete remission. At this point, the patient was discharged from the psychiatric hospital and was subsequently lost to follow up. However, he had no further known episodes of UGIB.

DISCUSSION

The most common cause of LSPH is splenic vein thrombosis from pancreatitis or pancreatic malignancies. Since the splenic vein is located directly posterior to the pancreas, other pancreatic diseases such as abscesses and pseudocysts can also involve the splenic vein. However, there are also many nonpancreatic disorders that have been reported to cause splenic vein obstruction, such as surgical procedures, metastatic malignancies, lymphoma, splenic artery aneurysms, and hypercoagulation disorders. Whether splenic vein obstruction occurs internally or externally, the end result is the same with elevated left-sided portal pressures; IGV develop as collateral pathways to decompress this high-pressure system.

Compared with esophageal varices, gastric variceal bleeding generally follows a more severe course in terms of blood loss, length of hospital stays, and mortality. Although IGV-1 has a relatively low prevalence (7%), a high proportion of these patients (73%–78%) present with moderate to massive bleeding. Initial management of gastric variceal bleeding is focused on reducing portal pressures with vasoactive drugs such as octreotide, terlipressin, or somatostatin. Gastric variceal obliteration with cyanoacrylate glue is accepted as the first-line endoscopic treatment for gastric variceal bleeding because other endoscopic interventions such as sclerotherapy and band ligation have fallen out of favor because of lower initial hemostasis and higher rebleeding rates. However, cyanoacrylate glue is not currently approved by the Food and Drug Administration in the United States for this particular indication. In appropriate candidates, interventional radiology procedures such as transjugular intrahepatic portosystemic shunt (TIPS) and balloon retrograde transvenous
oblation (BRTO) have also proven successful at controlling gastric variceal bleeding and are especially effective at preventing rebleeding complications.6 Our institution does not routinely perform gastric variceal obliteration; all cases of gastric variceal bleeding that present to our institution, whether they are actively bleeding, are evaluated for TIPS or BRTO if the patient is not a suitable candidate for TIPS. In cases of massive bleeding, balloon tamponade may be used as a bridging therapy to TIPS or BRTO.

We report a rare case of DLBCL causing IGV-1 in the absence of any appreciable liver disease or evidence of splenic vein thrombosis. Extensive neoplastic involvement of regions that largely overlap with the splenic portion of the portal venous system suggested that mass effect on the splenic vein was the most likely etiology of IGV-1 development. To our knowledge, we have only found 5 case reports published in peer-reviewed journals of lymphoma causing UGIB through IGV.4,10–13 Our patient was managed conservatively with a nonselective β-blocker, vasoactive agent, and proton pump inhibitor. The decision to manage our patient conservatively was multidisciplinary in nature, with active communication between the medical oncology, gastroenterology, and internal medicine teams. Considering that (i) the patient had presented after weeks of chronic melena without frank hematemesis or hematochezia; (ii) the esophagogastroduodenoscopy had revealed no active bleeding; and (iii) the patient had agreed to begin chemotherapy immediately, the decision was made to monitor the patient for several days and then to begin treatment of the underlying condition instead of pursuing more invasive measures such as TIPS or BRTO at the start. As such, our patient successfully completed R-CHOP and had no further known episodes of UGIB.

Financial disclosure: None to report.

Informed consent was obtained for this case report.

REFERENCES

1. Sarin SK, Lahoti D, Saxena SP, Murthy NS, Makkvna UK. Prevalence, classification and natural history of gastric varices: A long-term follow-up study in 568 portal hypertension patients. Hepatology. 1992;16(6):1343–9.
2. Koduk S, Coban S, Yüksel O, Arhan M. Left-sided portal hypertension. Dig Dis Sci. 2007;52(5):1141–9.
3. Pereira P, Peixoto A. Left-sided portal hypertension: A clinical challenge. GE Port J Gastroenterol. 2015;22(6):231–3.
4. Chen BC, Wang HH, Lin YC, Shih YL, Chang WK, Hsieh TY. Isolated gastric variceal bleeding caused by splenic lymphoma-associated splenic vein occlusion. World J Gastroenterol. 2013;19(40):6939–42.
5. Trudeau W, Prindiville T. Endoscopic injection sclerosis in bleeding gastric varices. Gastrointest Endosc. 1986;32(4):264–8.
6. Angeli P, Bernardi M, Villanueva CA, et al. EASL Clinical Practice Guidelines for the management of patients with decompensated cirrhosis. J Hepatol. 2018;69(2):406–60.
7. Little AG, Moossa AR. Gastrointestinal hemorrhage from left-sided portal hypertension: An unappreciated complication of pancreatitis. Am J Surg. 1981;141(1):153–8.
8. Chang CJ, Hou MC, Liao WC, et al. Management of acute gastric varices bleeding. J Chin Med Assoc. 2013;76(10):539–46.
9. Garcia-Pagán JC, Barrufet M, Cardenas A, Escorsell A. Management of gastric varices. Clin Gastroenterol Hepatol. 2014;12(6):919–28.e1.
10. Lenchart A, Fernandez-Castillo J, Mullins K, Salgia R. A rare case of gastric variceal hemorrhage secondary to infiltrative B-cell lymphoma. Case Rep Gastroenterol. 2016;10(3):518–24.
11. Parekh R, Walia S, Zalawadia A, Siddiqui Y. A rare case of splenic lymphoma in a patient with polymyositis manifesting as gastric variceal bleeding. Clin J Gastroenterol. 2013;5(2):92–6.
12. Vida Pérez L, González Galilea A, Fraga Rivas E. Bleeding from gastric varices as the initial manifestation of primary pancreatic lymphoma. Gastroenterol Hepatol. 2010;33(3):165–70.
13. Shih I, Hu W, Wei C, Liu K. Retroperitoneal follicular lymphoma presenting with gastric varices and splenic artery pseudoaneurysm. Ann Hematol. 2013;92(9):1283–4.

DISCLOSURES

Author contributions: All the authors contributed equally to the manuscript. S. Ahlawat is the article guarantor.

Written work prepared by employees of the Federal Government as part of their official duties is, under the U.S. Copyright Act, a “work of the United States Government” for which copyright protection under Title 17 of the United States Code is not available. As such, copyright does not extend to the contributions of employees of the Federal Government.