Gastric Varices from Metastatic Ovarian Cancer with Splenic Involvement

by Allyce Caines, Ghassan Allo, Yousuf Siddiqui

Left-sided portal hypertension (LSPH), also known as splenoportal hypertension, is a rare but life-threatening cause of upper gastrointestinal bleeding. LSPH often occurs in non-cirrhotic patients as a consequence of splenic vein obstruction. We present a case of isolated gastric varices due to mass effect on the splenic vein and likely tumor thrombus due to metastatic ovarian cancer.

INTRODUCTION

Portal hypertension refers to increased pressures in the hepatic portal system. This in turn leads to complications such as variceal hemorrhage from gastric or esophageal varices, ascites and hepatic encephalopathy. Upper gastrointestinal bleeding secondary to portal hypertension in the form of variceal hemorrhage is a recognized life-threatening cause of gastrointestinal bleeding. In patients with gastrointestinal bleeding due to portal hypertension, bleeding from gastric varices is the cause in 5%-10% of patients.[1] Increased resistance to portal flow due to a stiff, cirrhotic liver is often the cause of portal hypertension. However, portal hypertension can also be caused by isolated obstruction of the splenic vein, which is often referred to left sided portal hypertension (LSPH). LSPH accounts for less than 5% of all patient with portal hypertension[1]. Most of the reported cases of splenic vein obstruction have been the result of malignancy involving the spleen. Pancreatic disorders, including pancreatic carcinoma, acute and chronic pancreatitis, cysts, and pseudocysts which may block the splenic vein via thrombus formation or mass effect, are the most common causes of LSPH. These patients often have no known prior liver disease and have no evidence of cirrhosis on presentation. Bleeding can be catastrophic due to high portal pressures. In these cases, it is important to consider causes of left sided portal hypertension and the available treatment options.

Case Report

A 59 year-old female with a past medical history of metastatic ovarian cancer and obstructive uropathy, secondary to extrinsic compression, presented to her primary gynecologic oncologist with complaints of fatigue, lightheadedness and melena for one week. On laboratory evaluation, she was found to have a hemoglobin of 5.7 g/dL prompting emergency evaluation. Four years prior she was diagnosed with stage IIIC serous carcinoma of the ovary, at which time she underwent total abdominal hysterectomy, omentectomy, pelvic lymphadenectomy, tumor debulking and subtotal colectomy with diverting ileostomy followed

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by chemotherapy. She remained disease free for approximately nine months when imaging showed disease recurrence. She developed significant disease progression involving the splenic hilum, retroperitoneal lymph nodes and pelvis in addition to bilateral hydronephrosis.

On presentation to the emergency department she was hemodynamically stable with unremarkable physical exam. An episode of hematemesis consisting of 500cc of bright red blood occurred in the emergency department with repeat hemoglobin of 4.5 g/dL. Endoscopy revealed type 1 isolated gastric varices (IGV1). She received a total of 6 units of packed red blood cells. The patient was then transferred for consideration of balloon retrograde transvenous obliteration (BRTO). Repeat endoscopy confirmed the findings of type 1 isolated gastric varices with red wale signs (Figure 2).

BRTO, splenectomy, gastric vessel ligation and cyanoacrylate injection were discussed as potential therapeutic options. The patient had no further episodes of bleeding and her hemoglobin remained stable. Magnetic resonance imaging (MRI) revealed a hypovascular mass in the splenic hilum with minimal central enhancement concerning for metastatic disease. Areas of hypovascular nodularity around the lesion extending into part of the splenic vein at the hilum and branches were suggestive of tumor thrombus. Liver lesions concerning for metastatic disease were also present. On MRI the hepatic vein, celiac artery, hepatic

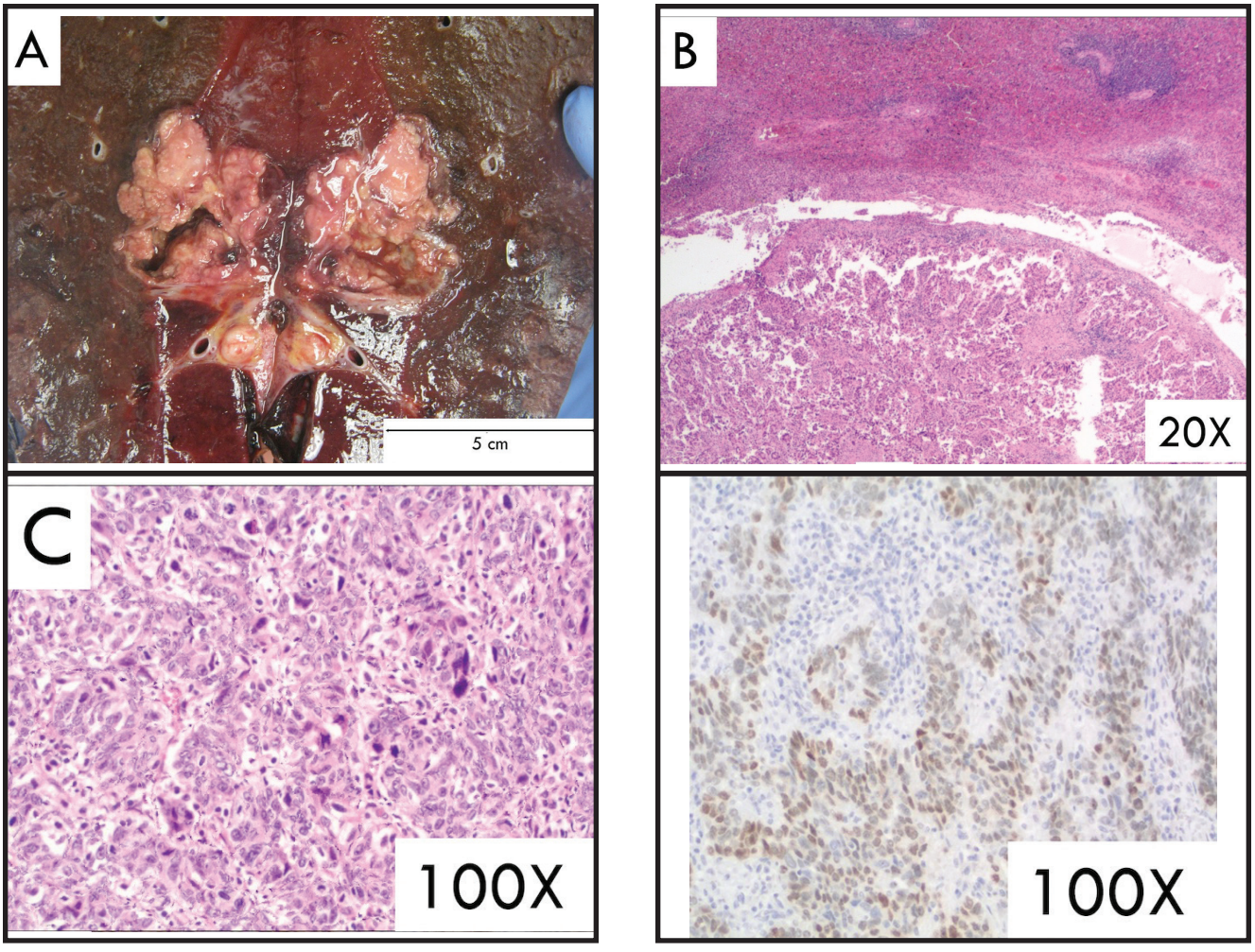

Figure 1. A. Tumor within splenic hilum. Inset 5 cm B. Poorly differentiated carcinoma within spleen. Hematoxylin and Eosin. 20X C. Poorly differentiated carcinoma within spleen. Hematoxylin and Eosin. 100X D. Estrogen receptor (ER) immunohistochemistry. 100X

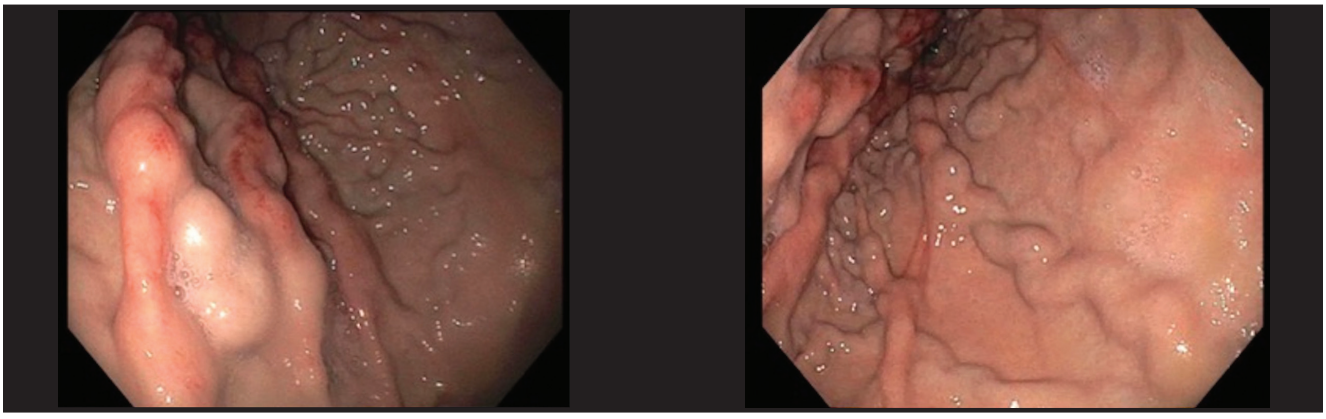

Figure 2. Type 1 isolated gastric varices with red wale signs

artery, portal vein and superior mesenteric vein were patent. The splenic artery appeared tortuous and the splenic vein was engorged.

The patient was not a candidate for BRTO due to the lack of portosystemic collaterals. She underwent splenic artery embolization to decrease inflow to the spleen and splenectomy the following day. Pathology revealed a 4.4 x 3.7 x 3.1 cm ill-defined mass located within the splenic hilum (Figure 1A), which consisted of poorly differentiated adenocarcinoma (Figure 1B-1C). Immunohistochemically, the adenocarcinoma was positive for cytokeratin AE1/AE3, CK7, PAX-8 and estrogen receptor (Figure 1D). The adenocarcinoma was negative for CK20. The morphology and immunohistochemistry were consistent with metastatic adenocarcinoma of müllerian origin, from the patient's known ovarian adenocarcinoma.

Discussion

LSPH, also known as splenoportal or sinistral hypertension is a rare, but life-threatening cause of upper gastrointestinal bleeding.[2] LSPH often occurs in non-cirrhotic patients as a consequence of splenic vein obstruction. Pancreatic disorders, including pancreatic carcinoma, acute and chronic pancreatitis, cysts, and pseudocysts which may block the splenic vein via thrombus formation or mass effect, are the most common causes of LSPH.[3] To our knowledge there is only one other case of LSPH with bleeding gastric varices secondary to metastatic ovarian cancer published by Wallace et al in 2004.[4]

In our case, mass effect on the splenic vein as well as likely tumor thrombus caused splenic venous outflow obstruction. Obstruction of the splenic vein results in venous hypertension in collateral pathways that carry splenic arterial blood via the short gastric, coronary, and gastroepiploic veins to the superior mesenteric and portal veins. In the gastric wall veins of the fundus, blood flow and pressure increase, and submucosal structures consequently dilate, producing gastric varices.[4,3] Risk factors for gastric variceal hemorrhage include the size of fundal varices (as there is a linear relationship between size of varices and risk of variceal hemorrhage), endoscopic presence of variceal red spots, and Child-Pugh class in patients with cirrhosis.[5] Gastric varices are less frequent than esophageal varices and are present in 5%-33% of patients with portal hypertension. The reported incidence of bleeding from gastric varices is approximately 25% in two years, with a higher bleeding incidence for fundal varices.[6] Compared to esophageal varices, gastric varices are larger, more extensive, and lie deeper in the submucosa. As a result, standard endoscopic treatments for esophageal varices, including band ligation and sclerotherapy are largely ineffective for gastric varices.[7]

Management of gastric varices is dependent on the etiology, presence of collaterals and the available treatment modalities. LSPH, for example, has distinct therapeutic management options that are not appropriate for the management of generalized portal hypertension**.** Currently the gold standard for treatment of fundal (IGV1) varices as a result of splenic vein thrombosis is splenectomy. Surgical

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removal of the spleen decreases venous outflow through collateral circulations and decompresses IGV to prevent future bleeding.[8] Additional therapies have been used to control gastric variceal bleeding and prevent re-bleeding from occurring. These include band ligation and endoscopic sclerotherapy (frequently by cyanoacrylate glue injection). The American Association for the Study of Liver Diseases (AASLD) Society guidelines recommend endoscopic variceal sclerotherapy in patients who bleed from gastric fundal varices otherwise when available, endoscopic variceal ligation is an option. AASLD also recommends that transjugular intrahepatic portosystemic shunt (TIPS) should be considered in patients in whom hemorrhage from fundal varices cannot be controlled or in whom bleeding recurs despite combined pharmacological and endoscopic therapy.[5]

Left-sided portal hypertension should be considered in the presence of gastrointestinal bleeding with normal liver function and unexplained splenomegaly.[1] Isolated gastric varices type 1 should immediately raise the clinician's suspicion for splenic vein obstruction. Although a rare cause, in patients with prior malignancy or without evidence of pancreatic pathology, malignancy should remain on the differential as a cause of splenic vein obstruction. ■

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