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Case Report

Atypical Presentation of Isolated Gastric Variceal Bleeding Requiring Multidisciplinary Management

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Abstract

Variceal bleeding is the most lethal manifestation of portal hypertension, most commonly due to esophageal varices in the setting of liver cirrhosis. Isolated gastric varices (IGV) are a rare cause of upper gastrointestinal bleeding, often of pancreatic origin. In this case we present a complex patient with a history of remote splenic injury leading to portopulmonary hypertension and the development of refractory bleeding due to IGV. We discuss the extensive multidisciplinary approach taken to provide comprehensive care and to control bleeding, which included endoscopic (sclerotherapy), radiologic (balloon-occluded antegrade transvenous obliteration [BATO] and splenic artery embolization) and surgical interventions (splenectomy).

Keywords: gastric varices; non-cirrhotic portal hypertension; splenic injury

Introduction:

IGV are observed in up to 5% of patients with liver cirrhosis, and up to 10% of patients with non-cirrhotic portal hypertension (NCPH) [1,2]. If a branch of the portal venous system becomes obstructed, such varices can form in order to decompress the segment [2]. Although gastric varices are 50% less likely to bleed compared to oroesophageal varices, bleeding from gastric varices is more severe [1,3]. Studies have shown that gastric variceal bleeding have a significantly higher mean blood transfusion requirement [1]. Gastric variceal bleeds also portend a higher mortality risk than esophageal varices with some studies reporting mortality as high as 45% [1,4]. Management of initial bleeding of IGV presents a challenging problem since information about the etiology may not be available at the time of initial presentation. While cirrhosis is the most common cause of portal hypertension, there are numerous causes of NCPH which can be classified by the location of resistance to blood flow into prehepatic, hepatic, and post hepatic groups [5]. We present a rare case of refractory bleeding from IGV in a patient with NCPH due to remote splenic injury.

Case Presentation:

A 40-year-old woman with a history of remote splenic injury due to a motor vehicle accident during infancy was admitted with one episode of large hematemesis. On arrival, vital signs were unstable (blood pressure to 80/50 mmHg and tachycardia of 130/min) and initial hemoglobin was

11.7 g/dl. She was resuscitated with normal saline and started on intravenous pantoprazole. She was intubated and transferred to the intensive care unit for an urgent esophagogastroduodenoscopy (EGD), which demonstrated IGV with a large nipple sign and no active bleeding [Figure 1].

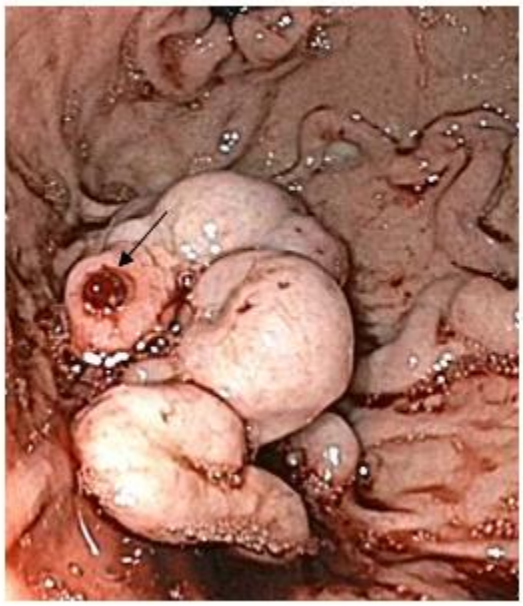

Figure 1: Isolated Gastric Varix in the Fundus with nipple sign (black arrow) from recent bleed

No endoscopic intervention was performed given the unknown etiology of IGV and inactive bleed. Abdominal CT scan revealed splenomegaly and confirmed prominent gastric varices without a discrete gastro-renal shunt; there was no evidence of splenic vein stricture or thrombosis on imaging [Figure 2].

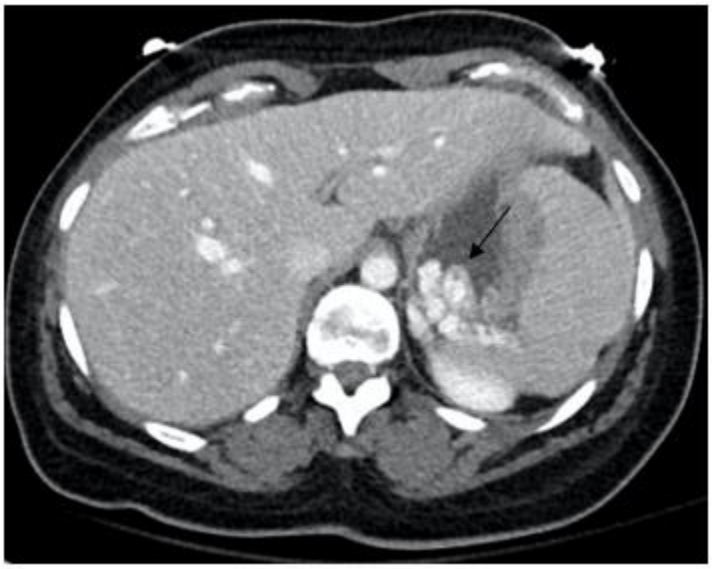

Figure 2: Contrast enhanced axial image confirming the presence of dilated gastric varicosities (black arrow) protruding into the gastric lumen.

Transthoracic echocardiogram (TTE) showed moderate diastolic dysfunction and was suggestive of pulmonary hypertension with elevated right ventricular systolic pressure (RVSP). Percutaneous transhepatic obliteration of IGV with coil embolization was performed by Interventional Radiology [Figure 3]. On post-op day one, a repeat episode of hematemesis prompted follow-up EGD and sclerotherapy with N-Butyl Cyanoacrylate (NBC). She was discharged home four days later.

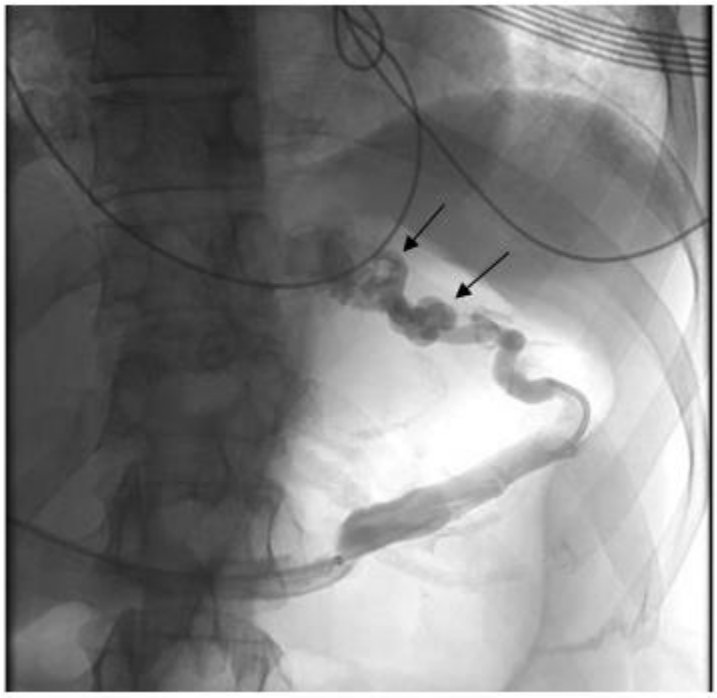

Figure 3: Fluoroscopic image of percutaneous transhepatic cannulation and venography of the gastric varices (black arrows) from the splenic vein prior to embolization.

One day after discharge on post-op day six, the patient again presented to our facility after another episode of hematemesis. After initial resuscitation, right heart catheterization confirmed pulmonary hypertension with a pulmonary artery (PA) systolic pressure of 67 mmHg (normal 15-25 mmHg), mean PA pressure of 41 mmHg (normal <24 mmHg), PA wedge pressure of 13 mmHg (normal <14 mmHg) and right atrial pressure of 12 mmHg. Fick cardiac output was measured at 8.53 L/min and cardiac index was measured at 4.59 L/min/m2. A repeat EGD and NBC sclerotherapy of IGV was again performed. At this point, our assessment was NCPH; confirmatory transjugular liver biopsy was then performed revealing minimal fibrosis. Her hepatic venous pressure gradient was normal at 5 mmHg, reflecting that a transjugular intrahepatic portosystemic shunt (TIPS) would not provide benefit.

Ultimately, the patient underwent partial splenic artery embolization followed by total splenectomy as a definitive therapy. She tolerated splenectomy well. On post-surgical inspection, her spleen measured 19.4 cm (normal: 11 cm) in the greatest dimension and weighed 777 grams (normal is up to 150 grams). For her pulmonary hypertension she was started on epoprostenol infusion and then transitioned to oral riociguat and macitentan. Two months later, she was doing well with no further episodes of bleeding.

Discussion:

We present, to our knowledge, the first case of splenic injury resulting in bleeding IGV from NCPH, a case which was a diagnostic puzzle in which the patient experienced four life-threatening bleeding episodes before curative multidisciplinary management.

In recent years better understanding of the physiology of the portal venous system has improved management of bleeding IGV. Studies have demonstrated that almost half of IGV are secondary to abnormalities of the pancreas [6]. Classically, patients have imaging suggestive of splenomegaly and/or splenic vein thrombosis. Lin et al describe a patient with GV bleeding due to splenic vein occlusion arising from abdominal lymphadenopathy [7]. Similarly, mass effect from lymphomas, abscesses, or tumors of the pancreas, stomach, colon or kidney may cause splenic vein stenosis or thrombosis with resultant GV bleeding [2, 8]. Another case has described IGV resulting from abdominal tuberculosis [9].

Sclerotherapy of IGV with NBC is the optimal endoscopic intervention, supported by evidence-based guidelines, and routinely performed, though not approved for this purpose in the United States. Radiological interventions for managing IGV include TIPS, balloon-assisted antegrade or retrograde transvenous obliteration (BATO or BRTO), and partial splenic artery embolization [10]. For patients with IGV bleeds related to portal hypertension, TIPS directly reduces portal pressures, portending significant survival benefit [10]. BATO/BRTO is indicated for those in whom TIPS fails or is contraindicated. Transvenous obliteration of IGV entails systemic venous access with subsequent injection of a sclerosing agent and/or embolization of the culprit varix. Blood flow is diverted toward the portal circulation, and unlike a TIPS procedure, does not reduce portal pressures [10,11]. Alternative therapies for patients with IGV include partial splenic artery embolization or splenectomy to mitigate portal hypertension and its associated physiological effects. Partial splenic artery embolization is a safe and effective adjunctive consideration to address bleeding IGV, particularly in patients who cannot immediately undergo splenectomy or may not be good surgical candidates [9]. Surgical splenectomy can be pursued for more definitive therapy.

Our patient had large IGV, a normal pancreas, minimal hepatic fibrosis, a normal HVPG and pulmonary hypertension. Her marked splenomegaly from a remote splenic injury led to the development of presinusoidal portal hypertension followed by pulmonary hypertension and ultimately, refractory IGV bleeding. Portopulmonary hypertension is a well-established sequela of cirrhosis; however, it was recently described in patients with non-cirrhotic portal hypertension (NCPH) as in our case [12]. The underlying pathogenesis remains largely obscure. In patients with NCPH, vasoactive substances such as endothelins have been hypothesized to result in vascular remodeling, leading to increased pulmonary pressures. Decreased production of vasodilators, including nitric oxide and prostaglandins, will contribute to elevated pulmonary pressures as well. Another proposed mechanism is that established

portosystemic shunts help to deliver these vasoactive substances directly into the pulmonary circulation, expediting pathologic changes. Portopulmonary hypertension leads to stress on the right ventricle, and prognosis is highly correlated with the degree of subsequent right ventricular dysfunction [12].

In order to serve our patient, the expertise of numerous disciplines was required. This unique case illustrates the challenges of managing gastric varices in a patient without cirrhosis caused by remote splenic injury and the need for a multidisciplinary approach to ensure the best outcome [13].

Conclusion:

In conclusion, we present the unique case of a patient whose remote splenic injury led to the development of NCPH followed by pulmonary hypertension and refractory bleeding from IGV. While most IGV are caused by pancreatic abnormalities, there are multiple other etiologies including malignancy, abscess, and trauma leading to splenic vein stenosis or thrombosis. While sclerotherapy of IGV with NBC is considered optimal endoscopic therapy, there are variable options for refractory IGV including radiologic and surgical interventions. For such an unusual case of IGV without cirrhosis, we therefore recommend a multidisciplinary approach to patient care.

Conflicts of interest: Our authors have no conflicts of interest to disclose.

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