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CASE REPORT | ESOPHAGUS

Downhill Esophageal Varices Associated With Central Venous Catheter-Related Thrombosis Managed With Endoscopic and Surgical Therapy

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ABSTRACT

Downhill esophageal varices are a rare cause of upper gastrointestinal hemorrhage. We present a case of downhill variceal bleeding due to superior vena cava thrombosis resulting from a prior central venous catheter. The patient was managed with endoscopic band ligation and later with surgical axillary vein to right atrium bypass grafting. Successful long-term resolution of varices was achieved at 1 year of follow-up. This is the longest follow-up described for combined endoscopic and surgical management in the existing literature for catheter-associated downhill varices.

INTRODUCTION

Downhill varices of the esophagus have been described as a rare cause of upper gastrointestinal tract hemorrhage. These varices are generally related to occlusion of the superior vena cava (SVC), which leads to blood flowing "downhill" through the azygous vein and esophageal venous plexuses. The occlusion may result from extrinsic compression of the SVC by mass effect or intrinsic stenosis such as from venous thrombosis.[1] The first reported case of varices from hemodialysis catheter-related thrombosis was reported in 1998.[2]

CASE REPORT

A 32-year-old woman presented with 2 days of persistent melena and 2 episodes of hematemesis. There was no red blood in the stool. There was associated subjective weakness without syncope. Her past medical history was significant for end-stage renal disease due to mesangial proliferative glomerulonephritis requiring hemodialysis, known SVC thrombosis from prior central venous catheters, and mixed connective tissue disorders. Angioplasty had previously failed to recanalize the SVC. The patient was taking warfarin for recurrent arteriovenous graft thromboses. There was no history of abdominal pain, antiinflammatory medication, or alcohol use.

Physical examination was significant for tachycardia to 114 beats per minute, dilated superficial veins on the anterior chest wall, and melenic stool. There were no orthostatic vital sign changes, scleral icterus, abdominal distention, or hepatomegaly. The patient was transferred to the medical intensive care unit. Laboratory studies were as follows: hemoglobin (Hb) 6.0 g/dL (baseline 10.0 g/dL), blood urea nitrogen (BUN) 49 mg/dL, creatinine 11.2 mg/dL, bilirubin 0.7 µmol/L, aspartate aminotransferase (AST) 7 U/L, alanine aminotransferase (ALT) 6 U/L, international normalized ratio (INR) 1.1, and albumin 3.3 g/dL.

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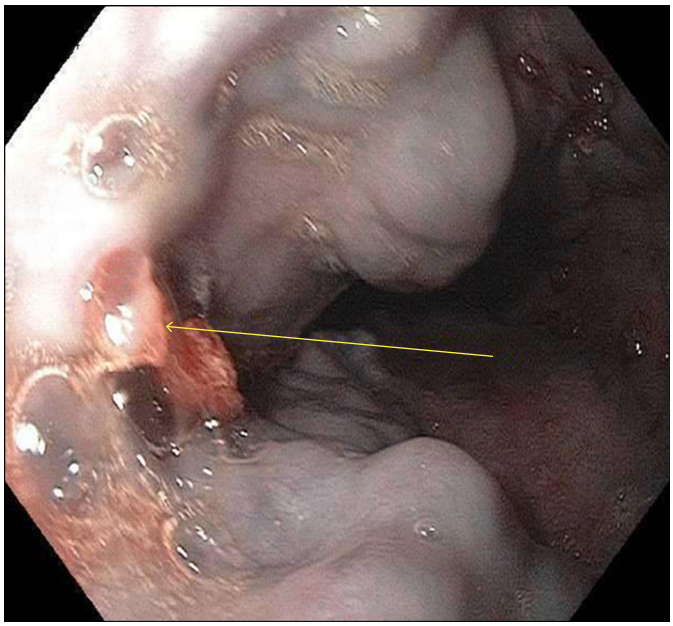

Figure 1. Esophagoscopy of the midesophagus demonstrating Grade III varices and red wale sign/hematocystic spot (arrow).

She received transfusion of 2 units of packed red blood cells. The tachycardia resolved. Esophagogastroduodenoscopy demonstrated Grade I/II esophageal varices in the upper esophagus and Grade III esophageal varices in the midesophagus with the presence of red wale sign and white nipple sign (Figure 1). There were no varices seen in the distal esophagus. Fresh blood was found in the gastric cardia and fundus. Endoscopic band ligation of a proximal varix was performed. The melena resolved, and the hemoglobin level remained stable. Abdominal ultrasound with Doppler demonstrated a normal liver with patent portal and hepatic veins. Thoracic and abdominal computed tomographic angiography demonstrated numerous abdominal and chest wall collateral vessels, occlusion of the SVC at its junction with the right brachiocephalic vein, and a prominent azygous vein (Figure 2). The diagnosis was downhill esophageal varices secondary to SVC thrombosis due to multiple prior central venous catheters.

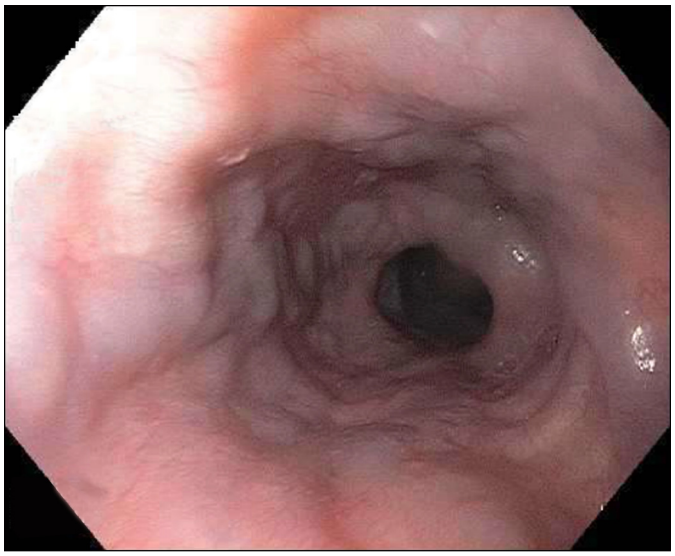

Figure 3. Esophagoscopy at 12 months after operation demonstrating Grade I varices with no signs of bleeding in upper- and midesophagus.

The patient underwent repeat esophagogastroduodenoscopy 1 month later demonstrating improvement of the varices, which were Grade I. At that time, the patient underwent right axillary vein to right atrium appendage bypass grafting. At this time, it was felt safe to resume anticoagulation. Follow-up endoscopy was performed at 1 year post operation, which demonstrated Grade I varices and no signs of bleeding (Figure 3).

DISCUSSION

Downhill esophageal varices are a rare cause of bleeding. Occlusion of the SVC results in increased pressure to the

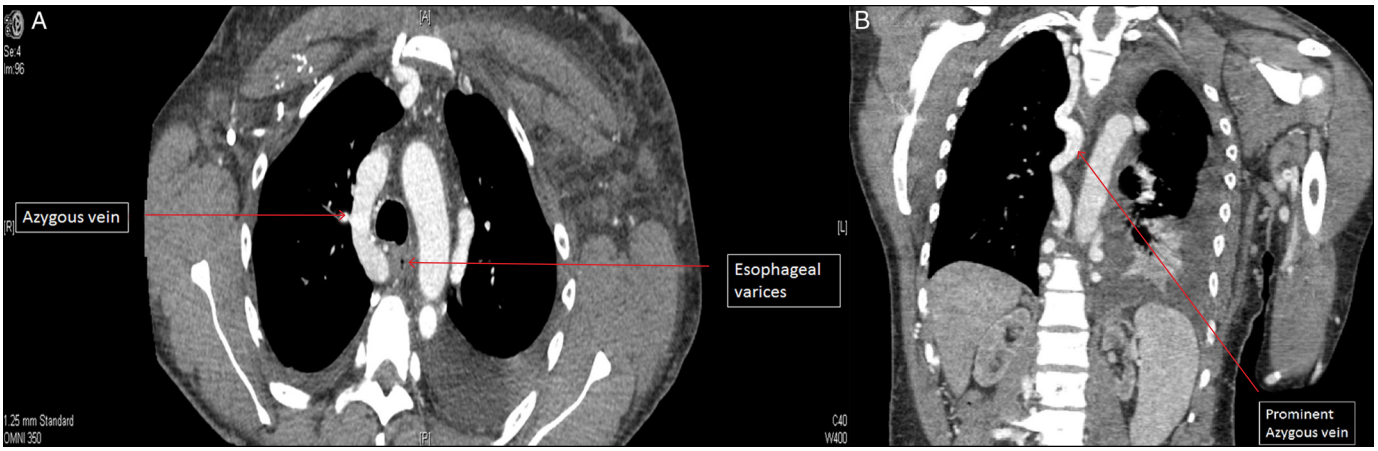

Figure 2. Computed tomographic angiogram of the chest demonstrating (A) dilated azygous vein and midesophageal varices and (B) dilated azygous vein and chest wall collateral vessels.

azygous and hemiazygous systems responsible for drainage of the upper two-thirds of the esophagus. This leads to varix formation. Varices due to intrinsic narrowing of the SVC from thrombosis have been described, such as in the setting of hemodialysis catheters and Behcet's disease.[3] Extrinsic compression has been described from intrathoracic goiters[4] as well as thymoma[5] and lung cancer.[6] Our patient's occlusion was proximal to the azygous vein-SVC junction with varices involving only the upper two-thirds of the esophagus. Occlusion distal to the azygous vein's drainage results in varices throughout the esophagus.[2,7]

Treatment of downhill varices focuses on restoring venous drainage. Options include balloon angioplasty,[8] SVC stenting,[9] and open surgical therapies. In the present case, angioplasty had been unsuccessful in the past at treating the SVC stenosis, and therefore the more invasive surgical option was necessary. Endoscopic treatment options include banding, with recent emphasis placed on banding proximally.[10] Sclerotherapy is avoided due to concerns over embolization and spinal cord infarction.[11] In general, endoscopic surveillance is recommended; however, the optimal interval is unknown.[12]

DISCLOSURES

Author contributions: JC Berkowitz, S. Bhusal, and S. Inamdar drafted the manuscript. D. Desai and MA Cerulli critically revised the manuscript. S. Inamdar is the article guarantor.

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Informed consent was obtained for this case report.

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