When Going With the Flow Sends You Upstream

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Question: A 55-year-old man with decompensated nonalcoholic steatohepatitis cirrhosis complicated by portal hypertension and ascites underwent routine upper gastrointestinal endoscopy examination for follow-up of previously documented grade 1 esophageal varices. He also had metabolic syndrome (severe obesity, type 2 diabetes mellitus with peripheral neuropathy, and hypercholesterolemia), chronic anemia secondary to portal hypertensive gastropathy, lung atelectasia, hypothyroidism, and hypogonadism owing to primary gonadal failure. Physical examination revealed large-volume ascites and marked peripheral edema. His bloods showed pancytopenia (platelets, 17 \times 10^9/L; red blood cells, 2.12 \times 10^{12}/L; white blood cells, 2.24 \times 10^9/L), anemia (hemoglobin, 60 g/L), International Normalized Ratio of 1.5, preserved liver function, and elevated creatinine (144 \text{ m}\text{mol}/L) and C-reactive protein (30 mg/L).

The upper gastrointestinal endoscopy showed a lesion of multiple small vessels and small column varices in the upper esophagus (less than 25 cm from the incisor teeth), which were not suitable for banding (Figure A). The distal esophagus was devoid of varices and there was moderate portal hypertensive gastropathy. To investigate the origin of the varices described in the proximal esophagus, the patient underwent a computed tomography scan with contrast, which showed an abnormal soft tissue mass in the posterior mediastinum, not clearly characterized owing to the phase of contrast enhancement (Figure B, C). Ascites, an enlarged spleen, multiple collateral vessels, patent portal, splenic and superior mesenteric veins were also described. Given the patient’s renal impairment, in his best interests, a repeat contrast scan was avoided.

Based on the clinical history and radiographic abnormalities, what is the most likely diagnosis?

Look on page 22 for the answer and see the Gastroenterology website (www.gastrojournal.org) for more information on submitting your favorite image to Clinical Challenges and images in GI.

Conflicts of interest
The authors disclose no conflicts.
Contrast-enhanced computed tomography images performed 3 years earlier revealed that the soft tissue mediastinal mass was owing to multiple enlarged collateral vessels, extending from the carina to the gastroesophageal junction. The azygos vein was also dilated (Figure D).

In cirrhosis, esophageal varices form as a result of increased intrahepatic resistance, portal venous hyperemia, and portal venous shunting. This “classical” evolution of varices is most notable in the distal esophagus but can occur in the stomach, duodenum and distal gastrointestinal tract. In contrast, so-called “downhill varices” (DV) form owing to obstruction of blood flow from the superior vena cava into the right atrium and are frequently due to superior vena cava obstruction, as a result of thrombosis or direct compression. Causes include thoracic tumors, giant intrathoracic goiter, and Castleman’s disease.1 In this case, superior vena cava compression was due to mediastinal collateral vessels owing to portal hypertension. Occurrence of DV is also reported as a rare complication of venous access for hemodialysis or pacemaker implants.2 The location of the obstruction determines the extent of the DV, usually localized to the upper esophagus, when the obstruction is proximal to the azygos vein, but may involve the whole esophagus, in cases of obstruction distal to the azygos vein.2 DV account for 0.4% to 11.0% of esophageal varices, and have a much lower risk of bleeding (9%–16% of cases) than seen in classical esophageal varices.3 Treatment of the underlying medical condition is sufficient in some cases to resolve the varices. Despite its relative rarity, it is important for clinicians to be aware of this entity, to promptly investigate the potential causes and offer the patient a personalized etiology-driven treatment.

Given the small size of our patient’s varices and his ongoing therapy with carvedilol, he did not receive any additional interventions. Sadly, he succumbed some weeks later to severe sepsis and subsequent multiorgan failure.

References

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