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When going with the flow sends you upstream

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Francesca Saffioti: acquisition of data, manuscript concept and design, drafting of the manuscript. Rajeshwar P. Mookerjee: critical revision of the manuscript for important intellectual content.

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QUESTION:

A 55-year-old man with decompensated NASH cirrhosis complicated by portal hypertension (PH) and ascites underwent routine upper gastrointestinal endoscopy (OGD), 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 due to portal hypertensive gastropathy, lung atelectasia, hypothyroidism and hypogonadism due to primary gonadal failure. Physical examination, revealed large volume ascites and marked peripheral edema. His bloods showed pancytopenia (platelets: 17x10^9/L, red blood cells: 2.12 x10^12/L, white blood cells: 2.24 x10^/L), anemia (hemoglobin 60 gL), INR 1.5, preserved liver function and elevated creatinine (144 umol/L) and C-reactive protein (30 mg/L).

The OGD showed a “leish” of multiple small vessels in the upper esophagus and small columns varices, above 25 cm, which were not suitable for banding (**Figure A**). The distal esophagus was devoid of varices and there was moderate portal hypertensive gastropathy. In order to investigate the origin of the varices described in the proximal esophagus, the patient underwent a CT scan with contrast, which showed an abnormal soft tissue mass in the posterior mediastinum, not clearly characterized due to the phase of contrast enhancement (**Figures B and 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 clinical history and radiographic abnormalities, what is the most likely diagnosis?

ANSWER: *Downhill varices* caused by portal hypertension.

Contrast CT images performed 3 years earlier revealed the soft tissue mediastinal mass was due to multiple enlarged collateral vessels, extending from the carina to the gastro-esophageal junction. The azygos vein (AV) 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 due to obstruction of blood flow from the superior vena cava (SVC) into the right atrium, and are frequently due to SVC obstruction as a result of thrombosis or direct compression. Causes include: thoracic tumours, giant intrathoracic goiter or Castleman's disease.[1] In this case, SVC compression was due to mediastinal collateral vessels due to PH. Occurrence of DV are 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 AV, but may involve the whole esophagus, in cases of obstruction distal to the AV.[2] DV account for 0.4–11% 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 that clinicians are aware of this entity in order to promptly investigate the potential causes and offer the patient a personalized etiology-driven treatment.

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

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