“Downhill” varices. A rare cause of esophageal hemorrhage

M. Areia, J. M. Romãozinho, M. Ferreira, P. Amaro and D. Freitas

Department of Gastroenterology. Coimbra University Hospital. Coimbra, Portugal

ABSTRACT

“Downhill” varices or upper esophageal varices are a rare cause of proximal digestive tract hemorrhage with only 16 cases described in the literature. In our series, hemorrhage due to “downhill” varices represents 0.1% of all acute esophageal variceal bleeding. Their etiology differs from that of the usual “up-hill” varices secondary to portal hypertension, and the clinical management should be directed to vascular obstruction if present. We report a case of an 89-year-old male with hemorrhagic “downhill” varices not associated, as usually, with superior vena cava obstruction or compression, but with severe pulmonary hypertension and drug-related hemorrhagic risk factors, whose removal proved sufficient to prevent rebleeding.

Key words: Esophageal and gastric varices. Gastrointestinal hemorrhage. Pulmonary hypertension.

INTRODUCTION

Esophageal varices can be associated with clinical conditions other than portal hypertension. “Downhill” varices or upper esophageal varices were first reported in cases of superior vena cava obstruction generally attributed to mediastinal tumors or masses. Most cases are diagnosed by upper digestive endoscopy as performed for complaints other than hemorrhage, with only 16 cases of hemorrhagic “downhill” varices reported in the literature (1). We describe a case of a patient with hemorrhagic “downhill” varices limited to the upper esophagus, with no mediastinal condition causing venous obstruction. Instead, a severe pulmonary hypertension associated with hemorrhagic risk factors (warfarin and NSAIDs) may explain the clinical presentation.

CASE REPORT

An 89-year-old male was admitted to our hospital with a 24-hour-standing melena. He was prostrated, pale and hypotensive (100/50 mmHg), with a heart rate of 100 per minute. No other abnormalities were present on physical examination. A nasogastric tube was inserted, and the presence of fresh blood in the gastric cavity was confirmed. Laboratory values at admission included: hemoglobin of 8.8 g/dL, hematocrit of 26%, and prothrombin time of 52.0 seconds. After a transfusion with two units of packed red blood cells and two units of fresh frozen plasma hemodynamic stabilization was achieved, and an upper digestive endoscopy was performed.

The endoscopic examination (Fig. 1) revealed grade-two esophageal varices just below the upper esophageal sphincter, with two red spots that extended to the mid esophagus in a grade-one score. No active bleeding was present but the exclusion of other hemorrhagic lesions suggested that the red spots might have been the origin of hemorrhage.

Therefore, the patient was admitted to our Intensive Care Unit with a diagnosis of hemorrhagic “downhill” varices. An abdominal ultrasonography with hepatic Doppler excluded the presence of portal hypertension, and
a dynamic computed tomography scanning of the neck, chest and abdomen showed no mass, lesion or vascular abnormality to suggest arterial or venous compression.

Echocardiography (Fig. 2) was repeated and confirmed a severe pulmonary hypertension, a right atrium/ventricle gradient of 64 mmHg, dilated right cardiac cavities, moderate tricuspid regurgitation, and severe aortic fibrosis without stenosis.

The patient was managed conservatively and switched from warfarin to low-molecular-weight heparin, and NSAIDs were discontinued. No recurrent bleeding was observed during immediate follow-up, and a second-look endoscopy 48 hours later also revealed no active bleeding. Finally, there was no rebleeding during the first three months after patient discharge from hospital.

DISCUSSION

In our thirteen-year series on severe acute upper digestive hemorrhage (n = 2,368), “downhill” varices contribute only 0.1% (1/908) of all esophageal variceal bleeding (2).

The normal venous drainage of the esophagus occurs predominantly by the azygous and hemi-azygous systems. Esophageal veins are usually not visible on upper endoscopy unless some obstructive pathologic event leads to their dilatation and the formation of varices that serve as collateral channels between the portal and azygous systems. “Downhill” varices or upper esophageal varices will develop when an obstruction in the superior vena cava flow forces the blood back into the right atrium using collateral mediastinal pathways to the inferior vena cava, causing a retrograde direction of blood flow, as opposed to the usual “uphill” varices, which develop when portal hypertension forces blood into the inferior vena cava system (3). The first report of this kind of esophageal varices was made in 1964 by Felson and Lessure (4) in a patient with a superior vena cava obstruction secondary to fibrous mediastinitis. To date, more than 100 cases have been reported with various etiologies described (5).

The diagnosis of “downhill” esophageal varices includes lung and thyroid carcinoma, chronic mediastinal fibrosis, metastatic carcinoma, mediastinal mass of unknown origin, venulitis, and complications in the placement of central hemodialysis catheters or the surgical ligation of the superior vena cava (11,12).

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The diagnosis of “downhill” varices is reached at endoscopy, generally in non hemorrhagic patients, and should be recognized as a separate entity from portal hypertension varices, since both the etiology and clinical management are very different. In fact, as already emphasized, “downhill” varices bleed very rarely when compared to “uphill” varices. One possible explanation is that portal hypertension varices are usually associated with coagulation abnormalities and more prone to erosions caused by esophagogastric reflux (13). In the reported case both risk factors were present as the patient was medicated with warfarin (causing a coagulation anomaly) and NSAIDs (which could lead to mucosal erosions).
Treatment should be directed to the underlying cause of vascular obstruction (14), since this is the only approach that will cure the clinical syndrome and also prevent its recurrence. Chemotherapy and radiotherapy have been used for malignant causes, and the angiographic placement of expandable metal stents has proved effective in resolving vascular obstruction (15-17). In surgical candidates a vascular bypass of the superior vena cava can be attempted (3). When all the above measures fail to resolve the obstruction, as in patients with no superior vena cava obstruction, which is the present case, only palliative measures can be applied (3).

Endoscopic therapy, either by ligation or injection sclerotherapy, is possible in “downhill” varices, but the associated complications can be higher in the upper esophagus when compared to portal hypertension lower varices. Indeed, the risk of hemorrhage or perforation seems higher on account of the weakness of the proximal esophageal posterior wall and overall lack of serosa (18, 19). Other complications have also been reported, including a case of vertebral infarction, possibly due to a retrograde flow of the sclerosing agent via the ayzygous vein into the vertebral vasculature (20), and a situation of cardiovascular failure due to massive foreign-body pulmonary embolism following an endoscopic injection with polidocanol and cyanoacrylate (21). Since the effectiveness of the available endoscopic techniques is unpredictable because of a lack of experience related to the rarity of these clinical cases, it seems wise to reserve the endoscopic treatment of “downhill” varices only for extreme situations.

In the present case, a non-aggressive therapeutic approach related to the non-existence of vascular obstruction and consisting only of the removal of hemorrhagic risk factors (warfarin and NSAIDs) proved a very effective option.

REFERENCES