An unusual cause of hematemesis: Goiter

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Abstract

Downhill varices are located in the upper part of the esophagus and are usually related to superior vena cava obstruction. Bleeding from these varices is extremely rare. We describe a 77-year-old patient with hematemesis due to downhill varices as a result of recurrent goiter. A right lobe thyroidectomy was carried out with disappearance of the varices.

INTRODUCTION

Bleeding from distal esophageal varices is a frequent complication of portal hypertension. The portal blood drains into the superior vena cava by gastric and esophageal collaterals, also called uphill varices, referring to the upward direction of blood flow to the superior vena cava. In contrast to uphill varices, downhill varices have a retrograde blood flow and are located in the proximal esophagus. Downhill varices are rare and usually caused by superior vena cava obstruction due to bronchogenic carcinoma and mediastinal tumors, etc.[1,2]. They serve as collaterals either to bypass superior vena cava obstruction via the azygos vein or to drain the superior system to the portal vein when both the superior vena cava and the azygos vein are occluded. In contrast to the high risk of hemorrhage from uphill varices in portal hypertension, bleeding from downhill varices is extremely rare. Although an increased variceal wall tension is the ultimate factor causing bleeding in both types of varices, several factors may underlie this difference in bleeding tendency. First, in patients with uphill varices, coagulation capacity may be reduced due to concomitant liver disease with an inherently increased bleeding tendency. Second, exposure to esophagogastric reflux damages distal rather than proximal varices. Third, because distal uphill varices predominantly distend at subepithelial levels compared to the submucosal location of downhill varices in the midthoracic and proximal esophageal wall, variceal rupture is much more likely to occur near the esophagogastric junction[3]. We report a rare case of bleeding downhill varices in the absence of superior vena cava obstruction. Detailed diagnostic work-up showed that the downhill varices were caused by goiter. The varices disappeared after subtotal thyroidectomy.
Endoscopic view demonstrating downhill varices (A) and almost complete disappearance of the downhill varices (B) before and 12 mo after thyroidectomy of the right lobe.

3D reconstruction of CT angiography with view at the dorsal wall of the trachea (T) demonstrating a venous plexus of downhill varices (V) on the wall between the esophagus and trachea connected with a thyroid vein at the goiter (G) of the right thyroid lobe; * out of plane, cut off level thick slice.

Figure 1

Figure 2

CT angiography demonstrating downhill varices one day after hemorrhage (A) and disappearance of the downhill varices 12 mo after thyroidectomy of the right lobe (B). * indicates varices around the esophagus; T, trachea.

Figure 3

DISCUSSION

Downhill varices are usually associated with superior vena cava obstruction due to bronchogenic carcinoma, different types of mediastinal tumor and fibrosis, venulitis, surgical caval ligation and thyroid masses. Occasionally, as in our patient, downhill varices may develop without superior vena cava obstruction. Relatively few case reports have been published on bleeding downhill varices in relation to thyroid pathology. In a study of 1051 patients with cervical and retrosternal goiter, 3% of patients developed non-bleeding downhill varices. Lagemann performed barium swallows in 50 patients with recurrent thyroid enlargement and demonstrated that more than 50% of the patients have non-bleeding downhill varices. Blood from the thyroid plexus flows through the inferior thyroid veins (also called thyroid ima veins) into the brachiocephalic vein. In case of obstruction of the inferior thyroid veins, blood flows via the deep esophageal veins leading to esophageal varices. The esophageal varices can drain into collaterals to the brachiocephalic, azygos, hemiazygos and accessory hemiazygos veins, all of which finally drain into the superior vena cava. In the present case, goiter caused compression of the internal jugular vein. Blood flow over the thyroid plexus draining into the inferior thyroid veins might bypass compression of the internal jugular vein. However, in this patient downhill varices developed and bypassed this compression, suggesting that the function of inferior thyroid veins is insufficient. Both previous thyroidectomy and recurrent goiter are possible explanations, since inferior thyroid veins can be occluded either by primary or recurrent thyroid tumors or by surgical ligation during thyroidectomy and fibrogenesis or mediastinitis secondary to surgery. In the present case, the downhill varices drained into the azygos vein as illustrated in Figure 4. Hemorrhage of downhill varices is an emergency. However, the experience with treatment is limited because of its rare bleeding propensity.
In contrast to uphill varices, endoscopic sclerotherapy is not generally performed to treat downhill varices up to 5 cm below the upper esophageal sphincter, because retrograde flow of sclerosant through the azygos vein could result in spinal cord and vertebral body infarction. Fatal pulmonary embolism of cyanoacrylate used for endoscopic embolization of downhill varices has also been reported. Therefore, downhill varices should be recognized and distinguished from uphill varices. The use of a Sengstaken-Blakemore tube can be lifesaving. Endoscopic band ligation has been shown to be effective in preventing recurrent bleeding of downhill varices.

Finally, definitive treatment is performed to eliminate the cause of venous obstruction. As in this patient, surgery can successfully relieve obstruction. For goiter-related downhill varices, jodium therapy can also be effective.

In conclusion, downhill varices although rare, can cause upper gastrointestinal bleeding and should be suspected in any patient with evidence of thyroid enlargement or having a history of thyroid surgery, even though signs of superior vena cava obstruction are absent. Management of the underlying cause, as in this case by thyroidectomy, can efficiently lead to recovery and disappearance of the esophageal varices.

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