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Downhill varices: An uncommon cause of upper gastrointestinal bleeding

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ABSTRACT

Background: Upper gastrointestinal bleeding (UGIB) is a common condition in gastroenterology, but "Downhill Varices" (DHV) or varices of the upper oesophagus are an uncommon cause of UGIB, with different aetiology from lower third oesophageal varices and different therapeutic implications.

Case report: A 28-year-old male patient, with a history of chronic kidney failure secondary undergoing haemodialysis and superior vena cava syndrome (SVCS) due to multiple catheter replacements, was admitted to the Emergency Department with haematemesis secondary to a varicose vein rupture in the proximal third of oesophagus, treated initially with ethanolamine. Subsequent diagnostic studies showed the collateral circulation secondary to the SCVS. No further endoscopic or endovascular therapy could be performed and the patient will finally undergo a surgical bypass.

Discussion: DHVs are a very uncommon condition and endoscopic band ligation emerges as the appropriate therapeutic approach for the bleeding event. The definitive therapy continues to be that for the cause of the SVCS.
INTRODUCTION
Upper gastrointestinal bleeding (UGIB) is a common condition in gastroenterology, with an approximate incidence of 170 patients/100,000 inhabitants/year. The most common cause is a peptic ulcer, followed by oesophageal or gastric varices and oesophagitis. "Downhill varices" (DHV) or varices of the upper oesophagus are part of the second group. It is a little known condition, with an aetiology different from lower third oesophageal varices and with different therapeutic implications than usual. Below is a case report of UGIB due to DHV.

CASE REPORT
A 28-year-old male patient with a history of chronic kidney failure secondary to bilateral cystic renal dysplasia with vesicoureteral reflux. For this reason he had had three kidney transplants and was undergoing haemodialysis. Due to multiple catheter replacements in the subclavian vein, to perform this renal replacement technique, he suffered from thrombosis of the subclavian vein with secondary superior vena cava syndrome (SCVS).
He was admitted to the Accident and Emergency Department with haematemesis starting 48 hours before, affecting laboratory (Hb 5.9 g/dl) and haemodynamic values. An oral endoscopy was performed that evidenced three vascular cords from Killian's area up to 35 cm and at 27 cm one of them bleeding in a jet. These cords respected the distal oesophagus. Since the origin of this abnormal vascularisation was not known and assuming that it was secondary to portal hypertension, the varicose vein was treated with 5 cc of 5% ethanolamine in two punctures with good immediate results. Later the presence of portal hypertension was ruled out after a chest CT angiogram had been performed, requested due to the patient's history and suspecting an abnormal venous drainage at oesophageal level. This ascertained the existence of upper third oesophageal varices that depended on the lower thyroid veins, along with
marked collateral circulation in the chest wall and paravertebral veins. These branches enabled venous drainage for a chronic occlusion in both braquiocephalic venous trunks.

After the acute episode endovascular rechanneling of both vessels was attempted, but it was not possible given the chronic nature of the occlusion. In a new reassessment endoscopic therapy was ruled out as secondary prophylaxis, as the possible consequences for the rest of the collaterals were not known or due to the possible emergence of new oesophageal varices. Finally, the patient will undergo a surgical bypass between the superior vena cava and the right atrium.

DISCUSSION
Downhill varices were first described in 1964 by Felson and Lessure (1). The obstruction of the superior vena cava, or its main tributaries, produces a branching of the venous flow through collaterals to return blood to the right atrium. Part of these collaterals are oesophageal varices, which will form varices whose length depends on whether the level of obstruction is above or below the azygos vein. In the first case it will be limited to the upper third, while in the second it will affect the entire oesophagus.

As regards the cause of the obstruction, it may be secondary to the mass effect produced by lung malignancies (2), intrathoracic goiter (3), thymomas (4) or post-radiotherapy mediastinal fibrosis (5). This disease has also been described in patients with haemodialysis catheters where thrombosis of the blood vessel in which they are located occurs because of the replacement and handling of such catheters, as is the case of our patient (6-8). There are cases associated with Behçet's disease (9) and on other occasions there is no venous obstructive compromise at mediastinal level and the onset of DHV is secondary to severe pulmonary hypertension (10).

Once diagnosed, in the case of having an acute UGIB episode, the varicose vein must be treated, taking into account that the treatment, given the venous flow, must in this case be proximal to the point of bleeding. The use of band ligation is recommended over sclerotherapy as there are cases that describe complications such as spinal heart attacks and pulmonary thromboembolism (11,12). Once the acute episode is resolved
a possible cause of the SVCS should be investigated, so that by resolving it we can re-establish the venous flow. In the case of malignancies, surgery or chemo/radiotherapy would be indicated, whereas, when thrombosis occurs angioplasty with or without stenting seems to be the most appropriate method, with a surgical bypass as the second option. In cases where it is not possible to return the appropriate venous flow, secondary UGIB prophylaxis could arise, but there is no sound scientific data to support it. A review of 130 patients with DHV showed a 9% lower incidence of UGIB than in varices secondary to portal hypertension, although this type of event is more life-threatening (13). In this regard, there is a case of SVCS secondary to Behçet's disease in which a partial SCV thrombosis gave rise to DHV. After the first bleeding episode, multiple endoscopic ligation sessions were conducted achieving variceal eradication (14).

Therefore, DHVs are a very uncommon condition that must be suspected in patients with SVCS-related diseases. In this way an appropriate therapeutic approach for the bleeding event can be provided that would prevent the development of complications arising from the treatment. The definitive therapy continues to be that for the cause of the SVCS in the first place, also considering that there are currently no scientific data to support one or other strategy of secondary prophylaxis in patients in whom venous flow cannot be re-established.

REFERENCES
Fig. 1. Point of haemostasis on varicose vein in upper third of the oesophagus.

Fig. 2. Radiological image (CAT) of oesophageal varicose vein in upper third of the oesophagus (arrow).

Fig. 3. Radiological image (CAT) of chronic occlusion of both braquiocephalic venous trunks (arrow).