

An unusual case of Esophageal Varices: “Downhill” type

Case Report

Abstract

Downhill esophageal varices is a rare condition, less common than the classical “uphill” varices characterized by the presence of esophageal varices in the absence of portal hypertension. Because of the rarity of this condition, there are no clear guidelines or recommendations regarding its management. This is a case of Downhill esophageal varices incidentally discovered during endoscopy.

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Introduction

“Downhill” esophageal varices is a rare condition, less common than the classical “uphill” varices [1-4], it was first reported in 1964 by Felson and Lessure [5]. Obstruction of blood flow through the inferior vena cava and/or its tributaries, forces the blood to find an alternative way back to the right heart. This is achieved through the portal circulation via the esophageal veins. The esophageal veins will have to accommodate the backflow of blood coming from the azygos, hemiazygos, superior intercostal, bronchial, and the inferior thyroid veins.

Case presentation

A 31 year old female presented to the Gastroenterology Clinic in 2009 for abdominal pain. Her past surgical history includes a successful renal transplant in 2006. Her past medical history includes renal insufficiency in 2000, managed with Dialysis until 2006, complicated by fistula thrombosis so multiple central lines were inserted. Other past medical history includes Lupus, High Blood Pressure, and Hypercholesterolemia. Her medical treatment included Amlodipine, Bisoprolol, Atorvastatin, Mycophenolic Acid, and Tacrolimus.

The upper gastrointestinal endoscopy (Figure 1,2) revealed small and medium size esophageal varices in the upper and middle thirds of the esophagus, without red signs, and “snake skin” antritis. The presence of esophageal varices warranted a workup for portal hypertension. ALTS, Albumin, Bilirubin were normal. Ferritin was elevated (771 ng/ml), Transferrin saturation was 38%. Genetic test for hemochromatosis revealed heterozygous mutation on Cys 282Tyr and His 63Asp, and no mutation on Ser 65Cys. Ceruloplasmin, 24h Urine copper, a1 Anti trypsin, Anti mitochondria, ANA, and Anti SM, were all normal. Anti LKM was border line. Abdominal US with Doppler confirmed normal portal venous system. A Chest CT scan and Abdominal MRI (Figure 3,4,5) revealed a thrombosed SVC with venous drainage into the heart through the IVC, azygos and hemiazygos veins and through the multiple collateral vessels in the anterior and posterior wall of chest and abdomen bilaterally. Liver and Portal network was normal. The retained diagnosis was Downhill Esophageal Varices, secondary to thrombosis of the Superior Vena Cava.

Etiologies of “Downhill” esophageal varices

Thrombosis of the Superior Vena Cava or its tributaries is the most common etiology of “Downhill” esophageal varices. There is a long list of etiologies described in the literature including central venous catheterization [2,6,7], mediastinal fibrosis [8-

14], primary and metastatic mediastinal tumors [3,10,11,15-19], mediastinal lymphadenopathy secondary to head and neck cancers such as carcinoma of the tongue [20], substernal goiters and thyroid masses [21-23], thyroid carcinoma [3,21-25], lung cancer [3,10,16,18-20,26, 27,28], thymoma [3,29], systemic venulitis [30], Behcet’s disease [31-36], Castleman’s disease (angiofollicular lymph node hyperplasia) [4], and as a late complication after correction of congenital heart defect [37]. Downhill varices may also develop without superior vena cava thrombosis [4,38], as seen in goiter [39-41,43-45,46], history of thyroid surgery [22,40-42], severe pulmonary hypertension [47], thyroid tumors [22], abnormal cricopharyngeal muscle constriction or any abnormal muscular constriction of the hypopharyngeal veins [48].



Figure 1: Esophagoscopy of the presented case showing a variceal vein situated at the upper third of the esophagus.



Figure 2: Esophagoscopy of the presented case showing esophageal veins at the middle third of the esophagus.

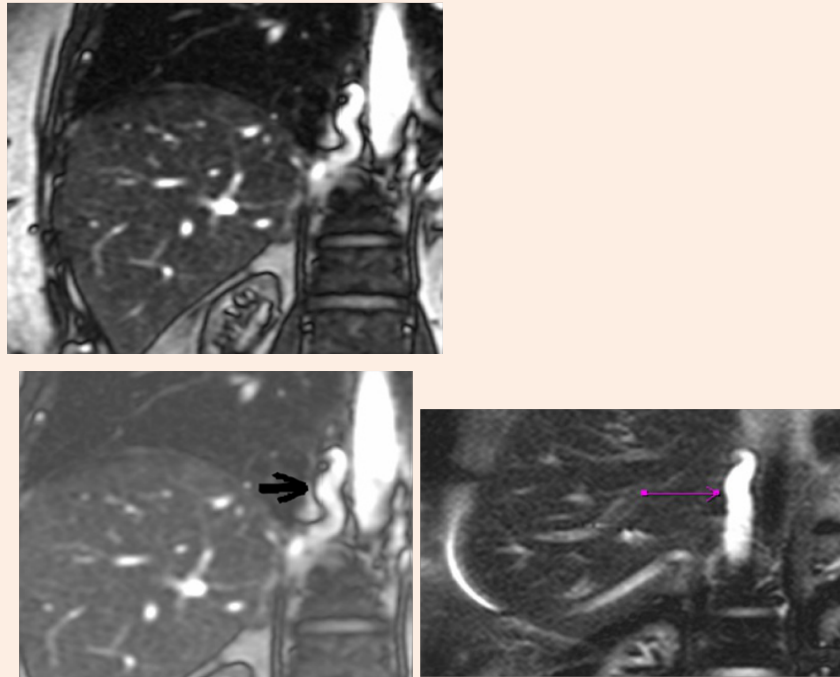


Figure 3: Prominent Azygos vein.

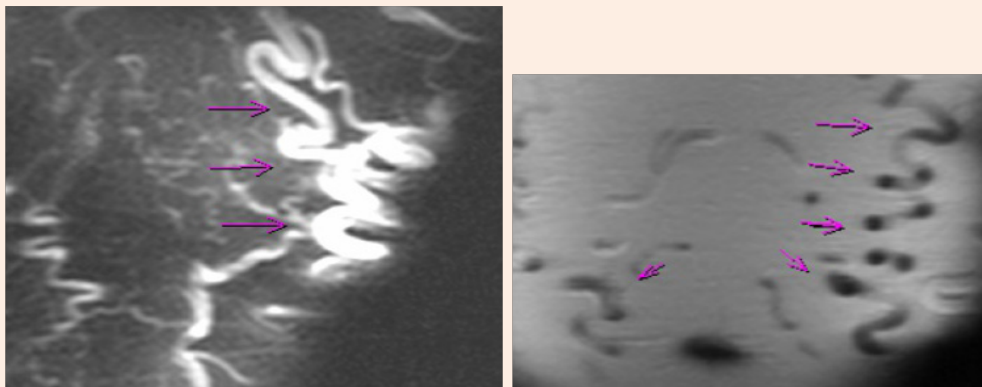


Figure 4: Chest and Abdominal wall collaterals.

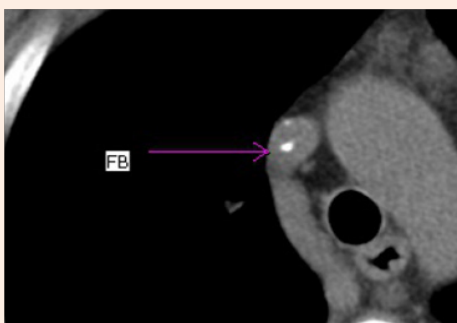


Figure 5: Thrombosed SVC.

Clinical picture of SVC syndrome

Clinical presentation of "Downhill" esophageal varices is dominated by clinical symptoms of Superior vena cava obstruction that were present in 91.4% of the cases described in the literature [3]. Patients may present with Face, neck, and arm swelling, tongue swelling, hoarseness, epistaxis, dyspnea, cough, headache, visual disturbance, syncope, dysphagia, upper gastro-intestinal bleeding, haemoptysis, and dilated chest collaterals.

Accidentally discovered "Downhill" esophageal varices and upper gastrointestinal bleeding may be the first presentation [31].

Incidence

Downhill varices are less common than the so called uphill

distal esophageal varices caused by portal hypertension [49,3,50,8,21,10,51,23,52]. We found little information about the incidence of downhill varices in the literature. In patients with thyroid disease, Downhill varices were found in 4% of patients with primary goiters, 12% of patients after thyroidectomy, and 54% of patients with recurrent thyroid tumors [22]. Similar results were found in a series of 1051 patients with cervical and retrosternal goiter, where 3% of patients developed non-bleeding downhill varices [46].

Mechanism of variceal formation

"Downhill" esophageal varices develop when there is an increase in blood flow in the esophageal plexuses.

Case of superior vena cava obstruction

Esophageal varices serve as collateral branches to bypass an obstruction in the superior vena cava, either via azygos vein if the obstruction is located above the level of the azygos vein (proximal), or via the portal system if the obstruction is involving the azygos vein [6,40,8,53,21,10,51,23,52,11,12,14]. The most important factors that determine the extension of "Downhill" esophageal varices are the level of Superior Vena Cava obstruction and its duration [3,8]. Obstruction of the Superior Vena Cava above the level of the azygos vein will result in the formation of varices in the upper third of the esophagus. In contrast, obstruction of the Superior Vena Cava below the level of the azygos vein will result in the formation of varices along the entire length of the esophagus.

Case without superior vena cava obstruction

Increase in blood flow in the esophageal plexuses may occur without superior vena cava obstruction. Examples include

- a. Castleman's disease [4]
- b. Thyroid diseases

In thyroid, blood from the thyroid plexus normally flows through the inferior thyroid veins into the brachiocephalic vein to reach the superior vena cava. In case of obstruction of the inferior thyroid veins, blood flows into esophageal venous plexus (via the deep esophageal veins) leading to "Downhill" esophageal varices. Inferior thyroid veins can be occluded by different mechanisms, such as accidental surgical ligation during thyroidectomy, fibrogenesis secondary to surgery [40,22], or by primary or recurrent thyroid tumors [22], leading to development of proximal esophageal varices.

- c. Primary esophageal motor disorders

"Nutcracker" esophagus and diffuse esophageal spasm were considered as a possible causes of proximal "downhill" varices esophageal [53,54].

- d. Unknown causes: three patients with downhill varices of unknown cause have been reported, probably as a consequence of obstruction of the posterior hypopharyngeal venous plexus by abnormal cricopharyngeus muscle contractions [48].

Complications of "downhill" esophageal varices

"Downhill" esophageal varices that are presenting with bleeding

are extremely rare in the literature [21,30, 51,18,12,14,48]. It was reported that 7 to 9 % of patients with "Downhill" esophageal varices presented with upper gastrointestinal hemorrhage [5,3,30], with some cases of life-threatening bleeding [6,30]. Most of the bleeding varices are caused by vena cava obstruction due to tumors or mediastinal fibrosis [21,51,18,12,14]. "Downhill" varices represent only 0.1% of all esophageal variceal bleeding [55]. Two explanations were suggested: the first is the lack of coagulopathy usually associated with chronic liver disease in varices secondary to portal hypertension, and the second is the higher location of "Downhill" varices in the esophagus, away from erosive gastroesophageal reflux. "Downhill" varices are located in the submucosa of the proximal esophagus, in contrast to the esophageal varices of portal hypertension that are located in the superficial subepithelium of the distal esophagus [49,40].

Treatment

Treatment plan needs to be individualized. Primary treatment of "downhill" esophageal varices is directed toward the etiology [49]. In superior vena cava thrombosis, removal of a catheter, chemical or mechanical thrombolysis of the clot [56] and/or venoplasty and stenting [2,57,58] has been reported to resolve the problem. In surgical candidates a vascular bypass of the superior vena cava can be attempted [6]. Treatment of the underlying medical condition was sufficient in some cases to resolve the varices, such as steroids and dapsone in systemic vasculitis [30], chemoradiotherapy or resection of a tumor [4,26,15,59,60], thyroidectomy [41,43,21,45] and radioactive iodine therapy [40] in case of goiter, and surgical resection for Castleman's disease [4]. Inferior thyroid artery embolization was attempted in a case of downhill varices caused by a goiter [39]. Sclerotherapy was complicated by spinal cord infarction in some cases [48,61,62], caused by flow of sclerosant from the azygos to spinal veins when injected at the level of the middle and upper esophagus [61]. Complication with pulmonary embolism was also described [63].

Variceal band ligation is effective for controlling bleeding in patients with uncorrectable underlying disorders [49,31,64]. The site of banding was not clearly defined. Some suggest banding proximal to the varices for hemodynamic reasons, as opposed to the distal banding in esophageal varices due to portal hypertension [67]. The risk of bleeding or perforation seems higher because of the weakness of the proximal esophageal posterior wall and overall lack of serosa [65,66]. Endoscopic treatment of "downhill" varices should be undertaken only in severe cases [46]. The use of a Sengstaken-Blakemore tube can be lifesaving in case of uncontrolled bleeding [40]. When all the above measures fail, palliative measures may be applied [6,46]. Possible technical complications of the treatment are not well studied because of the rarity of this condition.

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