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Downhill Esophageal Variceal Bleeding Complicating a Superior Vena Cava Syndrome in Behcet's Disease: Another Case Report of a Rare Situation

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Abstract: Superior vena cava (SVC) thrombosis, a possible complication of Behçet disease (BD), can lead to the development of venous collaterals in the upper esophagus known as downhill esophageal varices (DEV). It is a rare condition, and bleeding from this varices, althought rarer, can be life threatening. Presently describe is a case of a 25-year-old male patient with five years history of vasculo Behçet, presenting a SVC syndrome with upper gastro intestinal bleeding (GIB) consequent to the UEV.
Keywords: Behcet's disease, SVC syndrome, Down-hill esophageal varices, upper gastro intestinal bleeding.

INTRODUCTION

"Downhill" esophageal varices (DEV) are less common then the distal esophageal varices, also known as the uphill type (UEV), mostly seen in patients with portal hypertension. This is a little known condition, mostly described as a consequence of secondary obstruction or compression of the superior vena cava (SVC), bypassed via the azygus vein [1]. Bleeding from the DEV happens rarely, accounting for 0.1% of patients present with hematemesis [2]. This occlusion can be caused by compressive masses such as lung cancers, mediastinal and thyroid tumors or mediastinal lymphadenopathy [2, 3].

Less commonly, it can be caused by a venous thrombosis in systemic vasculitides such as Behçet disease (BD). The syndrome of SVC obstruction is a rare situation described in only 2% of patients with a Behçet history. We report herein a case of DEV bleeding complicating a SVC obstruction in a young adult patient with BD.

CASE REPORT

A 25 year old male patient was admitted to the hospital with sudden onset hematemesis with no abdominal pain of three days progression. He had a five years history of BD. The diagnosis was based on recurrent bipolar aphtosis and pseudofolliculitis. An internal jugular vein thrombosis was present at the time of the diagnosis. No other involvements (eyes, joints, nervous and cardio vascular system) were found. A combination of anticoagulant and immunosuppressive treatment (prednisolone relayed by cyclophosphamide) was initiated. There was no history of jaundice, hepatitis or familial liver disease. No past history of upper gastrointestinal bleeding was mentioned. He denied any NSAIDs use or alcohol intake. Physical examination found a pale non icteric patient, with normal vital signs. Rectal examination was positive for melena. No other abnormalities were noticed except for scares of scrotal ulcers and a chest collateral venous circulation (figure 1-2). Laboratory values on admission showed anemia (9.7 g/dL hemoglobin) with normal platelet count. The international rationalised ratio (INR) was 2,3. Anticoagulants were stopped and the patient received intravenous infusion of Somatostatine and Omeprazole. Upper gastrointestinal endoscopy, performed 9 hours after the bleeding, showed two grade II columns of proximal esophagus varices, with red color signs as stigmata of recent bleeding (figure 3). The distal part of the esophagus, the stomach and the duodenum had normal appearance and no peptic ulcer was found. Bleeding from these varices was the most likely diagnosis and variceal band ligation was performed. Four bands were deployed with excellent hemostasis. No portal hypertension due to chronic liver disease was found. Liver tests and abdominal doppler sonography were normal. Because of the underlying vasculitis, thrombosis of portal vein or hepatic veins was ruled out. To define the etiology of these varices, we performed a chest contrast –enhanced computed tomography scan. It showed a marked thrombosis of the SVC with cervicothoracic collateral circulation close to the esophagus (figure 4, 5). Anticoagulation therapy was differed; the patient remained stable and was discharged five days later without any complications or further bleeding. We managed to perform a second band ligation three weeks later. Our patient underwent a third endoscopy; varices were eradicated. Anticoagulation

was initiated again and no bleeding occurred during the last 12 months.

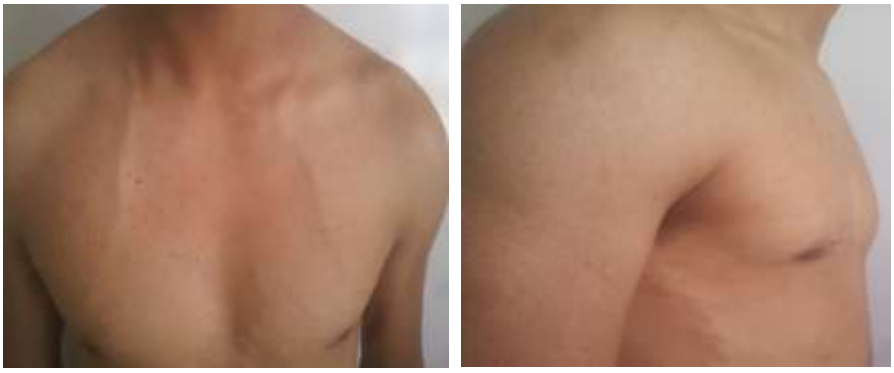

Fig-1-2: chest collateral venous circulation

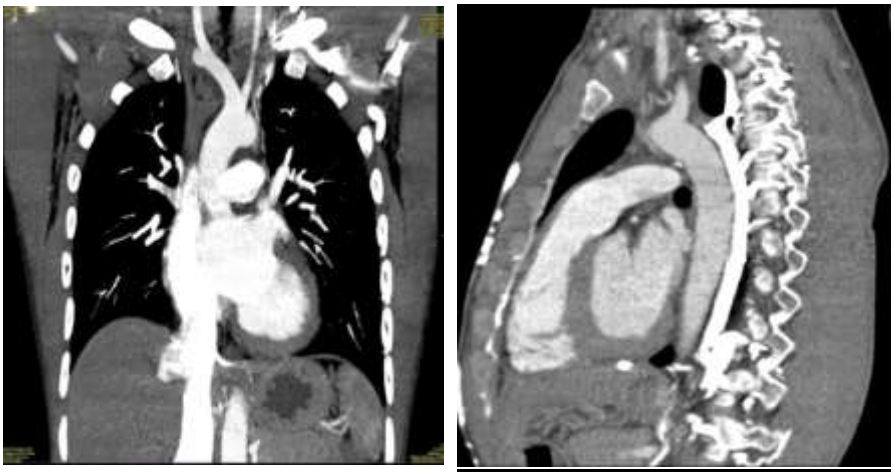

Fig-4-5: thrombosis of the SVC with cervicothoracic collateral circulation close to the esophagu

DISCUSSION

The first description of DEV was made by Felson and Lessure in 1964 [4]. These varices are located in the proximal third of the esophagus as a direct consequence of the blood flow obstruction in SVC. The development of these various collateral pathways aims to bypass the SVC obstruction via the portal circulation while directing blood flow "downwards" [3-5]. Mostly seen in the upper part of the esophagus, their downward extension depends on the level and the rapidity of the obstruction and also on its duration [6-7]. We believe that the obstruction of the proximal SVC is likely to cause DEV extended to the entire length of the esophagus, at the opposite of distal obstructions above the azygous inflow, which are more likely to cause downhill varices limited to the upper esophagus [8].

DEV are related to various etiologies, with a major predominance of malignancy. It is well known now that lung cancers, mediastinal neoplasms as lymphomas, mediastinal metastases, intra thoracic goiter, thyroid carcinoma, thymoma and are common causes of DEV. Central venous catheterization and mediastinal fibrosis can also lead to this condition. Exceptionally, they can be seen in patients with systemic vasculitides as BD or hypercoagulability [4-8]. Some cases of DEV related to procedures such as pacemaker insertion or hemodialysis access have been reported [9].

To today, 80 cases of DEV have been reported in adult literature [4]. In our knowledge, thirteen cases of DEV complicating a SVC obstruction in BD have been reported, among them only five had variceal bleeding [1-3-10-11-12-13-14-15-16]. Indeed, bleeding from DEV is exceptional, accounting for less than 9% of all DEVs, and 0,1% of upper gastrointestinal bleeding [17]. Because of the rarity of the condition, there is no consensus on the treatment of DEV bleeding, as they tend to disappear with the etiological treatment. We managed to do an elastic band ligation since stigmata of recent bleeding were present on grade II varices, which seemed to be a safe and effective choice. Some authors tend to disapprove the ligation, since the weakness of the proximal esophageal posterior wall and overall lack of serosa can lead to perforation.

As said earlier, the main pillar of the treatment is to solve the condition causing the DEV, and therefore should be individualized. The management of the underlying medical condition was discussed with the

department of internal medicine in our case. As known, BD is a systemic vasculitis affecting mostly the arterial system, less frequently the venous one. It is characterized by recurrent oral and genital aphtosis and uveitis [16]. Vasculo-Behçet patients are at risk for multiple vessel-related complications including thrombosis, stenosis, occlusions, and aneurysms. The primary reason for clot seems to be an inflammatory process in the vessel wall [18]. Venous thrombosis occurs in 25–35% of patients with BD while thrombosis of the SVC is seen in less than 2% of patients with BD [19, 20]. In our case, our patient was on prior anticoagulation therapy. Although the INR was in the optimal therapeutic range, we tend to believe that it might have facilitated variceal bleeding. The use of anticoagulant itself is controversial in BD thrombosis, knowing that it is adherent to the vessel and the risk of pulmonary embolism is negligent. The management of angio-Behçet is controversial, including mostly corticosteroids and immunosuppressive treatments. The combination of immunosuppressive and anticoagulation therapies does not do better than immunosuppression alone.

REFERENCES

1. Yaşar B, Kılıçoğlu G. Behçet's disease-related superior vena cava syndrome and bleeding downhill varices: A rare complication. Ulus Travma Acil Cerrahi Derg. 2017 Mar 1;23(2):170-2.
2. Areia M, Romãozinho JM, Ferreira M, Amaro P, Freitas D. " Downhill" varices. A rare cause of esophageal hemorrhage.
3. Tavakkoli H, Asadi M, Haghighi M, Esmaeili A. Therapeutic approach to" downhill" esophageal varices bleeding due to superior vena cava syndrome in Behcet's disease: a case report. BMC gastroenterology. 2006 Dec;6(1):43.
4. Gómez-Aldana AJ, Gómez-Zuleta MA. Varices esofágicas en Downhill secundarias a trombosis de vena cava superior por déficit de factor V. Revista de Gastroenterología de México. 2017 Jan 1;82(2):179-80.
5. Fleig WE, Stange EF, Ditschuneit H. Upper gastrointestinal hemorrhage from downhill esophageal varices. Digestive diseases and sciences. 1982 Jan 1;27(1):23-7.
6. Palmer ED. Primary varices of the cervical esophagus as a source of massive upper gastrointestinal hemorrhage. The American journal of digestive diseases. 1952 Dec 1;19(12):375-7.
7. Otto DL, Kurtzman RS. Esophageal varices in superior vena caval obstruction. The American journal of roentgenology, radium therapy, and nuclear medicine. 1964 Nov;92:1000-12.
8. Popr A, Cutler AF. Bleeding downhill esophageal varices: a complication of upper extremity hemodialysis access. Gastrointestinal endoscopy. 1998 Mar 1;47(3):299-303.
9. Nguyen LP, Sriratanaviriyakul N, Sandrock C. A rare but reversible cause of hematemesis:"Downhill" esophageal varices. Case reports in critical care. 2016;2016.
10. Ennaifer R, Hamzaoui SB, Larbi T, Romdhane H, Abdallah M, Hadj NB, M'rad S. Downhill oesophageal variceal bleeding: A rare complication in Behçet's disease-related superior vena cava syndrome. Arab Journal of Gastroenterology. 2015 Mar 1;16(1):36-8.
11. Ichikawa M, Kobayashi H, Mukai M, Saitoh Y. Superior Vena Cava Syndrome as Initial Symptom of Vasculo-Behçet's-diseaseCase Report. The Japanese journal of thoracic diseases. 1991 Oct 25;29(10):1344-8.
12. Ishikawa R, Noguchi T, Matsumoto K. A case of downhill esophageal varices--Behçet disease associated with abdominal aortic aneurysm and occlusions of the superior and inferior vena cava. Nihon Geka Gakkai zasshi. 1986 Dec;87(12):1576-82.
13. Tsuji S, Suzuki Y, Tomii M, Matsuoka Y, Kishimoto H, Irimajiri S. Behçet's disease associated with multiple cerebral aneurysms and downhill esophageal varices caused by superior vena cava obstruction: a case report. Ryumachi.[Rheumatism]. 1990 Oct;30(5):375-9.
14. Orikasa H, Ejiri Y, Suzuki S, Ishikawa H, Miyata M, Obara K, Nishimaki T, Kasukawa R. A case of Behçet's disease with occlusion of both caval veins and "downhill" esophageal varices. Journal of gastroenterology. 1994 Aug 1;29(4):506-10.
15. Suzuki H, Mizuguchi A, Fujii M, Nakagomi T, Okabe K. A case of vasculo-Behçet with esophageal downhill varices (author's transl). Nihon Shokakibyo Gakkai zasshi= The Japanese journal of gastro-enterology. 1982 Jan;79(1):93.
16. Han DS, Kim JB, Lee OY, Sohn JH, Park KN, Park CK. A case of Behçet's syndrome with supeior vena cava syndrome. The Korean journal of internal medicine. 1998 Feb;13(1):72.
17. Areia M, Romãozinho JM, Ferreira M, Amaro P, Freitas D. Unidade de Cuidados Intensivos de Gastrenterologia–13 anos de vida. GE–J Port Gastrenterol. 2005;12(3).
18. Calamia KT, Schirmer M, Melikoglu M. Major vessel involvement in Behçet's disease: an update. Current opinion in rheumatology. 2011 Jan 1;23(1):24-31.
19. Houman MH, Ghorbel IB, Salah IK, Lamloum M, Ahmed MB, Miled M. Deep vein thrombosis in Behcet's disease. Clinical and experimental rheumatology. 2001 Sep 1;19(5; SUPP/24):S-48.
20. B'chir SH, Harmel A, Bouslama K, Abdallah M, Ennafaa M, M'rad S, Ben MD. Behçet's disease in Tunisia. Clinical study of 519 cases. La Revue de medecine interne. 2006 Oct;27(10):742-50.