Case Report

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Budd-Chiari Syndrome in Behçet's Disease successfully managed with immunosuppressive and anticoagulant therapy: A case report and literature review

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Summary **Behçet's Disease (BD) is a rare, chronic and recurrent inflammatory multisystemic condition of unknown origin that can affect any tissue. The vascular system is involved in 5-40% of cases of BD, including venous and arterial beds and it has a relapsing course. Budd-Chiari syndrome (BCS) is a rare complication of BD with a frequency of < 5% among patients with vascular involvement and is more frequent in men (89.5%). Two clinical presentation groups of BCS related to BD have been described: the "symptomatic" form and the "silent" form. We present a case of BD in a young woman presented as symptomatic severe BCS with rapid progression of coagulopathy reaching a spontaneous INR of 1.74 and increased ascites by ultrasound control. BD was confirmed through clinical history. The patient was treated with a high-dose pulse of corticosteroids and cyclophosphamide with a strikingly favorable response in the first forty-eight hours. Although several studies have demonstrated a survival improvement with the use of transjugular intrahepatic portosystemic shunt in patients with severe BCS, it was discarded due to the lack of evidence of this procedure in patients with BD and the fact that it could trigger a vascular pathergy phenomenon. Vascular BD should be suspected in recurrent venous and/or arterial thrombosis since it is associated with high morbidity and mortality. Immunosuppressive treatment is critical for the management of vascular involvement in BD. However, the role of anticoagulation is debatable. We suggest an algorithm for the management of BCS associated with BD.**

Keywords: Behçet's disease, Budd-Chiari syndrome, immunosuppressants, anticoagulation, transjugular intrahepatic portosystemic shunt

1. Introduction

Behçet's Disease (BD) is a rare, chronic and recurrent inflammatory multisystemic condition classified as vasculitis of unknown origin that can affect any tissue of the economy (mucocutaneous, ocular, cardiovascular arterio-venous, central nervous system, gastrointestinal, joint, among others). An aberrant response of different immunological pathways in relation to triggers (infectious or environmental) in predisposed subjects has been postulated in the pathogenesis of the disease, typically the human leukocyte antigen (HLA) class I gene: HLA-B51. BD is sometimes referred to as the "Silk

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Route disease" because it is mainly distributed in those regions. The mean age of onset is usually in the third decade and both genders are affected equally. To date there is no specific test to confirm the diagnosis of BD, which is based on clinical criteria (*1-3*).

BD is unique among other vasculitis as it usually affects the venous rather than the arterial vessels. BD shows a tendency to thrombosis associated with vascular inflammation. In fact, lower extremity vein thrombosis (LEVT) can be considered as the hallmark of the vascular involvement. Vascular involvement is an early manifestation of BD that affects predominantly men, and its prevalence shows a wide range (5-40%) in the published literature (*1,4*).

Budd-Chiari syndrome (BCS) is a rare complication in BD with a frequency of < 5% among patients with vascular involvement. It is developed due to thrombosis of suprahepatic veins and/or inferior vena cava (IVC), and has two different clinical presentations: a symptomatic presentation, with a high mortality (up to 60%) and manifested as abdominal pain, ascites and collateral circulation on the abdominal wall; and a silent presentation, with a better prognosis (10% mortality) and manifested without ascites but with efficient collateral formation (*5*).

Immunosuppressants, with or without glucocorticoids, are essential in the management of vascular involvement in BD. They have been shown to reduce the relapse rate and to prolong survival in several retrospective studies. In patients with BD, conditions associated with higher mortality as BCS require an early and aggressive medical treatment, including cyclophosphamide and glucocorticoid pulses. In resistant cases, anti-tumor necrosis factor (TNF) agents could also be effective (*5,6*). Whether to add anticoagulants to prevent recurrent thrombosis has been debated (*5,7*). Several retrospective studies showed the inefficacy of anticoagulation alone or added to immunosuppressants in preventing recurrences (*8*) and it could increase the risk of aneurysmal rupture (*6,9,10*). In the last decade, several studies have demonstrated a survival improvement with the use of transjugular intrahepatic portosystemic shunt (TIPS) in patients with severe BCS, remarking the use of TIPS as a definitive treatment prior to liver transplantation, and not only as a bridging treatment (*11,12*). However, there is scarce evidence regarding the efficacy and safety of this procedure in patients with BCS in the setting of BD and the fact that it could trigger a vascular pathergy phenomenon should be considered (*5,10,13*).

We present a case of BD in a young woman presented as symptomatic BCS successfully managed with immunosuppressive and anticoagulant therapy. Consequently, we review the management and prognostic implications and suggest an algorithm for the management of BCS associated with BD.

2. Case Report

A 29-year-old woman from Equatorial Guinea had a history of malaria one year before, long-term treatment with oral contraceptives, without toxic habits or cardiovascular risk factors. In April 2018, she presented to our emergency department with a 7-day history of diffuse abdominal pain, nausea and vomiting. In addition, the patient reported cough without expectoration, fever and dyspnea on moderate exertion. Physical examination revealed blood pressure 104/68 mm Hg, heart rate 96 bpm, oxygen saturation 98%, temperature 38.2°C. Lung auscultation disclosed crackles on the right base; abdomen was distended and painful to deep palpation focused on the upper right quadrant, with no signs of peritoneal irritation; pitting edema was found in both lower limbs. Laboratory tests showed hemoglobin 7.6 g/dL with a mean corpuscular volume of 73 fl, 8,200 leukocytes/uL, 140,000 platelets/µL, international normalized ratio (INR) of 1.53, fibrinogen 433 mg/dL, alanine aminotransferase 188 IU/L, gamma-glutamyl transferase 455 IU/L, alkaline phosphatase 355 IU/L, total bilirubin 0.5 mg/dL and creatinine 0.56 mg/dL. The urinalysis was normal. Doppler-ultrasound (US) of both lower limbs and abdominal computed tomography (CT) scan revealed extensive bilateral deep vein thrombosis involving IVC and both common iliac veins with extension to femoral veins, signs of ischemia of the right hepatic lobe, mild ascites and thrombosis of the right suprahepatic vein (SV) (Figures 1A-1E). Echocardiogram was remarkable for normal biventricular function with normal ejection fraction, with the presence of minimal pericardial effusion. An upper endoscopy did not show pathological findings.

The patient was admitted to the intermediate care unit; unfractionated heparin and diuretics were started. Initial evolution was poor, with rapid progression of coagulopathy reaching a spontaneous INR of 1.74 and increased ascites by ultrasound control. Considering the extension and atypical location of the thrombosis, and the presence of fever, differential diagnosis was broad and included infectious, autoimmune and malignant causes, mainly hematological.

Other tests were performed. Human immunodeficiency virus, hepatitis C virus and blood cultures were negative. A thick drop test and Plasmodium antigen ruled out the presence of acute infection by *Plasmodium spp*. A pulmonary arteries angio-CT showed no signs of pulmonary embolism. Antinuclear antibodies, neutrophil cytoplasmic antibodies and antiphospholipid antibodies were negative and the immunoglobulin count was normal. Hematological findings were compatible with iron deficiency anemia and Coombs tests were negative.

Taking into account the clinical picture, especially the extension and the atypical location of thrombosis, BD was suspected and clinical history was extended. The patient recognized recurrent painful oral and genital ulcers for the last 10 years, and intermittently painful

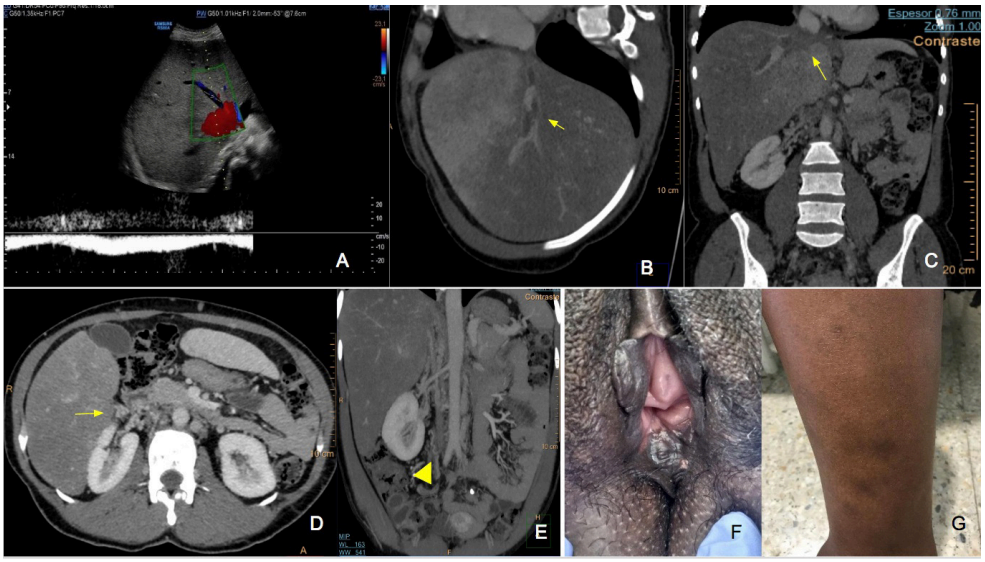

Figure 1. (A), Hepatic doppler-ultrasound revealed thrombosis and absence of flow involving the right suprahepatic vein and partially the inferior vena cava; **(B)**, Oblique coronal CT scan image showed thrombosis of the right hepatic vein and low attenuation of the right hepatic lobe in the venous phase; **(C)**, Coronal CT image shows inferior vena cava chronic thrombosis and right hepatic vein thrombosis with thrombus head leaning out into the cava; **(D)**, Axial CT scan shows low attenuation in the right hepatic lobe as an ischemic finding and chronic juxtarenal inferior vena cava thrombosis with hemiazygos hyperthropy; **(E)**, Coronal CT scan image shows complete inferior vena cava thrombosis; **(F)**, The gynecological examination revealed a well-defined painful ulcer in the left lower vaginal lip; **(G)**, Intermittently painful pretibial nodules with residual hyperpigmentation for the last 2 years that resembled erythema nodosum.

pretibial nodules with residual hyperpigmentation for the last 2 years that resembled erythema nodosum. The gynecological examination revealed a well-defined painful ulcer in the left lower vaginal lip (Figures 1F and 1G). The patient met criteria for BD with vascular involvement, associated with BCS. Pathergy test was negative.

The patient was treated with high-dose pulse of corticosteroids and cyclophosphamide with a strikingly favorable response in the first forty-eight hours after the beginning of immunosuppressive therapy, including defervescence, improvement of ascites and edema in lower limbs, as well as correction of coagulopathy (INR 1.15). Previous to immunosuppressive therapy, serology tests to assess chemoprophylaxis were gathered: anti-HBs negative and anti-HBc positive, with DNA quantification for hepatitis B virus of 17 IU/mL (1.23 *log*), and interferon gamma release assay test was positive. Lamivudine and isoniazid were started as prophylaxis.

The patient received seven cycles of cyclophosphamide every fifteen days. An abdominal US performed one month later showed significant recovery of the venous blood flow in the right suprahepatic vein, without ascites. Besides, an US of both lower limbs disclosed partial recanalization in both iliac and femoral veins. Nine months later, she progressed favorably with occasional appearance of genital ulcers without other complications, and with good tolerance to oral treatment: she is currently treated with prednisone, azathioprine and colchicine. In addition, anticoagulant treatment with acenocoumarol has been maintained.

3. Discussion

The vascular system is involved in 5-40% of cases of BD, including venous and arterial beds and it has a relapsing course. BD is unique among other vasculitis as it usually affects veins rather than arteries and it has significant thrombotic tendency associated with vascular inflammation, which cannot be explained by thrombophilic factors. LEVT is the most common type of vascular involvement. On the other hand, some rare presentations of vascular affection as pulmonary artery aneurysms, cardiac involvement and BCS are associated with a significant morbidity and mortality (*4,5,10*).

BCS is defined as an outflow obstruction at one or several levels from small hepatic venules to the IVC junction with right atrium, as a result of thrombosis or secondary fibrosis of the mentioned territory. BCS can be primary or secondary-due to compression or external invasion of the venous system-, and its presentation varies from asymptomatic, subacute-chronic or acute-fulminant states (*14-16*). Clinically, patients usually present with abdominal pain, fever, ascites, hepatosplenomegaly, collateral circulation and lower limb edema; other more severe findings include upper gastrointestinal bleeding and hepatic encephalopathy. Jaundice is not frequent, but the increase in direct bilirubin levels is a poor prognosis factor. In addition, BCS has been associated with an increased risk of long-term development of hepatocellular carcinoma. The diagnosis of BCS is based on clinical history, analytical data (impaired liver function and a serum albumin-ascites concentration

gradient ≥ 1.1), and by demonstrating obstruction to the flow of the hepatic venous territory by non-invasive imaging techniques: hepatic doppler ultrasound (first step), three-phase CT-scan and magnetic resonance imaging. The biopsy is relegated to unclear cases and it shows a centrilobular pattern predominance which is not exclusive of the disease. Among the most frequent causes of BCS are hematological conditions, predominantly myeloproliferative syndromes. A therapeutic strategy has been proposed to manage primary BCS and consists of anticoagulation, correction of risk factors, diuretics and prophylaxis for portal hypertension. Other procedures including angioplasty for short-length venous stenoses, TIPS or liver transplantation are reserved for refractory cases (*12,15,16*).

The association between BCS and BD has previously been described and is a rare complication of BD with a frequency between 2-5% in those patients with vascular involvement. However, in "endemic" areas of the disease, BD is one of the main causes of BCS. Some distinctive features have been described in BCS associated with BD including: younger age, higher frequency in males, IVC occlusion rather than isolated thrombosis of the hepatic veins, rare involvement of the portal system, better response to the immunosuppressive therapy compared to anticoagulation, and poor response to vascular interventions. Two clinical presentation groups of BCS related to BD have been described: the "symptomatic" form and the "silent" form. The former is presented with liver failure (ascites, increased bilirubin and prolonged INR) and is associated with 60% of mortality, and the later, with a better prognosis (10% of mortality) is manifested without ascites but with efficient collateral formation (*5,17*).

We performed a systematic review in PubMed using the terms "Budd-Chiari syndrome", "Behçet's disease" and "Behçet's syndrome" in English and Spanish languages on the 10th of January 2019 and 65 articles were found. We excluded case reports, review articles and case series of BD with incomplete information of the subgroup of patients with BCS. Therefore, ten articles were included (Table 1 and Table 2). The vast majority were men (89.5%) with a mean age between 18 and 41 years. The most common clinical features were ascites (46-100%), abdominal pain (40-100%), hepato-splenomegaly (29-100%), abdominal collateral circulation (25-100%), jaundice (19-100%) and fever (16-50%). Our patient had most of those symptoms (abdominal pain, fever and ascites). Interestingly, clinical findings of the patients are similar to our case report, however, the management has evolved over time. In other words, surgical management has decreased after the beginning of the use of immunosuppressants. Immunosuppressants and biological therapy are the most common therapeutic options in the last decade for BCS and BD (*7,17-25*).

Immunosuppressants, with or without glucocorticoids, are essential in the management of vascular involvement in BD. They have been shown to reduce the relapse rate and to prolong survival in several retrospective studies. In patients with BD, conditions associated with higher mortality like BCS require an early and aggressive medical treatment, including cyclophosphamide and glucocorticoid pulses. In resistant cases, anti-tumor necrosis factor (TNF) agents could also be effective (*1,4,6,8*). Whether to add anticoagulants to prevent recurrent thrombosis has been debated (*5,7*). Several retrospective studies showed the inefficacy of anticoagulation alone or added to immunosuppressants in preventing recurrences (*8*). Anticoagulation could increase the risk of aneurysmal rupture (*6,9,10*). Nevertheless, the tolerance of anticoagulation therapy was satisfactory in patients with low bleeding risk after ruling out pulmonary artery aneurysms and it could be used in refractory venous thrombosis (*4,6*).

In the last decade, several studies have demonstrated a survival improvement with the use of angioplasty/stenting or TIPS in patients with BCS, remarking the use of TIPS as a definitive treatment prior to liver transplantation, and not only as a bridging treatment (*11,12*). However, there is very limited experience in patients with BCS and BD (only a few case reports). A case of a 45-y-o male with BD presented with acute BCS and was treated with percutaneous transluminal angioplasty showing a dramatic reduction of portal venous pressure. Immunosuppressive agents and anticoagulation were used for prevention of recurrent thrombosis (*26*). A case series reported 5 patients with BD and acute BCS showing reversal of liver damage and correction of hemodynamic disturbances, prolonged survival and good quality of life when side-to-side portacaval shunt was performed early in the course of BCS (*22*). There is no specific mention about the role of TIPS in the subgroup of BCS associated with BD in the latest update of the EULAR (European League Against Rheumatism) recommendations (*6,12*). In addition, it is important to note the risk of vascular pathergy phenomena after manipulating vessels in patients with BD, triggering vascular inflammation and consequently extension of the thrombosis (*5,9,10*). This is an important question that needs to be answered, taking into account the high mortality of BCS in the setting of BD and the management of BCS of any etiology includes TIPS for the most severe cases.

We suggest an algorithm for the management of BCS in the setting of BD (Figure 2). In case of BCS without a known etiology, every patient should undergo a quick revision of the clinical criteria for BD, especially if the patient is young (< 35 y-o). Once BD diagnosis is established, we should consider we are facing a case of vascular involvement of BD. BCS is a severe manifestation of vascular involvement of BD, and therefore it should be treated promptly and aggressively. The recommended treatment for BCS in

Table 1. Clinical features of patients with Budd-Chiari syndrome associated with Behçet's disease (case series published in literature)

Author (year) (*ref.*)	N	Mean age	Male (%)	Symptoms and Signs (%)						
				Abdominal pain	Ascitis	Jaundice	Hepato-splenomegaly	Fever	Abdominal collateral circulation	Portal thrombosis
Seyahi *et al*. (2015) (*17*)	43	30	40/43 (93)	17/43 (40)	33/43 (77)	8/43 (19)	29/43 (71)	7/43 (16)	27/43 (63)	0/43
Desbois *et al*. (2014) (*7*)	14	33	11/14 (79)	6/13 (46)	6/13 (46)	-	6/13 (46)	-	-	-
Ben Ghorbel *et al.* (2008) (*18*)	7	29	7/7 (100)	-	6/7 (86)	1/7 (14)	7/7 (100)	-	7/7 (100)	-
Seyahi *et al.* (2007) (*19*)	3	18	3/3 (100)	2/3 (67)	2/3 (67)	-	2/3 (67)	-	-	-
Korkmaz *et al.* (2007) (*20*)	4	26	4/4 (100)	4/4 (100)	4/4 (100)	4/4 (100)	4/4 (100)	-	-	-
Kuniyoshi *et al.* (2002) (*21*)	2	41	1/2 (50)	2/2 (100)	2/2 (100)	-	1/2 (50)	1/2 (50)	-	-
Orloff *et al.* (1999) (*22*)	5	25	4/5 (80)	5/5 (100)	5/5 (100)	3/5 (60)	5/5 (100)	-	5/5 (100)	0/5 (0)
Bayraktar (1997) (*23*)	14	37	12/14 (86)	-	10/14 (71)	-	4/14 (29)	-	2/8 (25)	4/14 (29)
al-Dalaan *et al.* (1991) (*24*)	3	29	2/3 (67)	3/3 (100)	3/3 (100)	-	2/3 (67)	1/3 (33)	1/3 (33)	1/3 (33)
Bismuth *et al.* (1990) (*25*)	20	29	19/20 (95)	3/-	19/20 (95)	9/20 (45)	2/-	-	-	0/20 (0)

Table 2. Management of patients with Budd-Chiari syndrome associated with Behçet's disease (case series published in literature)

Author (year) (*ref.*)	N	Management							Mortality (%)
		AC	Colchicine	IS	anti-TNF	CS	Interventionist	Thrombolytic	
Seyahi *et al*. (2015) (*17*)	43	14/43	19/33	36/43	5/43	34/43	2/43 Angioplasty, 1/43 TIPS	5/43	20/43 (47)
Desbois *et al*. (2014) (*7*)	14	14/14	1/14	12/14	4/14	12/14	2/14 Endovascular, 1/14 OLT, 1/14 Surgery♦	3/14	2/14 (14)
Ben Ghorbel *et al.* (2008) (*18*)	7	7/7	-	2/7	0	6/7	0	-	1/7 (14)
Seyahi *et al.* (2007) (*19*)	3	0	0	3/3	3/3	3/3	0	0	2/3 (67)
Korkmaz *et al.* (2007) (*20*)	4	4/4	2/4	4/4	-	4/4	0	-	1/4 (25)
Kuniyoshi *et al.* (2002) (*21*)	2	1/2	-	0	-	1/2	2/2 Surgery♦	-	1/2 (50)
Orloff *et al.* (1999) (*22*)	5	5/5	-	-	-	-	5/5 SSPCS	-	1/5 (20)
Bayraktar (1997) (*23*)	14	-	-	-	-	-	-	-	10/14 (71)
al-Dalaan *et al.* (1991) (*24*)	3	3	-	1	-	3	2/3 Surgery♦	-	1/3 (33)
Bismuth *et al.* (1990) (*25*)	20	15/20	15/20	-		15/20	5/20 Surgery♦	-	9/17 (52)

♦Includes the following techniques: peritoneovenous shunt, mesocaval or mesoatrial shunting, portosystemic shunt. AC, anticoagulation; IS: immunosuppressants; anti-TNF, anti tumoral necrosis factor; CS, corticosteroids; OLT, orthotopic liver transplantation; SSPCS, side-to-side portacaval shunt; TIPS, transjugular intrahepatic portosystemic shunt.

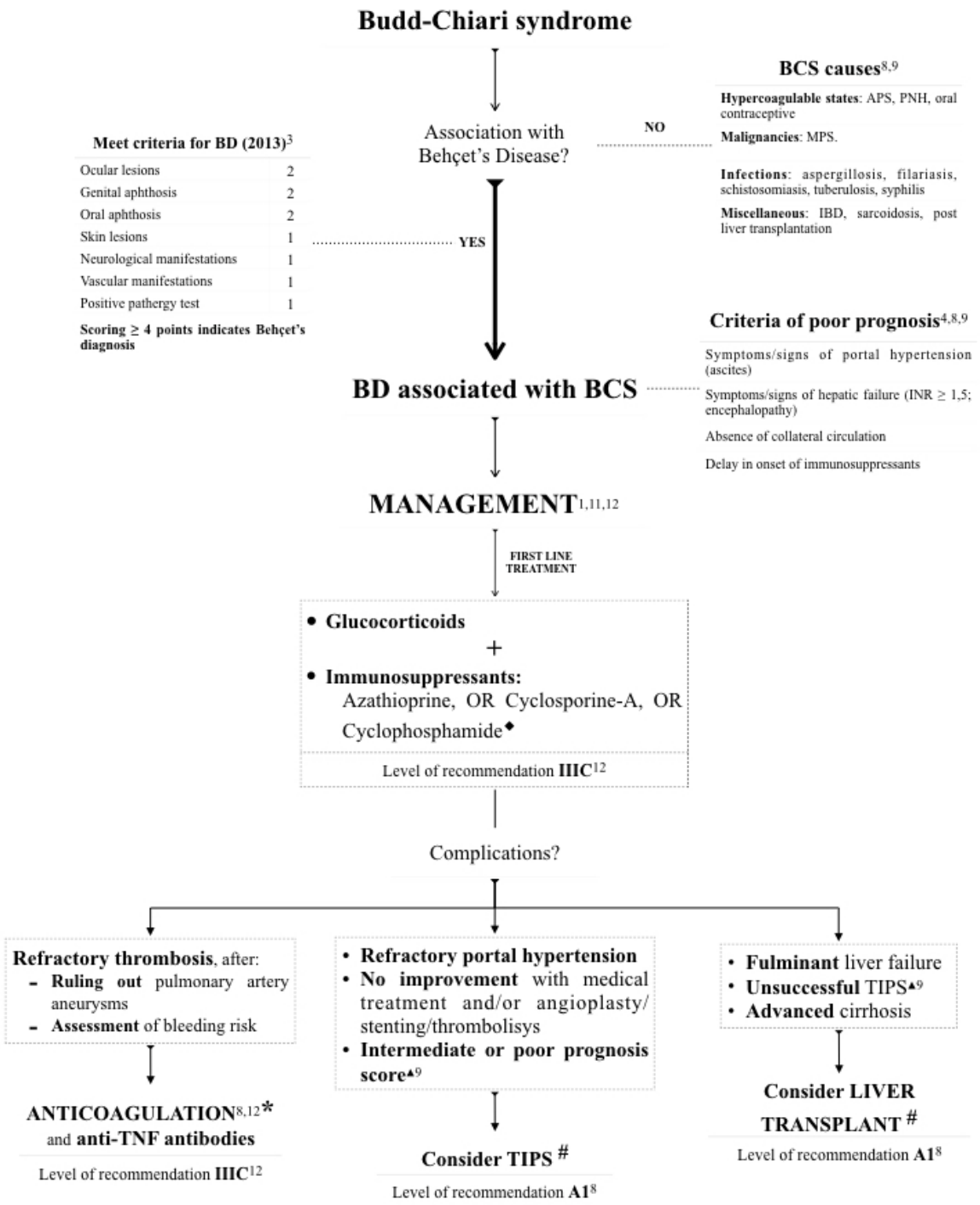

Figure 2. Algorithm for management of Budd-Chiari syndrome in Behçet's disease. ♦Cyclophosphamide may be reserved for patients with extensive thrombosis or larger veins like IVC due to potential adverse events. *Level of recommendation for anticoagulation is A1 in BCS according EASL guidelines, but level of recommendation for anticoagulation is IIIC in BCS associated with BD according to EULAR guidelines. ▲There have been proposed three prognostic scores: the Rotterdam BCS score, the BCS-TIPS index score, and the revised Clichy score. ●Consider angioplasty/stenting as the first line decompressive procedure in patients with short hepatic vein stenosis or IVC stenosis. If there is no response TIPS is the treatment of choice (EASL 2015 for all causes of BCS). #Individualized decision is recommended due to the absence of specific evidence for BCS in the setting of BD; Consider the risk of vascular pathergy phenomenon in BD. anti-TNF antibodies, anti tumoral necrosis factor antibodies; APS, antiphospholipid syndrome; BCS, Budd-Chiari syndrome; BD, Behçet disease; DVT, deep vein thrombosis; IBD, Inflammatory bowel disease; IVC, inferior vena cava; MPS, myeloproliferative syndrome; PNH, Paroxysmal nocturnal hemoglobinuria; TIPS, Transjugular intrahepatic portosystemic shunt.

BD is to start glucocorticoids and immunosuppressants. In case of no response, various options are possible, including anticoagulation and invasive procedures (Figure 2).

BD usually has a diagnosis delay of several years since the first medical visit. In our case, the presentation was an acute symptomatic BCS. The diagnostic process was quick and allowed a prompt therapeutic intervention.

Different therapeutic options were proposed, including anticoagulants, immunosuppressants and TIPS. We opted for intravenous heparin and a high-dose pulse of intravenous corticosteroids and intravenous cyclophosphamide and rejected TIPS due to the lack of evidence of this management in BD and the fact that could trigger a vascular pathergy phenomenon. Despite that our patient had an acute symptomatic presentation and liver failure, she fortunately showed a satisfactory and rapid response to the treatment.

In conclusion, acute BCS presented in the setting of BD is a severe condition with high mortality. Vascular involvement in BD is managed with immunosuppression. Other therapeutic options like anticoagulation or interventional vascular therapy have a secondary role and should be considered on an individual basis.

References

1. Yazici H, Seyahi E, Hatemi G, Yazici Y. Behçet syndrome: A contemporary view. Nat Rev Rheumatol. 2018; 14:107-119.
2. Criteria for diagnosis of Behçet's disease. International Study Group for Behçet's Disease. Lancet. 1990; 335:1078-1080.
3. International Team for the Revision of the International Criteria for Behçet's Disease (ITR-ICBD). The International Criteria for Behçet's Disease (ICBD): A collaborative study of 27 countries on the sensitivity and specificity of the new criteria. J Eur Acad Dermatol Venereol. 2014; 28: 338-347.
4. Emmi G, Bettiol A, Silvestri E, Di Scala G, Becatti M, Fiorillo C, Prisco D. Vascular Behçet's syndrome: An update. Intern Emerg Med. 2018; Nov 29.
5. Seyahi E. Behçet's disease: How to diagnose and treat vascular involvement. Best Pract Res Clin Rheumatol. 2016; 30: 279-295.
6. Hatemi G, Christensen R, Bang D, *et al*. 2018 update of the EULAR recommendations for the management of Behçet's syndrome. Ann Rheum Dis. 2018; 77:808-818.
7. Desbois AC, Rautou PE, Biard L, Belmatoug N, Wechsler B, Resche-Rigon M, Zarrouk V, Fantin B, de Chambrun MP, Cacoub P, Valla D, Saadoun D, Plessier A. Behcet's disease in Budd-Chiari syndrome. Orphanet J Rare Dis. 2014 Sep13; 9:104.
8. Saleh Z, Arayssi T. Uptodate on the therapy of Behçet Disease. Ther Adv Chronic Dis. 2014; 5:112-134.
9. Wang H, Guo X, Tian Z, Liu Y, Wang Q, Li M, Zeng X, Fang Q. Intracardiac thrombus in patients with Behcet's disease: Clinical correlates, imaging features, and outcome: A retrospective, single-center experience. Clin Rheumatol. 2016; 35:2501-2507.
10. Galeano-Valle F, Demelo-Rodríguez P, Álvarez-Sala-Walther L, Pinilla-Llorente B, Echenagusia-Boyra MJ, Rodriguez-Abella H, Del-Toro-Cervera J. Intracardiac thrombosis in Behçet's Disease successfully treated with immunosuppressive agents: A case of vascular pathergy phenomenon. Intractable Rare Dis Res. 2018; 7:54-57.
11. Garcia-Pagán JC, Heydtmann M, Raffa S, *et al*. TIPS for Budd-Chiari syndrome: Long-term results and prognostics factors in 124 patients. Gastroenterology. 2008; 135:808-815.
12. European Association for the Study of the Liver. EASL Clinical Practice Guidelines: Vascular diseases of the liver. J Hepatol. 2016; 64:179-202.
13. Park MC, Hong BK, Kwon HM, Hong YS. Surgical outcomes and risk factors for postoperative complications in patients with Behcet's disease. Clin Rheumatol. 2007; 26:1475-1480.
14. Demelo-Rodríguez P, Del Toro-Cervera J. Platypnea-orthodeoxia syndrome and Budd-Chiari syndrome: An unreported association. Med Clin (Barc). 2015; 144: 94-95. (in Spanish)
15. Valla DC. Primary Budd-Chiari syndrome. J Hepatol. 2009; 50:195-203.
16. Goel RM, Johnston EL, Patel KV, Wong T. Budd-Chiari syndrome: Investigation, treatment and outcomes. Postgrad Med J. 2015; 91: 692-697.
17. Seyahi E, Caglar E, Ugurlu S, Kantarci F, Hamuryudan V, Sonsuz A, Melikoglu M, Yurdakul S, Yazici H. An outcome survey of 43 patients with Budd-Chiari syndrome due to Behçet's syndrome followed up at a single, dedicated center. Semin Arthritis Rheum. 2015; 44:602-609.
18. Ben Ghorbel I, Ennaifer R, Lamloum M, Khanfir M, Miled M, Houman MH. Budd-Chiari syndrome associated with Behçet's disease. Gastroenterol Clin Biol. 2008; 32: 316-320.
19. Seyahi E, Hamuryudan V, Hatemi G, Melikoglu M, Celik S, Fresko I, Yurdakul S, Yazici H. Infliximab in the treatment of hepatic vein thrombosis (Budd-Chiari syndrome) in three patients with Behcet's syndrome. Rheumatology (Oxford). 2007; 46: 1213-1214.
20. Korkmaz C, Kasifoglu T, Kebapçi M. Budd-Chiari syndrome in the course of Behcet's disease: Clinical and laboratory analysis of four cases. Joint Bone Spine. 2007; 74:245-248.
21. Kuniyoshi Y, Koja K, Miyagi K, Uezu T, Yamashiro S, Arakaki K, Mabuni K, Senaha S. Surgical treatment of Budd-Chiari syndrome induced by Behcet's disease. Ann Thorac Cardiovasc Surg. 2002; 8:374-380.
22. Orloff LA, Orloff MJ. Budd-Chiari syndrome caused by Behçet's disease: Treatment by side-to-side portacaval shunt. J Am Coll Surg. 1999; 188:396-407.
23. Bayraktar Y, Balkanci F, Bayraktar M, Calguneri M. Budd-Chiari syndrome: A common complication of Behçet's disease. Am J Gastroenterol. 1997; 92: 858-862.
24. al-Dalaan A, al-Balaa S, Ali MA, Huraib S, Amin T, al-Maziad A, al-Fadda M. Budd-Chiari syndrome in association with Behçet's disease. J Rheumatol. 1991; 18:622-626.
25. Bismuth E, Hadengue A, Hammel P, Benhamou JP. Hepatic vein thrombosis in Behçet's disease. Hepatology. 1990; 11:969-974.
26. Han SW, Kim GW, Lee J, Kim YJ, Kang YM. Successful treatment with stent angioplasty for Budd-Chiari syndrome in Behçet's disease. Rheumatol Int. 2005; 25:234-237.

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