Behçet’s Disease with Vascular Involvement: Case Report

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Behçet’s disease is an inflammatory, multisystemic, relapsing syndrome of unknown etiology. It involves heterogeneous clinical manifestations such as: recurrent oral ulcers, ocular inflammation, genital ulcers, and skin lesions. More severe manifestations may occur due to vasculitis of small and large arteries and/or veins with formation of arterial aneurysms or thrombosis, in addition to neurological or gastrointestinal involvement. Diagnosis is clinical and mainly symptomatic, and treatment may include corticosteroids with or without immunosuppressants and, eventually, other interventions for more severe manifestations.

Clinical case

Male patient, 34 years of age, previously healthy, admitted to the Emergency Room (ER) of Instituto Dante Pazzanese de Cardiologia reporting a 3-month history of pain in the right lower limb (RLL) and limping for distances of 100 meters in the 3 weeks prior to admission. The patient evolved with worsening pain in the right calf and spontaneous pulsatile bulging a week before admission. He mentioned the use of Rivaroxaban for deep venous thrombosis (DVT) in the RLL five months before, weight loss (15kg in 6 months), and appearance of aphthous lesions in the oropharyngeal region. RLL pulses were present, except for the tibialis posterior with a slight thermal gradient in the right foot. A new vascular ultrasound examination showed occlusion of the TPT endoprosthesis (Figure 1D). Clinical follow-up and reintroduction of Rivaroxaban 20mg plus maintenance of ASA were carried out.

At the outpatient follow-up, after two weeks, the patient reported improvement in pain in the RLL, however accompanied by worsening of pain in the right foot, associated with paresthesia, coldness, and stable oral lesions. RLL pulses were present, except for the tibialis posterior with a slight thermal gradient in the right foot. A new vascular ultrasound examination showed thrombus in the right peroneal artery (Figure 1C). The patient was discharged with a prescription for Clopidogrel (75mg/day) and ASA (100mg/day); the use of Rivaroxaban was discontinued.

After 3 weeks, the patient was readmitted to the ER reporting a new, spontaneous, pulsating bulge in the right thigh, with progressive increase associated with significant pain at the site and impaired walking. The complaint of paresthesia in the right foot persisted. He also reported worsening of the aphthous lesions in the oral cavity and the appearance of ulcerated lesions in the scrotal region (Figures 2A and 2B). Physical examination indicated a large pulsating bulge in the distal right thigh and loss of pedal pulse. A new vascular echography with RLL Doppler was performed, which showed, in the right superficial femoral artery (RSFA), the presence of a pseudoaneurysm of the posterior wall, in the distal segment, measuring approximately 3.06 cm x 4.01 cm (AP x LL), in addition to another pseudoaneurysm measuring approximately 0.5 cm x 0.8 cm, 2.5 cm away from the femoral bifurcation, and two bulges in the posterior wall with apparent intimal rupture, maintaining the integrity of the adventitia. Images of partially thrombosed pseudoaneurysms were observed in the right (RCFA) and left common femoral arteries (LCFA) measuring approximately 1.1 cm and 1.0 cm, respectively. He also presented intraluminal hypoechogenic material in the right femoral vein (RFV) and right popliteal vein (RPV), compatible with acute DVT. Ultrasound findings are shown in Figures 3A–F.

Laboratory tests: normal white blood cell count, hypochromic and microcytic anemia — Hemoglobin (Hb) = 10.9 g/dL, Mean Corpuscular Volume (MCV) = 75.9, Mean Corpuscular Hemoglobin Concentration (MCHC) = 31.5 —, C-reactive protein (CRP), erythrocyte sedimentation rate (ESR) and high fibrinogen (CRP = 25.2 mg/dL, ESR = 44mm/h, and fibrinogen= 636mg/dL); high values of oxaloacetic transaminase (TGO) = 233 U/L, pyruvic transaminase (TGP) = 153 U/L, gamma glutamyl transferase (GGT) = 490 U/L; and alkaline phosphatase (FA) = 264 U/L; negative serologies for hepatitis and syphilis.
The hypothesis of autoimmune vasculitis was suggested. Given the disease activity, no invasive investigation was performed at that time and a rheumatological evaluation was requested. Considering the arterial and venous involvement — arterial pseudoaneurysms and venous thrombosis, elevated inflammatory tests, and oral and genital ulcers —, the diagnosis of Behçet’s disease was concluded. Pulse therapy with methylprednisolone and cyclophosphamide was onset. There was a reduction in symptoms, oral and genital ulcers, and inflammatory evidence after the first session (Figure 2C). In reassessment, after the second session of pulse therapy (30 days), a new intervention in RLL was indicated.

Endovascular treatment of the RSFA pseudoaneurysm was then performed with Viabahn Gore (7 mm x 100 mm), with good therapeutic results (Figures 4A-D). The patient had an uneventful postoperative period and was discharged from the hospital.

Discussion

Behçet’s disease consists of a multisystem inflammatory process of unknown etiology characterized by recurrent episodes of oral and genital ulcers, other skin lesions, and ocular lesions. It was first described by Hülüsî Behçet in 1937 as a triad of oral, genital, and uveitis ulcers.¹

Currently, it is known that the involvement goes beyond this triad and extends to several systems, including neurological, pulmonary, gastrointestinal, cardiac, articular, and vascular. Manifestations are not the same in all patients, clinical phenotypes are very heterogeneous and disease progression varies according to ethnicity, geography, and individual differences.²

The increase in evidence indicating the possibility of immunological mechanisms in the pathogenesis of the disease suggests an autoimmune etiology. Vascular alterations, for example, result from endothelial cell dysfunction caused by a probable immune-mediated reaction. Vasculitis appears to be the pathogenic basis of the various systemic manifestations.³
Series of patients confirmed that young men are more prone to vascular involvement of the disease. Large vessel disease is one of the manifestations associated with systemic symptoms and laboratory evidence of acute phase response. There is no specific laboratory test for Behçet’s disease, but the presence of the HLAB-51 gene is suggestive of the disease.

The arteries most affected by the formation of aneurysms are the pulmonary, femoral, iliac, aorta, and popliteal arteries. The main pathological findings in the aneurysm wall are: thickening of the adventitia, fibrosis, perivascular lymphocytic infiltration, decrease in muscle and elastic fibers in the media layer, and increase in spongy cells and fibroblasts in the intimal layer. The inflammatory process is acute and causes destruction of the arterial wall, resulting in rapid formation of aneurysms or pseudoaneurysms, increasing the incidence of rupture and bleeding.

Venous involvement is the most frequent (88%), while arterial involvement is responsible for a smaller number of cases (12%). Venous thrombosis is the most common finding, which can occur in different territories and progress despite anticoagulation. Thrombi, aneurysms or pseudoaneurysms can occur as complications of surgery or invasive procedures. Treatment is based on immunosuppression. Anticoagulants are used in the treatment and prevention of thromboembolic phenomena and surgical or endovascular procedures may be necessary.

The use of endovascular repair has been increasing in Behçet’s disease, with the aim of reducing complications resulting from surgical trauma. Surgical treatment in the acute phase of the disease is associated with higher rates of thrombosis and suture dehiscence. It is suggested that the combined use of corticosteroids and immunosuppressants, and possibly the use of antiplatelet agents or anticoagulants, may be effective in reducing complications associated to the procedures.

Aneurysms and pseudoaneurysms of the distal arteries are infrequent in these patients and each case must be evaluated individually. The literature describes few cases of
TPT involvement in Behçet’s disease. The main advantage of the endovascular technique is that it is less invasive and allows access to the affected area from a different site, without direct manipulation.

Author Contributions
Conception and design of the research: Tavares IR, Petisco ACGP, Folino CB, Dornas VLBL, Saleh MH, Rossi FH, de Sousa RP; acquisition of data: Tavares IR, Miranda MC, Petisco ACGP, Barros DS, Folino CB, Dornas VLBL, de Sousa RP, de Carvalho LCN; analysis and interpretation of the data: Miranda MC, Petisco ACGP, de Carvalho LCN; statistical analysis: Tavares IR; writing of the manuscript: Tavares IR, Miranda MC, Petisco ACGP, Barros DS; critical revision of the manuscript for intellectual content: Petisco ACGP, Saleh MH, Rossi FH.

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This article does not contain any studies with human participants or animals performed by any of the authors.

References

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