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Case Report

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Carotid Dissection Secondary to Neuro-Behcet's Disease - Case Report

Frazao IC[1*], Linford HT[2], Bronner GE[1], Massaro CDS[1], and Henning PR[1]

[1]Federal University of Mato Grosso do Sul-UFMS, Clerkship at the Neurology Service of UFMS, Campo Grande, MS, Brazil
[2]Campbell University, SOM, Buies Creek, NC, USA

[*]**Corresponding Author:** Frazao IC, Federal University of Mato Grosso do Sul-UFMS, Clerkship at the Neurology Service of UFMS, Campo Grande, MS, Brazil, Tel: 910-670-4754, E-mail: ii.ca.fz@gmail.com

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Abstract

Objectives: This objective of this article is to describe by case report, the diagnosis and treatment carried out in a case of stroke due to internal carotid dissection in a Neuro-Behcet patient, with cardiological manifestations due to vasculitis. This paper also presents a flowchart to summarize the approach and treatment of this case to aid others that might be facing the same rare disease.

Method: E.F.B., 43 years old, male, with history of smoking and untreated hypertension, was admitted at the University Hospital of Campo Grande (HUMAP) due to right hemiparesis and hemiparesthesia, associated with hemicranial headache with blurred vision. Patient evolved with acute myocardial infarction (AMI).

Results: Brain CT showed hypodensity in left thalamus–nuclear region. Cerebral arteriography revealed left internal carotid dissection. EKG showed new left bundle branch block (LBBB), and blood work with troponin elevations. Rheumatologic interconsult corroborated with Behcet diagnosis, and treatment with corticoid pulse therapy and specific cardiology and neurology protocol was started.

Conclusion: The neuro and cardiac Behcet's disease diagnosis is predominantly clinical. Therefore, the anamnesis focused in clinical presentations of disease, especially oral and genital ulcers, is important to the early diagnosis and proper treatment. The vasculitis causing neurologic and cardiologic findings was controlled with corticoid pulse therapy treatment.

Keywords: *Neuro-Behcet; Carotid Dissection; Pulse corticosteroid therapy; Cardiac Behcet's disease; Acute myocardial infarction*

Introduction

Hulusi Behçet, a Turkish dermatologist, described the triad of recurrent oral and genital ulcers with uveitis in 1937. Behçet disease (BD) is more prevalent along the ancient Silk Road, including countries in the Far East, the Middle East, and the Mediterranean basin [1]. The incidence of BD in America is rare [2]. This difference may be attributed to genetics factors, but also may be due to underdiagnosis of BD in the west [2].

BD is a remitting-relapsing, multisystem, autoimmune disease that may be associated with HLA-B51 and triggered by environmental factors [3]. Venous pathology and thrombotic complications are hallmarks of BD. However, it has been increasingly recognized that cardiac involvement and arterial complications are also important aspects of the course of the disease [4].

Diagnostic criteria include recurrent oral ulcers plus two of the following: Genital ulcers; Skin lesions; Eye injuries; Positive pathergy test [5].

Neurological involvement may occur and can lead to high morbidity and mortality, often affecting males [2,3]. Neuro-BD may be parenchymal, non-parenchymal or a mixed brain disease [6-8].

Cardiac involvement in BD is also referred to as cardiac BD. Cardiac involvement may occur in the form of intracardiac thrombus, endocarditis, myocarditis, pericarditis, endomyocardial fibrosis, coronary arteritis, myocardial infarction, and valvular disease [4].

The incidence of large arterial complications, like carotid dissection, is rare and ranges from 2% to 6% in patients from 20 to 40 years of age with Behçet's disease [9-11]. Our case report includes unilateral carotid dissection associated with coronary artery involvement. The presentation of arterial complications can vary as demonstrated by a case report in 2002 that presented a patient with BD who developed bilateral internal carotid artery (ICA) occlusions and another report in 2005 showed a large pseudoaneurysm of the carotid artery in a BD patient [9,12].

The objective of this paper is to report, through a clinical case, the diagnosis and treatment realized in a case of Behcet's disease with neurological and cardiological manifestations. The disease resulted in stroke due to carotid dissection and myocardial infarction, caused by vasculitis in the carotid and coronary arteries. A flowchart is included to summarize the approach and treatment of this rare disease.

Patients and Methods

E.F.B., a 43 year old male, light smoker for 22 years, with previous history of untreated hypertension, was checked in the Emergency Care Unit of the University Hospital of Campo Grande (HUMAP) in July of 2015, due to subtle right hemiparesis and hemiparesthesia. The patient also presented with intense left hemicranial headache, described as pulsatile, with blurred vision that started one hour before the loss of strength in the right arm. The headache responded to dipirone.

The patient was responsive, time and space-oriented, complaining of blurring vision, headache, and weakness in the right arm and leg. At the neurological examination, the patient presented with paresthesia in the right superior and inferior limbs, decrease of strength in the right arm 4/5, and hypertonia of right upper limb. In addition to these symptoms, anisocoria and ptosis on the right eye were present. No other abnormalities were registered.

Brain CT showed hypodensity in left thalamus–nuclear region. Cerebral arteriography revealed left internal carotid dissection (Figure 1).

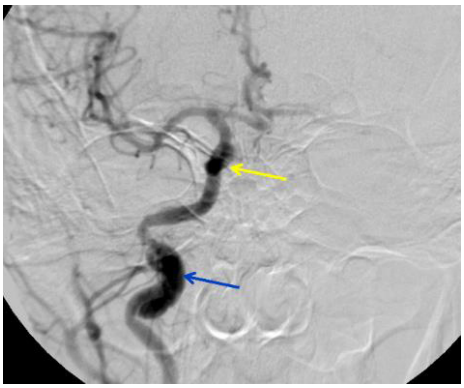


Figure 1: Cerebral Arteriography

Arteriography, revealing spontaneous dissection of the left internal carotid artery with 90-95% stenosis (blue arrow) and fusiform aneurismatic dilation in C3-C4 segment (yellow arrow).

During further investigation, the patient revealed a history of intermittent headaches for 10 years, associated with oral ulcers, genital ulcers, joint pain and blurred vision without motor deficit. There was no history of photosensitivity, Raynaud's phenomenon and previous thrombosis.

Rheumatologic interconsult corroborated with BD clinical diagnosis, and treatment with corticoid pulse therapy was started.

During the hospitalization, patient evolved with acute myocardial infarct (AMI), diagnosed by new left bundle branch block (LBBB) and elevated troponin levels. The patient was treated with specific cardiologic protocol.

The patient remained hospitalized until August 2015, with improved global condition, being discharged home with referral for rehabilitation, cardiologic and rheumatologic follow up.

In view of the results of the requested tests and evaluation of Rheumatology, the patient was diagnosed with Neuro and cardiac Behcet's disease. He was prescribed oral Enalapril at the dose of 10mg/day, Metoprolol 100mg/day, Warfarin 5mg/day, Aspirin 100 mg/day, Simvastatin 40 mg/day, Clopidogrel 75 mg/day, continuously. He is currently under follow-up with cardiology, neurology and rheumatology. He is indicating a significant improvement in his quality of life, and remains stable.

Discussion

Behcet's disease consists of vasa vasorum vasculitis of large vessels associated with hypercoagulable state. Lesions occur in vessels in approximately 25% of cases, primarily in the venous system, however arterial lesions can occur and are more severe [13].

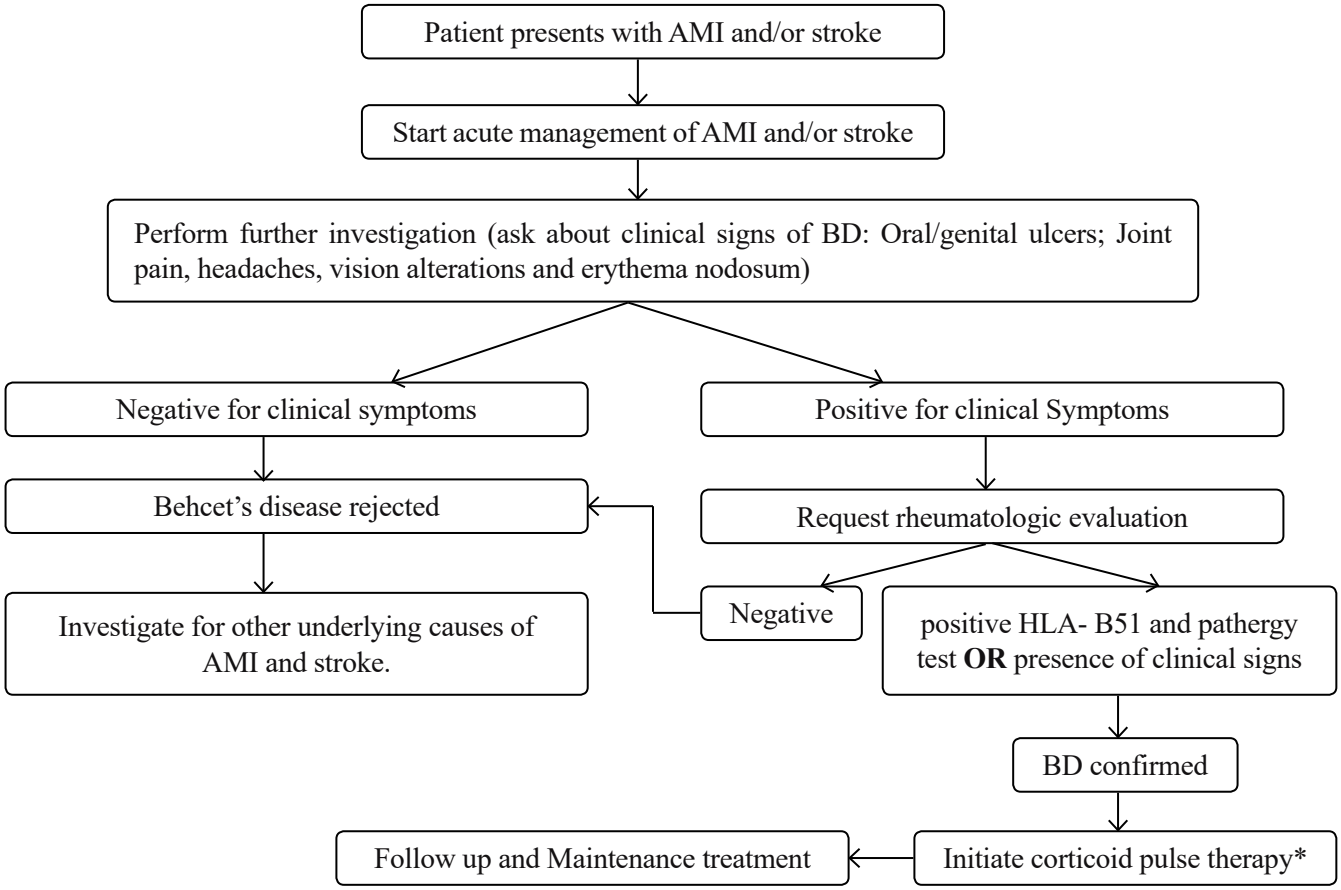
In BD the neurological involvement occurs in 5% to 25% of patients and it develops after the manifestation of the syndrome [2,13]. The parenchymal form is the most common, occurring in 81% of cases and often manifests, with subacute meningoencephalitis and rarely stroke [14]. In non-parenchymal, the main clinical presentations are cerebral venous thrombosis, intracranial hypertension, intracranial aneurysm and extracranial aneurysm/dissection [15].

After the first event, there is higher risk of new events, worsening the prognosis, and demands more aggressive treatment [2,13].

The purpose of the treatment is to control symptoms, improve the patient's quality of life, prevent complications, and when possible, the avoidance of the progression and recurrence of the disease. Regarding BD, the objective is to decrease the vasculitis to near normality, in order to cease symptoms.

Corticoid pulse therapy has proven to be efficient in patients with Behcet's disease. It is important to request a PPD test before the start of corticoid pulse therapy, to avoid reactivating latent Tuberculosis (Appendix 1).

In order to summarize and aid others in the management of this severe complication in this rare disease, the following flowchart was created:



- BD: Behcet's Disease.
- AMI: Acute myocardial infarct.

*High-dose intravenous methylprednisolone for example.

Conclusion

The neuro and cardiac Behcet's disease diagnosis are predominantly clinical. Therefore, the anamnesis focused in clinical presentations of disease, especially oral and genital ulcers, is very important to the early diagnosis and proper treatment with corticoid pulse therapy to prevent future events and reduce sequelae. In this case, the vasculitis causing neurologic and cardiologic findings was successfully controlled with corticoid pulse therapy treatment.

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