Annals of Clinical Case Reports

Case Report
Published: 12 Apr, 2023

Neuro-Behcet Disease Presenting as a Bilateral Thalamic Infarct Secondary to Cerebral Vein Thrombosis and Multiple Cerebral Arteritis; A Case Report

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Abstract

Background: Behcet's Disease is a systemic autoimmune disease characterized by chronic inflammation primarily affecting the eyes, mouth, and genitals. Its prevalence is very rare in Southeast Asia with only 31 recorded cases from 1988 to 2014 in a cluster of rheumatology clinics. The involvement of the central nervous system in Bechet's Disease is termed as Neuro Behcet's Disease, with a prevalence of 4% to 9%. However, prevalence of Neuro-Behcet disease in female is uncommon. CNS involvement mimics other conditions such as neoplasm, infection, and stroke. Herein, we report a rare case of Neuro-Behcet disease who presented with bilateral thalamic infarct from deep cerebral vein thrombosis and arteritis.

Case Report: A 34-year-old Filipino female was admitted to our hospital due to progressive headache and somnolence with a significant history of recurrent genital ulcers and arthritis. Her CT scan showed ischemic stroke on bilateral thalami with MRI and vascular study of subacute hemorrhagic conversion from thrombosis on vein of Galen and straight sinus with multiple segmental arteritis. Other causes of thrombosis such as neoplasm, cardiovascular etiology, and other autoimmune diseases were ruled out. Due to the absence of pathognomonic laboratory features, the patient was diagnosed with Neuro-Behcet's Disease after satisfying the clinical criteria. Clinical and radiographic improvement was noted after initiation of treatment with steroid, azathioprine, and anti-coagulant.

Conclusion: These findings suggest that the patient's bilateral thalamic infarct could have been due to cerebral vein thrombosis and segmental arteritis as a rare complication of Neuro-Behcet Disease.

Keywords: Cerebral Venous Sinus thrombosis; Neuro-Behcet; Neuroimmunology

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Received Date: *29 Mar 2023*
Accepted Date: *07 Apr 2023*
Published Date: *12 Apr 2023*

Citation:
Rabadon CR, Juangco DN. Neuro-Behcet Disease Presenting as a Bilateral Thalamic Infarct Secondary to Cerebral Vein Thrombosis and Multiple Cerebral Arteritis; A Case Report. Ann Clin Case Rep. 2023; 8: 2406.
ISSN: 2474-1655.

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Introduction

Behcet's Disease is a multisystem auto-inflammatory disease characterized by triple symptom complex of recurrent oral ulcers, genital ulcers, and uveitis [1]. It typically runs a relapsing and remitting course [2]. Disease etiology remains unclear but advances in genetics and immunology have led to a better understanding of the immunopathogenesis. A link with Human Leukocyte antigen B51 has been proposed, however, only 50% of patients were positive [3].

Among the extracutaneous manifestations, nervous system involvement is rare. Neurologic involvement is low with a prevalence of 4% to 10% and it predominantly affects males with a sex ratio of 2-5:1 and onset peaks during the 2nd and 3rd decade of life [4]. Neuro-Behcet's disease is a combination of neurologic signs and symptoms in patients with definite Behcet's Disease (BD). It generally occurs following the diagnosis of BD, but it can be the initial presentation of the disease.

Clinical and neuroimaging findings demonstrate two forms of Neuro-Bechet: The more common parenchymal disease (81%), and the less common nonparenchymal form (19%). Non-parenchymal disease tends to involve the veins more than arteries [4].

Case Presentation

A 34-year-old Filipino female was admitted to our hospital due to history of progressive headache for 4 days. Headache was described as dull in character, with pain scale of 6/10 at the temporo-occipital area without any history of trauma. On the day of admission, her sensorium deteriorated and became somnolent. She had a history of recurrent joint pains and genital ulcerations which spontaneously resolved even without treatment for almost 2 years. No history of mouth sores,

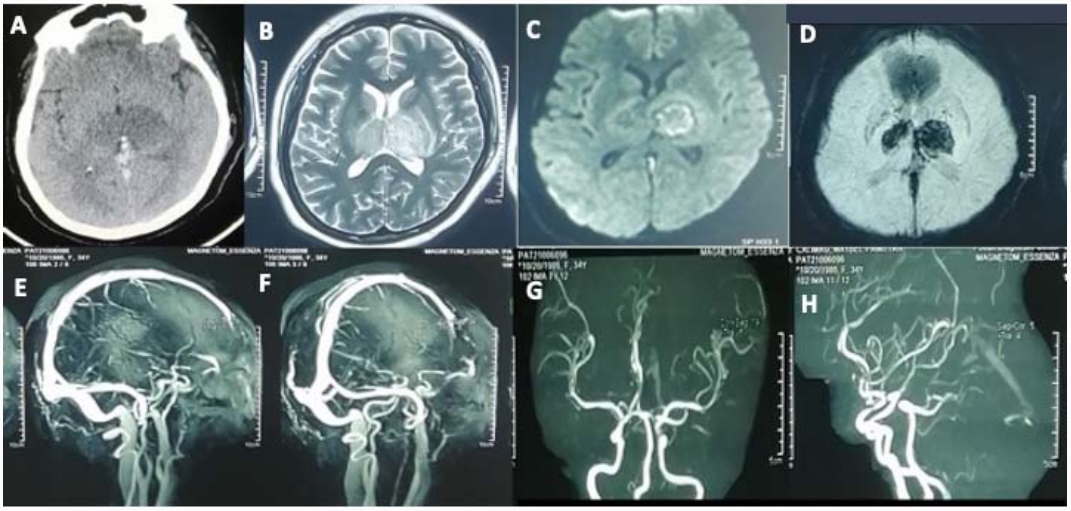

Figure 1: A) Hypodensity on bilateral thalami Plain Cranial CT scan on admission; (MRI images, B-H) B) T2 hyperintensity due to edema on bilateral thalami, C) DWI hyperintensity, D) SWI void on both thalami consistent with subacute hemorrhagic conversion, E/F) Absence of flow signal and non-opacification of the internal cerebral veins, vein of Galen, and straight sinus, G/H) Multiple short segmental intimal irregularity and luminal narrowing of the internal carotid branches ACA and MCA territories.

photosensitivity, and alopecia and she had an unremarkable OB-Gyne history with no history of contraceptive use.

On admission, her blood pressure, temperature, heart rate, respiration and oxygen saturation were normal. Physical examination findings on the head, neck, heart, lungs, abdomen, and genitals were normal. On neurologic examination, she has minimal eye opening to pain, no regard, and does not follow command. She had occasional spontaneous purposeful movement of all extremities with equal and symmetrical good muscle tone without spasticity or rigidity. Complete blood count showed anemia (Hemoglobin of 97 g/L). Results of serum chemistry were normal. ESR was elevated but work up for immunologic diseases such as ANA, anti-phospholipid panel, P-ANCA and C-ANCA were normal. Workup for secondary causes of vasculitis such as Hepatitis B testing, thyroid function tests and blood cultures were unremarkable. Cardiac 2-D echo was also negative.

Plain cranial CT scan revealed hypodensities on both thalami (Figure 1A). Metastatic work-up with chest and whole abdomen CT with IV contrast were unremarkable. Ovarian neoplasm markers were all negative. Cranial MRI with MRA and MRV revealed a hyperintensity on T2 and DWI (Figure 1B, 1C) with subacute hemorrhagic conversion on both medial thalami on GRE (Figure 1D) with thrombosis of vein of Galen, internal cerebral veins, and straight sinus (Figure 1E, 1F) with multiple minimal to moderate segmental arteritis (Figure 1G, 1H). Pathergy test was positive after an erythematous papule was noted on the intradermal injection site within 48 h. The patient was diagnosed with Neuro-Behcet's disease - having met the International Criteria for Scoring and International Consensus Recommendation.

The patient was admitted at ICU. Low-molecular weight heparin started and anemia was corrected. Methylprednisolone 1 gr per day for 5 days was given and subsequently shifted to oral prednisone thereafter. On the 20th day of admission, heparin was shifted to oral anticoagulant and Azathioprine was added. Gradual improvement was noted and she was discharged on the 24th hospital day with near complete resolution on repeat MRI on follow up.

Table 1: International Criteria for Behcet Disease Scoring, Pathergy test is optional, 1 extra point may be added for positive result; Score: <3-not having BD, 3-probable BD, > definitive BD.

Sign/Symptom	Points
Ocular lesions	2
Genital Aphthosis	2
Oral aphthosis	2
Skin lesions	1
Neurological manifestations	1
Vascular Manifestations	1
Positive pathergy test	1

Discussion

Behcet's Disease was originally described in 1937 by the Turkish dermatologist as a complex, multisystem, auto-inflammatory disease primarily involving the eyes, mouth, and genitals [5]. The International criteria for scoring was used to determine if the patient has Behcet's Disease with a sensitivity and specificity of 94% and 92% [3]. Our patient had a score of 5 based on diagnostic criteria (Table 1) - due to the history genital aphthosis, neurological manifestations, vascular manifestations, and positive pathergy test; leading to a definite diagnosis of Behcet's disease.

Pathergy was positive and is highly suggestive of BD with 87% specificity [6].

For definite Neuro-Behcet, the following criteria must be met; 1. Satisfy the criteria for Behcet's Disease, 2. Neurological syndrome (with objective neurological signs) supported by relevant and characteristic abnormalities seen on either neuroimaging, 3. No better explanation for the neurological findings. The patient has satisfied all the criteria for Definite Neuro-Behcet's disease.

Our patient had no mucocutaneous lesions during admission but had history of recurrent genital ulcers. The most common neurologic involvement is cerebral venous sinus thrombosis, which may be seen in up to 12% to 20% [7]. In this case, it affects the deep cerebral vein

and draining sinus, thus bilateral thalami was involved. Headache, the initial complaint of our patient, is also the most common neurologic symptom reported in Neuro-Behcet's. MR venography confirmed the diagnosis but arterial involvement is rare and the presence of arteritis in cranial MRI suggests that intra-axial arteries were involved.

There are no definite guidelines for the management of Neuro-Behcet's Disease. Treatment strategies depends on the clinical experience, availability of the imaging, and immunosuppressants.

The patient was managed with methylprednisolone of 1 g/day for 5 days, followed by oral Prednisone for 1 month before tapering. Steroid therapy was combined with Azathioprine for long-term immunosuppression upon discharge. In a multicenter study on the prognosis of patients with chronic progressive Neuro-Behcet, mortality rate and severe disability were 35.2% and 65.4% respectively [8]. Involvement of ocular, vascular, neurological, and gastrointestinal are associated with a poor prognosis [8].

Our patient was monitored closely since vascular and neurological involvement portend poor prognosis. To our knowledge, this is possibly the first reported case of Neuro-Behcet's disease in the Philippines which presented as stroke secondary to cerebral deep vein thrombosis and concomitant arterial stenosis.

Conclusion

This is a case of a young Filipina initially managed as a case of ischemic stroke. Further workup on imaging revealed thrombosis of deep cerebral veins thus workup for hypercoagulable state was done. Laboratory and imaging for neoplasm and other autoimmune diseases were unremarkable however marker for inflammation was elevated. Neuro-Behcet disease was considered due to history of recurrent genital ulcer, positive pathergy test, and elevated inflammatory marker. Bilateral thalamic lesions were attributed from cavernous sinus thrombosis however imaging showed a concomitant cerebral arteritis. The directed testing and review of the clinical consensus criteria confirmed the diagnosis of Behcet's disease. Thus, this is a Definite Neuro-Behcet disease with widespread involvement of both the venous and arterial system.

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