CASE REPORT

# Uveitis and Retinal Vasculitis—Harbingers of Neuro-Behcet's Disease: A Case Report

Pramod Kumar Samala[1], Sowmini Perumal[2], Sakthi Velayudham[3], Malcolm Jeyaraj[4], Kannan Vellaichamy[5], Viveka Saravanan Raju[6], Mugundhan Krishnan[7]

*Received on:* 20 July 2023; *Accepted on:* 17 September 2023; *Published on:* 21 December 2023

## Abstract

We report this case of Neuro-Behcet's disease who initially had recurrent episodes of uveitis over a period of 14 years and was under ophthalmological follow-up. Recently, she developed retinal vasculitis and multiple stroke-like episodes. She was evaluated for Behcet's disease. Pathergy test and HLAB51 were positive. She had characteristic diencephalic lesions on neuroimaging and responded well to immunotherapy.

**Key message:** Uveitis and retinal vasculitis may be harbingers of Neuro-Behcet's disease. Rare neurological manifestations such as stroke-like episodes can occur much later in the course of Behcet's disease with unique neuroimaging findings.

**Keywords:** Case report, Neuro-Behcet's disease, Retinal vasculitis, Stroke-like episodes.

*Bengal Physician Journal* (2023): 10.5005/jp-journals-10070-8022

## Case Description

We report a case of a 36-year-old lady with history of recurrent uveitis since 2007 (four episodes in the past 14 years) with the last episode being associated with retinal hemorrhages suggestive of retinal vasculitis in January 2021 (Fig. 1). She was treated by an ophthalmologist with tapering doses of oral steroids.

She presented to us in July 2021 with recurrent episodes of right-sided faciobrachial monoparesis, each episode lasting for about 2 to 3 minutes, which resolved completely and spontaneously. She had five to six such episodes per day over a period of 10 days, with the last episode being on the day of admission.

On admission, her neurological examination was unremarkable. Magnetic resonance imaging (MRI) brain revealed an acute left thalamic infarct (Fig. 2). Cardiac workup including ECG and 2DECHO was normal. Computed tomographic (CT) angiogram of the vessels in the brain and neck showed no significant abnormality. Routine biochemical blood investigations including complete blood picture and renal and liver functions were normal. Considering the possibility of autoimmune/inflammatory etiology, serum ACE levels and serum calcium were done which were normal. Computed tomography chest was normal. Tests for antinuclear antibody along with extractable nuclear antigen (ENA) profile were negative. Antiphospholipid antibody workup was negative. Mantoux and TB QuantiFERON gold were negative. Rheumatoid factor (RA factor) was negative. Test for rapid plasmin regain was nonreactive. Cerebrospinal fluid analysis was normal. Considering the possibility of Neuro-Behcet's disease, pathergy test was done, which was positive. Genetic analysis revealed HLAB51 positivity.

With the classical clinical picture of recurrent uveitis/retinal vasculitis and new onset neurological manifestations, pathergy positivity and HLAB51 positivity, the diagnosis of Neuro-Behcet's disease was made and the patient was treated with intravenous methylprednisolone 1 gm once daily for 5 days. After 2 days of commencement of the treatment, the patient was asymptomatic with no further stroke-like episodes. She was discharged with tapering dose of oral prednisolone, 75 mg of aspirin, and 100 mg of azathioprine. She is under regular follow-up without any further episodes.

[1–7]Department of Neurology, Stanley Medical College, Chennai, Tamil Nadu, India

**Corresponding Author:** Sowmini Perumal, Department of Neurology, Stanley Medical College, Chennai, Tamil Nadu, India, Phone: +91 9444256497, e-mail: drprsowmini@yahoo.co.in

**How to cite this article:** Samala PK, Perumal S, Velayudham S, *et al*. Uveitis and Retinal Vasculitis—Harbingers of Neuro-Behcet's Disease: A Case Report. Bengal Physician Journal 2023;10(3):102–104.

**Source of support:** Nil

**Conflict of interest:** None

**Patient consent statement:** The author(s) have obtained written informed consent from the patient for publication of the case report details and related images.

## Discussion

Behcet's disease is a rare immune-mediated small vessel vasculitis. The characteristic features include oral ulcers, genital ulcers, and ocular involvement. Knapp first reported the neurological involvement in Behcet's disease in 1941.[1] Cavara and D'Ermo coined the term Neuro-Behcet syndrome in 1954.[2]

Behcet's disease is commonly reported in younger people, usually in their twenties. In one of the largest case series published on Neuro-Behcet's disease with 200 cases by Akman-Demir et al.,[3] the average age of onset of Behcet's disease was 25.8 ± 7.8 years (median = 25 years), and the age at onset of neurological symptoms was 31.5 ± 8.9 years (median = 31), with a mean interval of 5.6 ± 5.5 years between these two ages.

Behcet's disease causes two different categories of neurological involvement, parenchymal and nonparenchymal involvement.[4] Meningoencephalitic presentations, hemispherical manifestations,

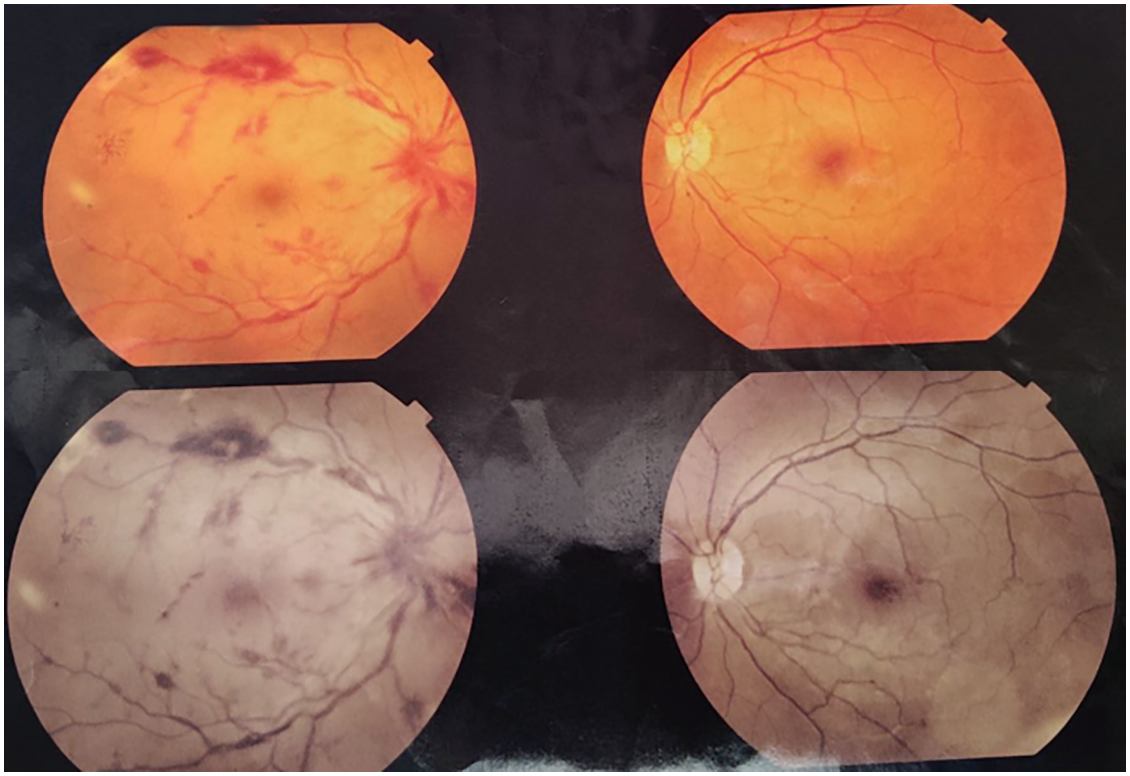

**Fig. 1:** Fundus picture showing multiple retinal hemorrhages, hyperemic disc, and roth spots in left eye

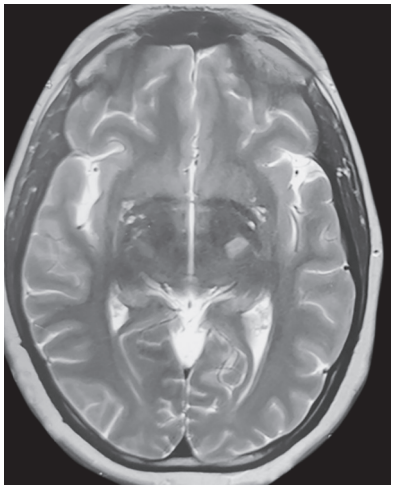

**Fig. 2:** Magnetic resonance imaging brain T2 image showing hyperintense lesion in left thalamus

brainstem manifestations, and spinal cord lesions are all the different manifestations of parenchymal involvement.

Although our patient did not have the history of oral or genital ulcers, which is the prerequisite for the diagnosis of Behcet's disease according to the International Study Group for Behcet Disease, neurologic involvement can occur in the absence of orogenital ulcers.

Lueck et al.[5] reported a case of persistent inflammatory central nervous system illness with uveitis, but without any oral or vaginal ulcers or arthritis. The neuropathological study of that case found histological alterations consistent with a Neuro-Behcet's disease diagnosis. As a result, Neuro-Behcet's disease without mucocutaneous-ocular symptoms may impact more patients than previously documented, as it may be underdiagnosed. The authors suggested the term "Behcet's MINUS" syndrome (multifocal intermittent neurological and uveitic syndrome) to describe such a condition.

Sigal[6] suggested that neurological symptoms can precede ulceration in Behcet's disease. The International Study Group for Behcet's disease also acknowledges that the disease can exist without oral ulceration.[7]

Sung Pil Joo et al.[8] reported a case of a young female diagnosed with Behcet's disease presented with repeated transient ischemic attacks of left hemiparesis who was later found to have moyamoya disease. However, our patient's CT angiogram was normal.

The mainstay of treatment for parenchymal Neuro-Behcet's disease is glucocorticoids (high-dose pulse intravenous and/or oral) and azathioprine. Azathioprine is commonly used as a first-line disease-modifying treatment for the serious manifestations of Behcet's disease, particularly Neuro-Behcet's disease, due to its relatively predictable and low side effect profile. Our patient responded well to the steroid therapy and is currently on azathioprine without any further relapse.

The unique and distinct features of our case are the long time gap between ocular manifestations and neurological symptoms, the recurrent stroke-like episodes and the absence of orogenital ulcers. The disease is more common among young males and brainstem manifestations are more common. In contrast, our patient is a middle-aged female with left thalamic infarct (hemispherical manifestation).

## Conclusion

We report this case to highlight the rare clinical presentation of Neuro-Behcet's disease with stroke-like episodes preceded by uveitis and retinal vasculitis without orogenital ulcers, the unique MRI findings and the marked response to immunotherapy.

## References


1. Knapp P. Beitrag zur Symptomatologie und Therapie der rezidivierenden Hypopyoniritis und der begleitenden aphthösen Schleimhauterkrankungen. Schweiz Med Wochenschr, Switzerland. 1941;71:1288–1290.
2. Cavara, V, D'Ermo F. A case of Neuro-Behcet's syndrome. Acta XVII Concili Ophtalmologici 1954:1489–1505.
3. Akman-Demir G, Serdaroglu P, Tasci B. Clinical patterns of neurological involvement in Behcet's disease: Evaluation of 200 patients. Brain 1999; 122(Pt 11):2171–2182. DOI: 10.1093/brain/122.11.2171.
4. Tascilar K, Melikoglu M, Ugurlu S. Vascular involvement in Behcet's syndrome: A retrospective analysis of associations and the time course. Rheumatology (Oxford) 2014;53(11):2018–2022. DOI: 10.1093/rheumatology/keu233.
5. Lueck CJ, Pires M, McCartney AC, et al. Ocular and neurological Behcet's disease without orogenital ulceration. J Neurol Neurosurg Psychiaty 1993;56(5):505–508. DOI: 10.1136/jnnp.56.5.505.
6. Sigal LH. The neurologic presentation of vasculitic and rheumatologic syndromes. A review. Medicine 1987;66(3):157–180. DOI: 10.1097/00005792-198705000-00001.
7. Editorial. Criteria for diagnosis of Behcet's disease. International Study Group for Behcet's Disease. Lancet 1990;335(8697):1078–1080.
8. Joo SP, Kim TS, Lee JH, et al. Moyamoya disease associated with Behcet's disease. J Clin Neurosci 2006;13(3):364–367. DOI: 10.1016/j.jocn.2005.03.040.