CASE REPORT

Challenges in Distinguishing Neuro-Behcet's Disease from Other Neurological Conditions: A Case Report from Pakistan

Rafi ullah[1*] and Arusa Lakhani[1]

[1]*Department of Nursing, Aga Khan School of Nursing and Midwifery, Karachi, Pakistan*

Abstract

Neuro-Behcet's Disease (NBD) is a rare complication of Behcet's Disease, being reported in approximately 5% of cases. This case study details a 36-year-old man with Behcet's Disease who has developed acute neurological symptoms, including right arm weakness, vision disturbance, facial numbness, and intermittent gait disturbance. The patient had a history of recurrent oral ulcers, genital ulcers and episodes of uveitis. Diagnostic imaging revealed characteristic findings such as perilesional vasogenic edema and nodular enhancement. Cerebrospinal fluid analysis showed elevated protein levels and neutrophils, consistent with NBD. However, the scarcity of screening and specific diagnostic tests makes it difficult to diagnose NBD in the early stage. Hence, it is realized that the complexities in NBD demand a multidisciplinary approach where advanced practice nurses, rheumatologists, neurologists, and infectious disease experts can work together to achieve early diagnosis and effective management for optimizing prognosis and enhancing the quality of life. This case also underscores the need for ongoing multispecialty care in the long-term management of NBD.

Keywords: *Neuro-Behcet's disease, Behcet's disease, diagnostic challenges, multi-disciplinary approach, differential diagnosis.*

INTRODUCTION

Neuro-Behcet's disease (NBD) is a rare neurological complication of Behcet's disease (BD), which is an autoimmune vasculitis disorder affecting blood vessels [1]. It is characterized by neurological symptoms and signs in an individual diagnosed with BD [2]. In NBD, approximately 50% of cases involve the brainstem, while diencephalon and other brain regions are affected in around 30%. Additionally, the spinal cord is impacted in roughly 10% of cases. These neurological complications can lead to movement disorders, epilepsy, psychiatric symptoms, and cognitive issues, which can result from both the brain disease itself, inflammation, and feelings of fatigue and despondency [3]. Behcet's disease typically impacts individuals aged 20 to 40 [4]. The condition affects both genders, with male predominance in Arab populations and female predominance in Korea, China, the US, and northern Europe, and has a more severe course in males and younger populations [5].

CASE PRESENTATION

The case begins with a 36-year-old male patient presenting with a sudden onset of neurological symptoms. Two days before admission, he was in his usual state of health when he experienced the abrupt onset of right arm weakness and facial numbness. These symptoms were intermittent and were accompanied by gait disturbance and fever. Additionally, he reported red spots in his eyes. On neurological examination, the patient had spontaneous eye-opening; pupils were bilateral, equal, and reacted to light, and measured 2 mm; tone and bulk examined normal in all four limbs; power was 2/5 in the right upper and lower limbs and 3/5 in the left upper and lower limbs; the plantar was bilaterally mute.

The patient had a history of recurrent oral ulcers, genital ulcers, and skin lesions that raised suspicion of Behcet's disease. These mucocutaneous symptoms are key features of Behcet's disease, and their presence, along with the patient's history of uveitis, further supported the diagnosis of Behcet's disease.

These recent symptoms prompted a medical evaluation to assess the potential neurological and ocular manifestations associated with Behcet's disease. CT head, MRI and lumber puncture were done. CT head without contrast revealed perilesional vasogenic edema, and MRI showed abnormal foci, diffused restriction, and nodular enhancement on the left frontoparietal lobe and basal ganglia. In contrast, the CSF showed a raised protein level. The diagnosis of Neuro-Behcet's Disease was established by a history of mucocutaneous symptoms and corroborated by characteristic neurological findings on neuroimaging. CSF analysis confirmed this diagnosis, showing that elevated protein and neutrophilic pleocytosis findings were consistent with NBD. Differential diagnoses like multiple sclerosis and systemic lupus erythematosus were taken, but later they were ruled out based on the clinical presentation and laboratory results. The patient was finally diagnosed with Neuro-Behcet's disease and managed according to the recommendations of rheumatology and the ID team.

Upon admission of the patient to the ward, steroid pulse therapy was started. By the fourth day of his

**Corresponding author: Rafi ullah, Aga Khan School of Nursing and Midwifery, Karachi, Pakistan, Email: rafii7062@gmail.com*
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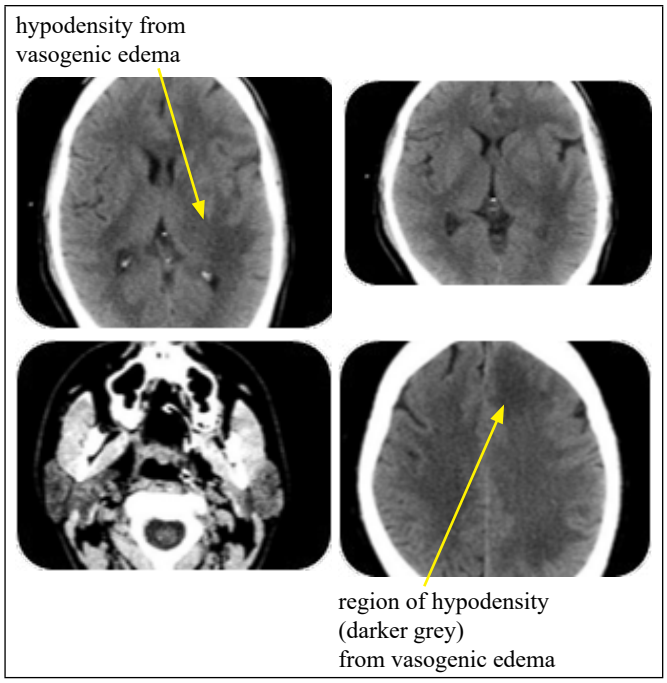

Fig. (1): CT head without contrast showing interval development of multifocal hypoattenuating areas in supratentorial brain parenchyma with perilesional vasogenic edema (highlighted with yellow arrows).

admission, the patient's symptoms worsened, and the patient became aggressive and abusive. This change in mental status prompted an evaluation by the psychiatric team; the Psychiatric team labelled it delirium and started Haloperidol. Other Medications include methylprednisolone pulse, cyclophosphamide, and broad-spectrum antibiotics. Despite these interventions, the patient continued to have multiple seizure episodes, which necessitated further evaluation and a change in the treatment plan.

Regardless of such interventions, the patient still had multiple seizure episodes. An electroencephalogram (ECG) was performed, which showed normal findings. On the ninth day of admission, the patient's condition further declined, and GCS dropped to 7/15 with recurrent seizures, mandating intubation to secure the airway and shifting to an intensive care unit.

After being in the ICU for eight days, the patient's condition showed signs of improvement and stabilization of vital signs; extubation was performed as the patient demonstrated improved neurological function and could maintain adequate ventilation independently. The patient was then transferred back to the ward, and discharge planning was initiated. Upon discharge, the patient's neurological status and vital signs were within acceptable ranges, the discharge plan included plans for ongoing patient monitoring and recovery support through nursing home care (**Figs. 1** and **2**).

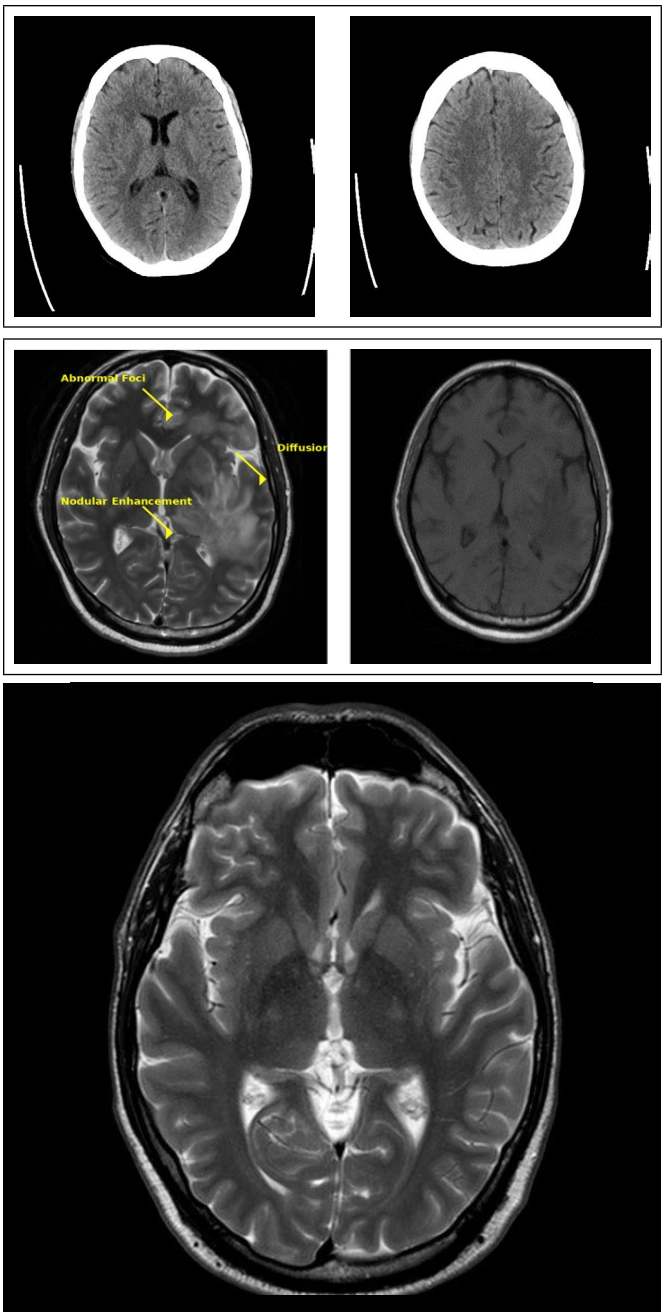

Fig. (2): MRI of the left frontoparietal lobe and basal ganglia showing abnormal foci with diffusion restriction and nodular enhancement (highlighted with yellow arrows).

The laboratory results strongly suggest Neuro-Behcet's Disease. The elevated level of CRP at 35.90 mg/L demonstrates active systemic inflammation. A significant LA ratio of 1.5 also supports an increased thrombosis susceptibility, a common complication of Behcet's Disease. Moreover, the raised white blood cell count and a high percentage of neutrophils in the CSF indicate substantial inflammation in the central nervous system. The elevated value of the CSF protein a, 150 mg/dl, is related to blood-brain barrier disturbance, representing a characteristic

feature of neurodegenerative brain disorders that reflect the severity of the neuroinflammatory process. The low serum potassium value of 3.2 mEq/L and the low lymphocyte percentage of 17.1% may relate to chronic inflammation or the consequences of immunosuppressive therapy. Such results consequently hold a strong inflammatory response involving the systemic and central nervous systems, thereby requiring urgent and aggressive intervention to minimize further neurologic insult and sequelae (**Table 1**).

DISCUSSION

NBD is one of the most serious and challenging manifestations of Behcet's disease, both to correct diagnosis and appropriate treatment. The absence of specific diagnostic tests for NBD makes it difficult to distinguish from other neurological disorders, necessitating careful evaluation and differentiation by clinicians. The diverse and non-specific symptoms of NBD, such as recurrent oral and genital ulcers, meningoencephalitis, and peripheral neuropathy, complicate the diagnostic process further, as these symptoms often overlap with other conditions like multiple sclerosis, stroke, or even infections [2].

Table 1: Laboratory investigation.

Lab Test	Unit	Reference Range	Result
Serum Mg	mg/dl	1.6-2.6 mg/dL	1.6
Serum Na	mEq/L	136-145 mEq/L	137
Serum K	mEq/L	3.5-5.1 mEq/L	3.2 ↓
Serum Chloride	mEq/L	98-107 mEq/L	99
Hb	g/dl	12.3-16.6 g/dl	13.1
HCT	%	38.4-50.7%	39.5
RBC	million/µL	4.25-6.02 million/µL	4.46
WBC	thousand/µL	4.8-11.3 thousand/µL	10.6
Lymphocytes	%	17.5-45%	17.1 ↓
Platelets	thousand/µL	154-433 thousand/µL	221
Creatinine	mg/dl	0.9-1.3 mg/dl	1.0
LA Screen	sec	31-44 sec	56.4 ↑
LA Confirmatory	sec	30-38 sec	39.0 ↑
LA Ratio	-	0.8-1.1	1.5 ↑
CRP	mg/L	0-10 mg/L	35.90 ↑
CSF protein	Mg/dl	15-45 mg/dL	150 ↑
CSF Chloride	mEq/L	122-132 mEq/L	119 ↓
CSF WBC	$\times 10^3/\mu L$	0-0.005 $\times 10^3$/µL	0.041 ↑
CSF Neutrophils	%	0-24%	60% ↑

The complexity of diagnosing NBD lies in its heterogeneous presentation and the lack of specific biomarkers. Practitioners must rely heavily on clinical insight, detailed patient history, and the exclusion of alternative causes. Collaborative efforts involving specialists such as rheumatologists and neurologists are crucial in navigating the diagnostic landscape of Behcet's disease [2]. The ongoing research into potential biomarkers and advanced diagnostic tools, including MRI with contrast, cerebrospinal fluid analysis, and genetic markers, is vital for improving the precision and efficiency of NBD diagnosis [6].

The case presented underscores the diagnostic complexity of NBD. The symptoms presented by the patient, including right arm weakness, facial numbness, and fever, might also be characteristic of different neurological conditions, thereby complicating the differential diagnosis. Imaging findings, such as perilesional vasogenic edema and abnormal foci on MRI, although suggestive of NBD, are not definitive and require careful interpretation alongside clinical features [7].

Systemic Treatment and Therapeutic Challenges were also present, as systemic anti-inflammatory and immune-modulating medications are the cornerstone of NBD treatment. The primary treatment involves glucocorticoids and immunosuppressants, with agents like low-dose methotrexate (MTX) and infliximab showing efficacy in halting the progression of neuropsychological symptoms. However, the response to these treatments can be unpredictable, as evidenced by the patient's worsening symptoms despite initial steroid therapy, which necessitated psychiatric intervention and intensive care management [6].

While effective in some cases, corticosteroids, cyclophosphamide, and azathioprine are not universally successful, further complicating the treatment landscape [6].

The steady improvement of the patient over time from a critically deteriorated condition illustrates the variable nature of NBD, highlighting the uncertainty in prognosis and the challenges in long-term management. This variability also complicates prognosis and necessitates ongoing multispecialty care. The chronic nature of the disease requires not only acute management but also long-term follow-up and support for potential relapses and persistent neurological deficits. A major challenge

in the long-term management of NBD is the lack of reliable diagnostic markers or techniques to accurately assess its severity in Behcet's disease [7].

The development of neuropsychiatric symptoms, including delirium, adds another layer of complexity to the management of NBD. These symptoms represent either the disease itself or the side effects of treatment, again pointing out that a comprehensive multidisciplinary approach is required for psychiatric care to be incorporated into traditional neurological and rheumatological intervention [8].

The complexity of NBD calls for an organized approach to its coordinated management. Rheumatologists, neurologists, psychiatrists, and other specialists must come together to deal with the variations in symptoms and complications that might develop. Hence, future research should aim to define specific diagnostic markers and lead to the formulation of standardized treatment protocols so that NBD can be treated with greater precision and effectiveness. Advanced neuroimaging, genetic markers, and large autoimmune panels will provide a more exact diagnostic process and lead to more effective management strategies [3, 6].

CONCLUSION

Neuro-Behcet's disease"is a rare and severe manifestation of Behcet's disease, marked by challenging neurological symptoms. The absence of specific diagnostic tests and the variable clinical presentation make NBD difficult to identify, often leading to a delayed diagnosis. A crucial aspect lies in the necessity for a multidisciplinary approach involving specialists from rheumatology, neurology, infectious diseases, and advanced practice nurses. This collaborative effort is vital for early detection, prompt intervention, and effective management of complications, emphasizing the intricate nature of NBD.

CONSENT FOR PUBLICATION

Approval for the publication of medical information and clinical photographs was obtained from the patient attendant.

CONFLICT OF INTEREST

I affirm that the authors have no financial or personal connections with individuals that might introduce bias into the research presented in this publication. They also have no conflict of interest.

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AUTHORS' CONTRIBUTION

Rafiullah is the main author of this case report and is responsible for identifying the case, gathering relevant clinical data, and drafting the manuscript.

Arusa Lakhani oversaw the planning and execution of the case report. Her contributions included directing its overall direction, offering mentorship, and critically reviewing the manuscript.

Both authors have reviewed and approved the final version of the manuscript and accept responsibility for its integrity and accuracy.

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