Unravelling the Mystery: A 19-Year-Old Male with Recurrent Strokes

Dr. Aishwarya N Udup[1], Dr. Arathi Darshan[2]
[1,2] *Jawaharlal Nehru Medical College Belgaum*

***Abstract*—Background Behçet 's disease (BD) is a chronic, multisystem autoimmune vasculitis that can affect various organs, including the central nervous system (CNS). Neuro-Behçet 's disease (NBD) is a rare but severe manifestation, often presenting diagnostic challenges due to its overlap with other neurological disorders.**

Case Presentation:

We report the case of a 19-year-old male who presented with sudden-onset right-sided weakness, slurred speech, and facial asymmetry. His history included recurrent oral ulcers but no prior neurological events. Neurological examination revealed right-sided hemiparesis with upper motor neuron facial palsy and gaze palsy. Imaging studies, including MRI, identified T2 and FLAIR hyperintensities in the left basal ganglia, brainstem, and periventricular white matter. A positive pathergy test and cerebrospinal fluid (CSF) analysis supported the diagnosis of NBD. Management & Outcome: The patient was treated with high-dose intravenous methylprednisolone followed by oral steroids in a tapering regimen. In light of thrombotic risk, anticoagulation therapy with apixaban was initiated. The patient showed significant neurological improvement and was discharged with no residual deficits at the one-month follow-up.

Conclusion:

This case underscores the importance of considering NBD in young patients with recurrent strokes and systemic BD features. Early diagnosis using neuroimaging and laboratory markers, coupled with aggressive immunosuppressive therapy, can improve clinical outcomes and reduce long-term complications. Further research is required to refine treatment protocols for NBD to prevent relapses and disability.

***Index Terms*—Neuro-Behçet 's disease, Behçet 's disease, recurrent stroke, vasculitis, immunosuppression**

I. INTRODUCTION

Behcet's disease (BD) is an autoimmune disease characterized by recurrent oral and genital ulcers and uveitis. The exact pathogenesis is still unclear, but the main histopathological feature is widespread vasculitis of arteries or venules of any size. Many organs may be affected; however, neurological involvement in Behcet's (neuro-Behcet's disease) is rare and remains a difficult diagnosis to establish as certain infections may also produce a similar picture. Here, we present a patient with neuro-BD with acute neurological involvement.

A. Case Report

A 19-year-old male presented with:

- Weakness of right upper and lower limb since 2 days.
- Deviation of the angle of mouth with slurring of speech since 2 days.
- The patient was apparently alright 2 days back, after which he developed sudden-onset, non-progressive weakness of the right upper limb upon waking up.
- The patient also complained of slurring of speech, deviation of the angle of the mouth towards the right side, and difficulty swallowing.
- The patient was evaluated at an outside hospital where a CT brain was advised.

B. Past Medical History

- History of repeated oral ulcers.

- Not a known case of diabetes (T2DM), hypertension, ischemic heart disease (IHD), tuberculosis, prior stroke.
- No history of trauma, surgery, immobilization, or central catheter insertions.

C. Personal History
- Mixed diet
- Normal appetite
- Normal bowel and bladder habits
- No habits

D. Family History:-
Nothing significant

E. Examination Findings
Vital Signs:
- PR: 110 bpm
- BP: 140/80 mmHg
- SpO2: 98% under room air
- Eye redness: Present

F. Systemic Examination:
- CVS: S1 S2 +, no murmurs
- RS: Normal vesicular breath sounds
- P/A: Soft

G. Neurological Examination:

- Conscious & Oriented
- Tone: Right hypertonia
- Right-sided hemiparesis (Power: 3/5)
- Brisk deep tendon reflexes on right side
- Extensor plantar reflex on right side
- Gait ataxia: Present
- Right UMN facial palsy
- Right lateral gaze palsy

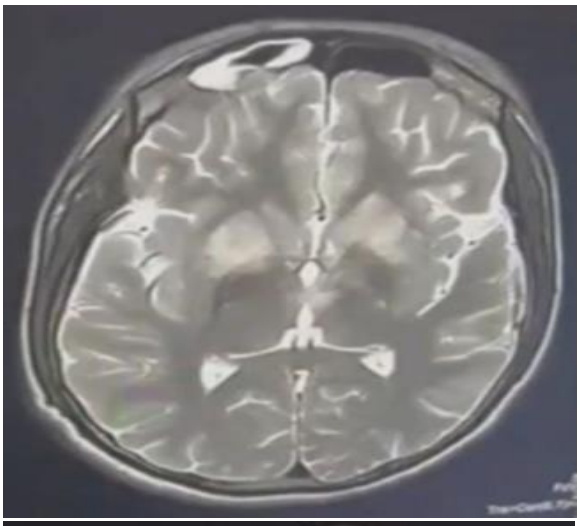
H. Investigations
- CT Scan: Acute/hyperacute infarct involving the left caudate nucleus, adjoining capsuloganglionic I. region.
- Cardiac Evaluation:
- ECG: Normal sinus rhythm
- Holter Monitoring: Normal
- MRI Brain Findings:
- T2 and FLAIR hyperintensities involving left centrum semi ovale, left periventricular frontal subcortical white matter, bilateral basal ganglia, brainstem (midbrain and pons) with no diffusion restriction.
- Pathergy Test: Positive
- CSF Analysis: Conducted

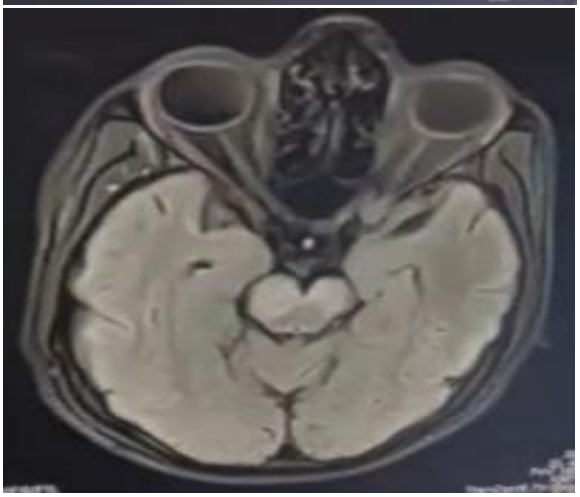
INVESTIGATIONS

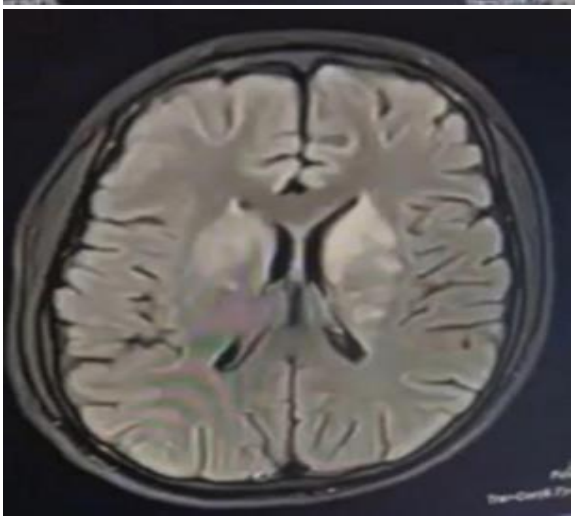
Haemoglobin	14.7	Cholesterol	151	CSF LDH	39
Red blood cell	515000	LDL	94	CSF ADA	0.8
WBC	7700	HDL	44	Indian ink	Negative
Platelet count	220000	Triglycerides	64	Ecg	Sinus tachycardia
Urea	36	CSF analysis		2D echo	Normal
Creatinine	0.95	Appearance	Clear	Usg	Normal
Potassium	4.17	Cell count	7cells/cumm	Holter	Normal
Chloride	95	Cell type	All lymphocytes	LFT	wnl
Bicarbonate	22	Gram stain	Few pus cells	Homocysteine	30
ANA EIA	negative	CSF protein	57.8		
VIT B12	354	CSF glucose	68		

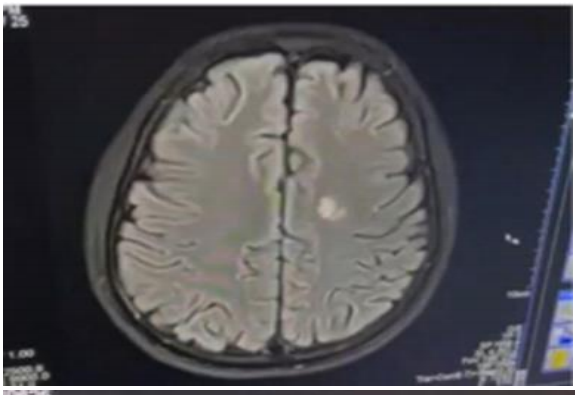
II. MRI BRAIN IMAGING

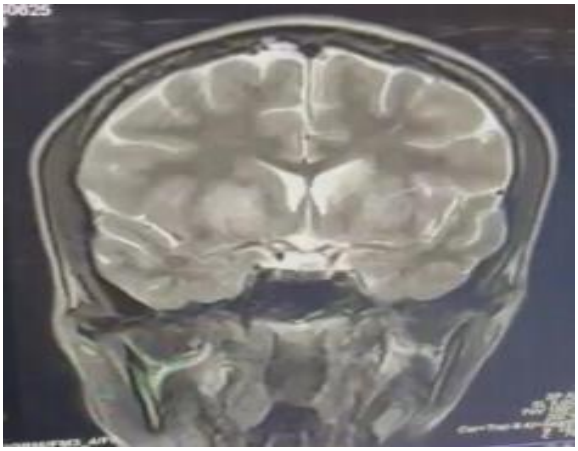
The following images represent MRI findings suggestive of Neuro-Behcet's disease.











A. Treatment & Discharge Summary

- The patient was started on:
- IV Methylprednisolone 500 mg once daily for 3 days.
- In view of thromosis patent was started on Tab Apixaban 2.5 mg (1-0-0).
- The patient's weakness improved significantly, and he had no residual weakness upon discharge.
- He was discharged on Apixaban and steroids in a tapering dose.
- Patient weakness improved drastically and doesnot have residual weakness on follow up after 1month

III. DISCUSSION

Neuro-Behçet ’s disease (NBD) is a rare but severe manifestation of Behçet’s disease (BD), characterized by neurological involvement due to systemic vasculitis. The presented case of a 19-year-old male with recurrent strokes, oral ulcers, and a positive pathergy test aligns with the

clinical spectrum of NBD. This discussion explores the pathogenesis, differential diagnosis, treatment, and prognosis of NBD with relevant literature.Pathogenesis and Clinical SpectrumBD is a multisystem inflammatory disorder
affecting small, medium, and large vessels. Neurological involvement is seen in approximately5
10% of BD cases, manifesting in two primary forms: parenchymal NBD and non-parenchymal
NBD (vascular involvement) (1). The current case demonstrates parenchymal NBD, evident from MRI findings of hyperintensities in the brainstem, basal ganglia, and periventricular white matter. Inflammatory vasculitis and perivascular infiltration are the key histopathological features leading
to ischemic or hemorrhagic lesions in the CNS .

IV. DIFFERENTIAL DIAGNOSIS

The neurological manifestations of NBD can mimic other cerebrovascular disorders, making diagnosis challenging. Differential diagnoses include multiple sclerosis, neuromyelitis optica, CNS infections, and primary vasculitis. Unlike multiple sclerosis, NBD lacks oligoclonal bands in cerebrospinal fluid (CSF) and shows a different MRI lesion distribution (3). Infections such as tuberculosis and viral encephalitis should also be considered, but the absence of systemic infectious markers and a positive pathergy test
favor NBD (4).Neuroimaging and DiagnosisMRI remains the gold standard for diagnosing NBD. T2 and
FLAIR hyperintensities in the basal ganglia, brainstem, and periventricular white matter are characteristic findings, as seen in this patient (5). CSF analysis typically reveals mild pleocytosis and elevated protein, further supporting the diagnosis. The positive pathergy test, although not exclusive to BD, strengthens the
clinical suspicion in this case.

V. MANAGEMENT AND TREATMENT

The cornerstone of NBD management is immunosuppression. High-dose corticosteroids, as administered
in this case, are the first-line treatment for acute exacerbations, effectively reducing inflammation and preventing further neurological damage (6). Additionally, anticoagulation with apixaban was initiated to mitigate the risk of recurrent strokes, which are common in NBD due to vasculitis-induced thrombosis.
Long-term immunosuppressants, including azathioprine or cyclophosphamide, may be required for recurrent or refractory cases (7).

VI. PROGNOSIS AND FOLLOW-UP

Early recognition and treatment of NBD are crucial for preventing disability.
While this patient showed remarkable improvement with steroid therapy, close monitoring is necessary to detect relapses. Long-term therapy with immunosuppressants may be required in severe cases to
prevent further neurological deterioration. The overall prognosis depends on the extent of CNS involvement and response to immunotherapy (8).

VII. CONCLUSION

This case highlights the importance of considering NBD in young patients presenting with
recurrent strokes and systemic BD features. Early diagnosis through neuroimaging and
laboratory tests, followed by aggressive immunosuppressive therapy, can significantly improve
clinical outcomes. Further research is required to establish optimal treatment protocols and long-term management strategies for NBD.

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