International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2024: 7.101

Unraveling Neuro-Behçet's Disease: Insights from a Case of Acute Brainstem Syndrome and the Clinical Value of the Cascade Sign

Dr. Priyanka Jangam[1], Dr. N. V. Sundarachary[2], Dr. G. Bindu Narmada[3], Dr. M. Bhargavi Devi[4], Dr. Kamalakar P[5]

[1]MD General Medicine, DM Neurology Resident, Guntur Medical College, Guntur, AP (Corresponding Author)

[2]MD DM, Professor of Neurology, GMC, Guntur

[3]MD DM, Assistant Professor of Neurology, GMC, Guntur

[4]MD DM, Assistant Professor of Neurology, GMC, Guntur

[5]MD General Medicine, Assistant Professor, JR Medical College, Avanampattu, TN.

Abstract: ***Introduction**: Behcet's disease (BD) is characterized by recurrent oral and genital ulcers, and uveitis. It is also known to affect various organs; however, central nervous system involvement is rare. Association of HLA-B5/B*51 has been recognized as the strongest genetic susceptibility factor for BD. NBD manifestations can be categorized into central nervous system (CNS) and peripheral nervous system (PNS) presentations. CNS manifestations fall into parenchymal and non-parenchymal subtypes. The parenchymal type is more prevalent and presents with brainstem, hemispheric, spinal, and meningoencephalitic syndromes. **Case Report**: A 35-year-old man presented with a 1-week history of progressive brainstem symptoms including slurred speech, dysphagia, diplopia, and facial weakness. This was followed 2 days later by acute-onset, symmetrical, distal-predominant quadriparesis. He had past history of quadriparesis 5 years prior, with residual foot weakness. On examination patient is Conscious and oriented. Tachypnea and tachycardia were present. Cranial nerve examination revealed palsies of CN III, IV, VI, VII, IX, and X. Motor examination showed symmetrical UMN-type quadriparesis (power 2/5 in all limbs). **Materials and Methods**: CBC, RFT, LFT, and CSF analysis were normal. ESR and CRP were elevated. Serum NMO, MOG, ANA, and CSF OCB were negative. MRI Brain showed T2/FLAIR hyperintensities in the brainstem, thalami, and basal ganglia with a "cascade sign." MRA suggested vasculitis. HLA-B*51 was positive. Final Diagnosis: Neuro-Behçet's Disease; **Discussion**: Behçet's Disease (BD) is a systemic vasculitis characterized by oral/genital ulcers and uveitis. Neurological involvement (Neuro-Behçet's) occurs in about 9% of cases and can sometimes be the presenting feature. Parenchymal brainstem syndrome is the most common neurological presentation. The "cascade sign" on MRI (lesions extending from thalamus to midbrain) is a typical radiological finding. HLA-B*51 is the strongest genetic susceptibility factor. **Conclusion**: Neuro-Behçet's should be considered in cases of acute brainstem involvement with upward extension into thalami, especially with a relapsing-remitting course and systemic features. Aggressive immunosuppression with steroids and IVIg can be life-saving and improve outcomes.*

Keywords: Behcet disease, Neuro-Behçet's, CNS- Central nervous system, PNS- Peripheral nervous system, HLA-B*51, cascade sign, vasculitis MCA- middle cerebral artery, NMO- Neuromyelitis optica, MOG- myelin oligodendrocyte antibody.

1. Introduction

Behcet's disease (BD) is characterized by recurrent oral and genital ulcers, and uveitis. It is also known to affect various organs; however, central nervous system involvement is rare. Association of HLA-B5/B*51 has been recognized as the strongest genetic susceptibility factor for BD. NBD manifestations can be categorized into central nervous system (CNS) and peripheral nervous system (PNS) presentations. CNS manifestations fall into parenchymal and non-parenchymal subtypes. The parenchymal type is more prevalent and presents with brainstem, hemispheric, spinal, and meningoencephalitic syndromes.

2. Case Report

35-year man presented with progressive neurological deficits in form of slurring of speech, regurgitation of food and choking, diplopia, incomplete closure of eyes, and drooling of saliva from a week duration. 2 days later, he developed sudden bilateral upper and lower limb weakness involving both proximal and distal muscles. No history of tingling or numbness / swaying / seizures or altered sensorium/ fever, rash, diarrhea, mucocutaneous ulcers/ urgency or incontinence. He also complained of low back pain prior to onset of these symptoms. He suffered Quadriparesis with predominant distal muscle involvement five years prior resulting in thinning of limbs. Residual distal foot muscle weakness persisted.

On examination: Patient is conscious, coherent and oriented. Tachypnea and tachycardia present. Cranial nerve examination revealed palsies of CN III, IV, VI, VII, IX, and X. With symmetrical UMN type quadriparesis of 2/5 power in all the limbs.

3. Materials and Methods

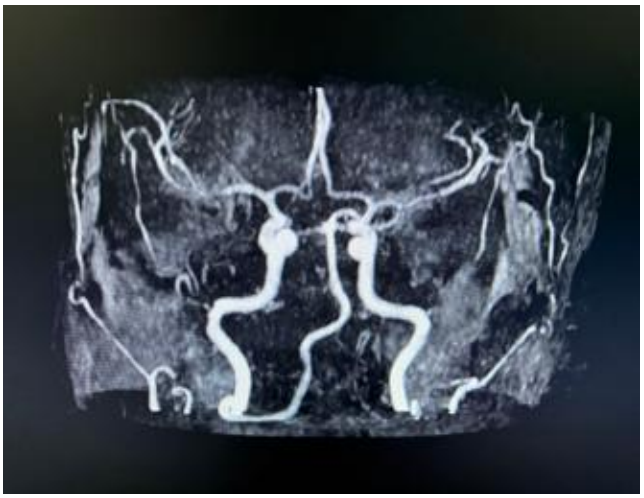
CBP, LFTS, RFTS, CUE, serum electrolytes, CSF analysis were normal. ESR and CRP were positive. Serum NMO and MOG, ANA profile, CSF OCB were negative. ECG was normal and 2D echo suggested global hypokinesia with EF 50%. CSF bacterial, viral and autoimmune panels were negative. MRI brain: T2 FLAIR hyperintensities in brain

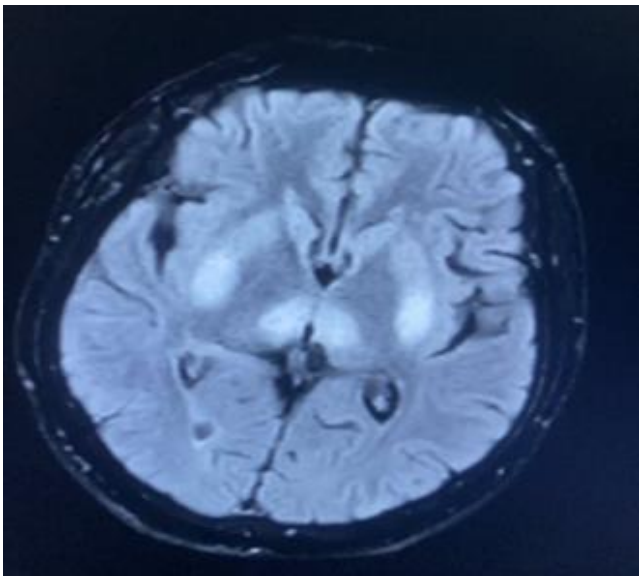
stem, bilateral thalami, basal ganglia with diffusion restriction present. Cascade sign present. MRA suggested focal areas of stenosis in bilateral MCA suggestive of vasculitis. NCS: all tested nerves were in excitable. MRI HIP: bilateral FLAIR hyperintensities in lower paraspinal muscles suggestive of myositis. HLA B 5 positive for 51 and negative for 52. Pathergy was negative as it was done after steroids and IVIG course.

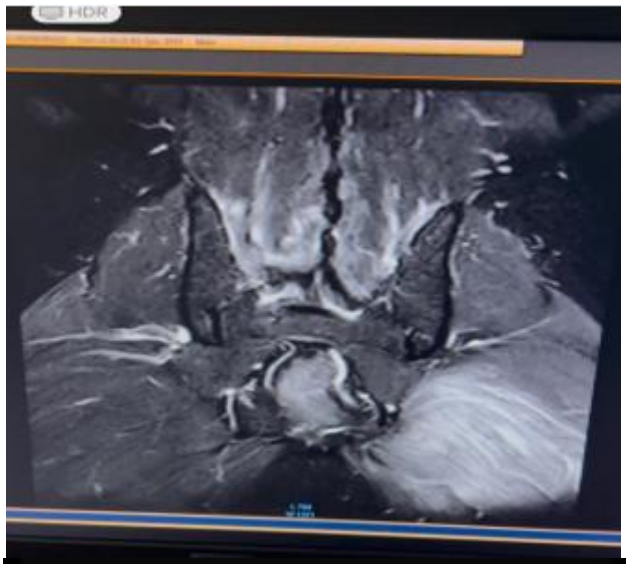
4. Results

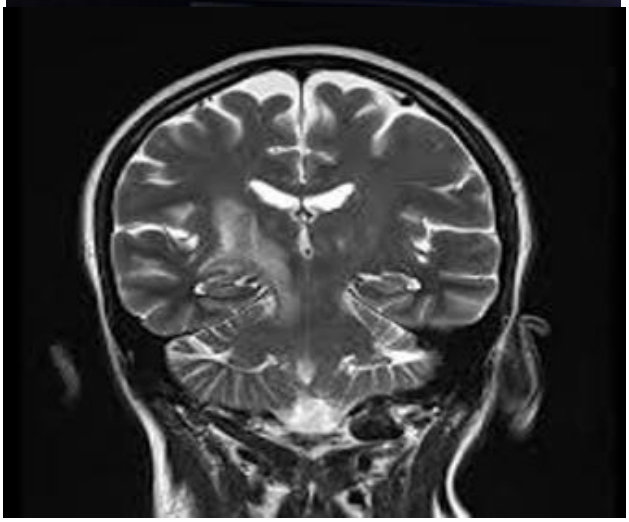
Course during hospital stay: Patient was suspected brainstem encephalitis initially and was started on broad-spectrum antibiotics, antivirals and Methylprednisolone 1gram pulse dose for 5 days, MRI - T2 FLAIR Hyperintensities seen in mid brain, pons, thalamus. Overall neurological and general condition of the patient deteriorated. Hypoxia and hypercapnia were present, and he was put on mechanical ventilator. Trial course of 125 gm IVIg was given for 5days. Other supportive treatment was also given. Post which patient had significant improvement in all his symptoms and able to walk by himself at discharge. He was discharged with tapering doses of oral prednisolone and azathioprine. Advised regular follow up.

Final Diagnosis: Neuro-Behcet's disease with parenchymal pattern of CNS, and PNS involvement.









5. Discussion

Behcet's disease (BD) is characterized by recurrent oral and genital ulcers, and uveitis. It is also known to affect various organs; however, central nervous system involvement is rare. Association of HLA-B5/B*51 has been recognized as the strongest genetic susceptibility factor for BD. The frequency of NBD amongst BD patients is approximately 9% (ranging 3–30%). In general, males may have more severe organ involvement, whereas skin lesions such as erythema nodosum are more common in females. While neurological involvement usually follows systemic Behçet's disease symptoms, in rare cases (around 6-10%), it can occur before or without prominent systemic features. [1]

NBD manifestations can be categorized into central nervous system (CNS) and peripheral nervous system (PNS) presentations. CNS manifestations fall into parenchymal and non-parenchymal subtypes. The parenchymal type is more prevalent and presents with brainstem, hemispheric, spinal, and meningoencephalitic syndromes. Non-parenchymal subtype includes cerebral venous sinus thrombosis (CVST)and arterial involvement; however, concomitant parenchymal and nonparenchymal involvement is rare. Arterial involvement includes stenosis, aneurysm formation, or dissection of the cervicocephalic arteries. Brainstem

presentations are the most common syndrome. [2] PNS manifestations, rarely seen in NBD, include neuropathies, myopathies, and neuromuscular junction disorders. [3,4]

The typical acute NBD lesions in brain MRI are mesodiencephalic lesions. The pattern of extension from thalamus to midbrain provides a cascade sign. Two spinal MRI patterns can be suggestive for NBD: "Bagel sign" and "motor neuron" sign. [1]

6. Conclusion

Neuro-Behçet's should be considered in cases of acute brainstem involvement with upward extension into thalami, especially with a relapsing-remitting course and systemic features. Classical MRI findings are Cascade sign and Bagel sign. HLA-B5/B*51 is strongest predisposing factor. Aggressive immunosuppression with steroids and IVIg can be life-saving and improve outcomes.

Ethical Considerations

Written informed consent of the patient has been taken during his hospital stay to use this clinical data related to him and to use the photographs relevant to this case report publication.

References

[1] Cleaver J, James R, Rice CM. Rhomboencephalitis. Practical Neurology. 2021 Apr 1;21(2):108-18.
[2] Jubelt B, Mihai C, Li TM, Veerapaneni P. Rhombencephalitis/brainstem encephalitis. Current neurology and neuroscience reports. 2011 Dec;11(6):543-52.
[3] Zhan H, Cheng L, Li Y. Neuro-Behçet's disease: an update of clinical diagnosis, biomarkers, and immunopathogenesis. Clinical and Experimental Immunology. 2025;219(1): uxae123.
[4] Saip S, Akman-Demir G, Siva A. Neuro-Behçet syndrome. Handbook of clinical neurology. 2014 Jan 1; 121: 1703-23.