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Neuro-Behcet's Disease: A Case Report of a 19-Year-Old Male with Recurrent Aphthous Ulcers, Headache, and Neurological Deficit

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Abstract: Neuro-Behcet's disease (NBD) is a rare and challenging manifestation of Behcet's disease (BD) that involves the central nervous system (CNS), affecting less than 10% of BD patients. The clinical spectrum includes neurological deficits such as motor dysfunction, memory impairment, and personality changes, often presenting subacutely. Here, we report a case of a 19-year-old male presenting with recurrent aphthous ulcers, headache, irritability, and left-sided limb weakness. MRI findings revealed hyperintensities in the left corona radiata, bilateral basal ganglia, midbrain, and pons, characteristic of NBD. The patient's clinical and imaging findings were consistent with a diagnosis of Neuro-Behcet's disease.

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I. INTRODUCTION

Behcet's disease (BD) is a systemic vasculitis characterized by recurrent episodes of oral and genital ulcers, uveitis, and skin lesions. While neurological involvement is rare, it is recognized as Neuro-Behcet's disease (NBD), which affects the central nervous system (CNS) in approximately 5–10% of BD patients (4). NBD primarily affects the brainstem, spinal cord, basal ganglia, thalamus, and internal capsule, with symptoms such as motor deficits, memory disturbances, and behavioral changes (8). The diagnosis of NBD is typically made by correlating clinical manifestations, MRI findings, and cerebrospinal fluid (CSF) analysis.

II. CASE PRESENTATION

A 19-year-old male presented to KLES Dr. Prabhakar Kore Hospital with a history of recurrent aphthous ulcers, headaches, irritability, and left-sided lower limb weakness. The patient's clinical course suggested a neurological involvement consistent with NBD, and further diagnostic work-up was initiated.

III. MRI FINDINGS

The patient underwent brain imaging, including axial T2-weighted and FLAIR MRI sequences. The results revealed hyperintensities in the left corona radiata, bilateral basal ganglia, midbrain, and pons. These findings were consistent with NBD .Diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) sequences showed no evidence of diffusion restriction, which is typically absent in NBD (7).

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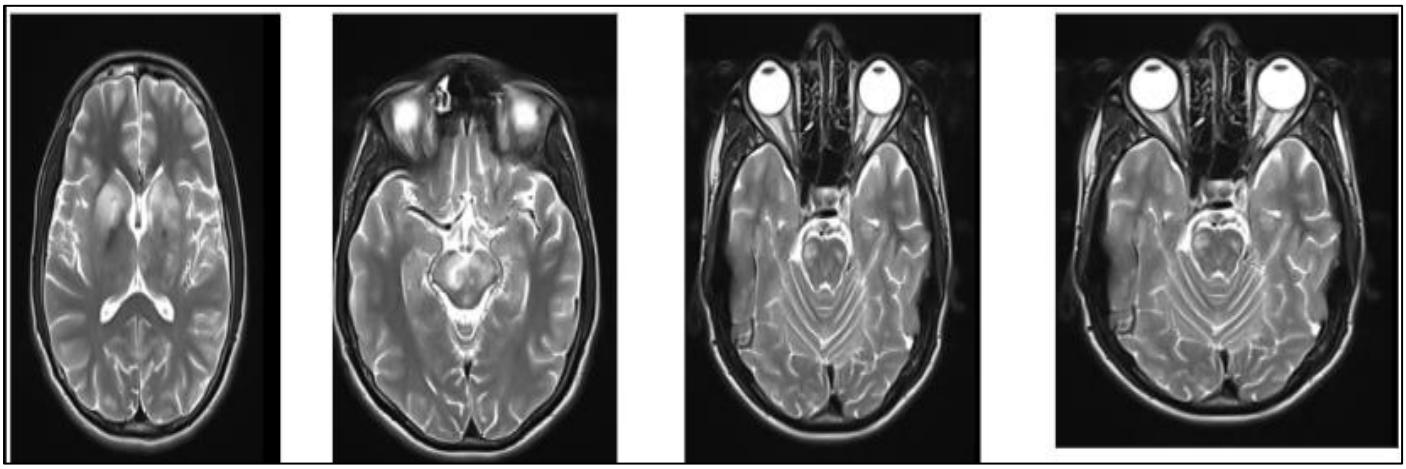

Fig 1: Axial T2w Images: Hyperintensities in the Left Corona Radiata, Bilateral Basal Ganglia, Midbrain and Pons

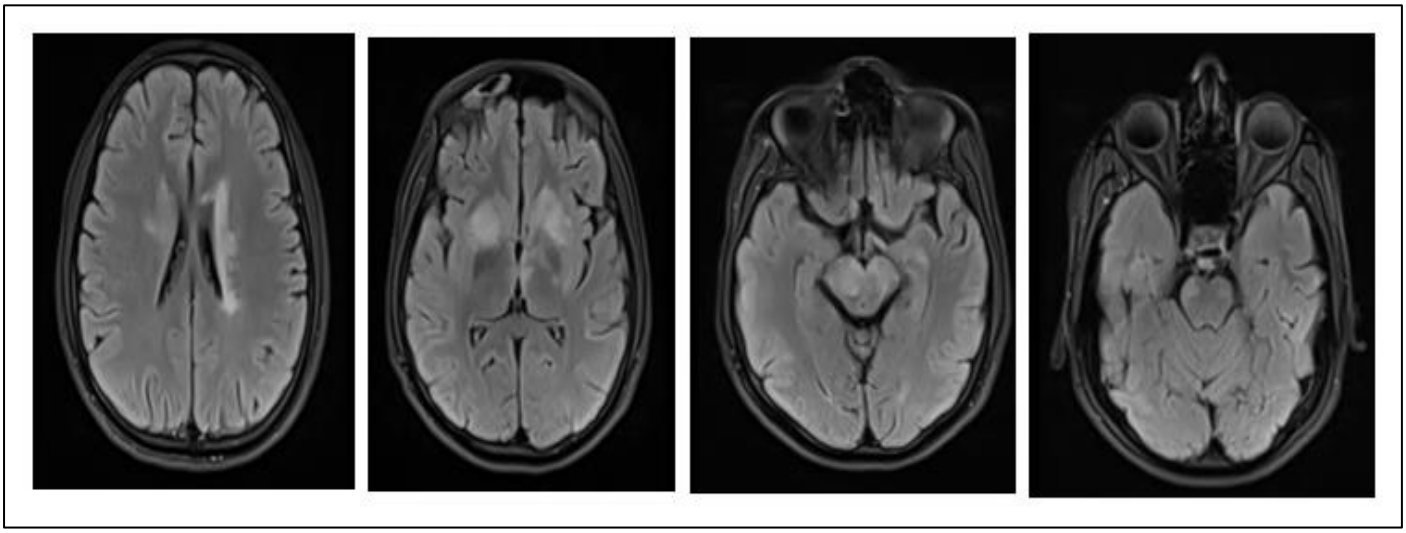

Fig 2: Axial Flair Images: Hyperintensities in the Left Corona Radiata , Bilateral Basal Gangla , Midbrain and Pons

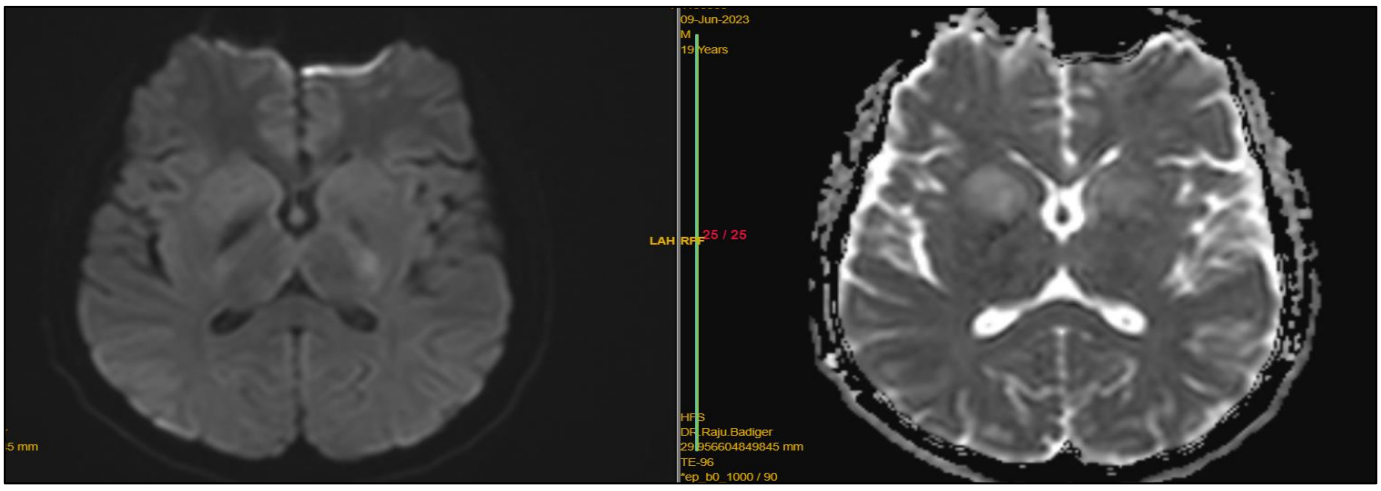

Fig 3: Axial DWI And ADC: No Evidence of Diffusion Restriction on DWI Sequence

IV. DISCUSSION

NBD is a rare manifestation of Behcet's disease that typically involves focal parenchymal lesions, often localized in areas such as the spinal cord, brainstem, basal ganglia, and thalamus. While parenchymal lesions are more commonly seen in adult patients, children with NBD tend to exhibit more non-parenchymal features (3). In this case, the MRI findings showed bilateral basal ganglia involvement, midbrain, and pons, which are common regions affected by NBD (1). These lesions present as hyperintensities on T2-weighted and FLAIR images, particularly in acute and subacute stages, and tend to enhance with contrast (2).

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Chronic NBD lesions typically show little to no contrast enhancement and may result in volume loss or atrophy of the brainstem (6). A lumbar puncture revealed an elevated protein level and an increased white blood cell count predominantly consisting of neutrophils, which is consistent with inflammatory changes observed in NBD (5).

Given the rarity of NBD, clinical diagnosis can be challenging, particularly in the early stages of the disease. The combination of clinical history, MRI findings, and CSF analysis is essential for the accurate diagnosis of NBD.

V. CONCLUSION

This case highlights the diagnostic challenges in a 19-year-old male with recurrent aphthous ulcers and neurological deficits, leading to a diagnosis of Neuro-Behcet's disease. Early recognition of NBD, supported by appropriate clinical and imaging investigations, is crucial for timely intervention and management. Although rare, NBD should be considered in patients with BD who present with neurological symptoms, particularly those involving the brainstem, basal ganglia, and other deep grey matter structures.

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