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Clinical Image

Central Nervous System Pseudomass in Neuro-Behçet's Syndrome: A Rare Radiological Manifestation and Diagnostic Challenge

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Neuro-Behçet's Syndrome (NBS) is a rare yet potentially severe neurological manifestation of Behçet's disease (BD). Although the condition frequently affects the brainstem, basal ganglia, and diencephalon, [1,2] its occurrence as a mass-like lesion (pseudotumor) is exceedingly uncommon. These tumefactive lesions can resemble neoplastic, infectious, or demyelinating conditions, which often hinders prompt diagnosis and treatment. [1-3] In this report, we discuss a 56-year-old female from Libya with a history of Behçet's disease, who presented with headaches, visual disturbances, and cognitive slowing. Magnetic Resonance Imaging (MRI) findings indicated a high T2/FLAIR signal in the bilateral basal ganglia and the left cerebellar hemisphere, extending into the middle cerebellar peduncle, accompanied by faint contrast enhancement.

Two months later, she experienced acute neurological deterioration with right-sided weakness, left eye closure, tremors, and seizures. Follow-up MRI demonstrated radiological progression with new diffuse encephalomalacia changes and the emergence of an enhancing, heterogeneous mass-like lesion in the left cerebellar hemisphere extending into the vermis (Figures 1a,b,c,d)).

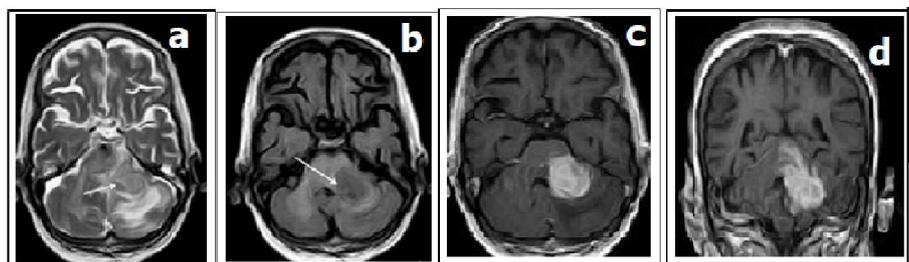


Figure 1a & 1b: Axial T2/FLAIR: High signal in left cerebellar hemisphere and middle cerebellar peduncle. **Figure 1c & 1d:** Coronal T1 contrast: Extension of pseudomass into cerebellar vermis.

After treating her with cyclophosphamide and infliximab, repeat MRI one-week post-treatment showed encephalomalacic areas stability and moderate pseudomass regression (Figure 2a,b).

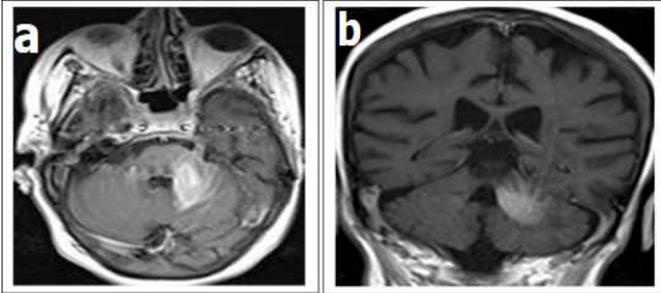


Figure 2a,b: Post-treatment axial T1 contrast: Partial regression of pseudomass lesion

Mass-like lesions in NBS are exceptionally rare. To date the number of reported cases did not exceed 28 cases. [4]

MRI findings typically include T2/FLAIR hyperintensity, peripheral or ring-like enhancement, and lack of significant mass effect (Figures 1a,b,c,d). These may resemble gliomas, primary central nervous system lymphomas, tuberculosis, fungal tumors, or tumefactive multiple sclerosis, underscoring the need for clinical-radiologic correlation.[3,4]

In this case, the lesion's atypical location (cerebellum), surrounding vasogenic edema, and therapeutic response pointed toward diagnosing pseudomass secondary to vasculitic inflammation. Prompt recognition and immunosuppressive therapy led to significant radiological improvement.

Learning points

- This case highlights the diagnostic and therapeutic relevance of recognizing pseudomass lesions in Neuro-Behçet's Syndrome. MRI remains indispensable in detecting and monitoring these lesions.
- Radiologists and neurologists should maintain a high index of suspicion for pseudomass in the appropriate clinical context to avoid invasive diagnostic approaches and ensure timely initiation of immunosuppressive therapy.

PATIENT CONSENT

A written informed consent was obtained from the patient for publication of this report.

AUTHORS' CONTRIBUTION

All authors contributed to the completion of this work. The final manuscript was read and approved by all authors.

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CONFLICT OF INTEREST

None.

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