BMC Neurology

CASE REPORT Open Access

Early atypical MRI findings in a pediatric patient with Neuro-Behçet's disease

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Abstract Behçet's disease (BD) is a multi-system recurrent inflammatory disease. Neuro-Behçet's disease (NBD), as a severe and rare manifestation of BD, is frequently misdiagnosed in its early stages. Pediatric NBD poses diagnostic challenges due to its insidious onset, highlighting the critical role of neuroimaging. This report describes a pediatric patient with atypical early clinical manifestations and magnetic resonance imaging (MRI) findings. The patient initially presented without mucocutaneous lesions despite persistently elevated inflammatory markers. MRI revealed prolonged T1/T2 signals in the posterior horns of the lateral ventricles. During years of follow-up, the patient gradually developed characteristic BD manifestations while MRI showed progressive intracranial lesions, eventually presenting typical NBD imaging features concurrently with cerebral venous thrombosis. This atypical case highlights the necessity of early multimodal MRI and close clinical monitoring of focal lesions in the posterior horns of the lateral ventricles. When infectious causes are excluded, NBD should be considered. The rarity of this case improves clinicians' ability to diagnose early NBD through MRI interpretation.

Clinical trial This case report did not involve a clinical trial.

Keywords Behçet disease, Neuro-Behçet's disease, Children, MRI

Introduction

Behçet's disease (BD) is a heterogeneous, multi-system inflammatory disease of unknown etiology. with its pathogenesis remaining poorly elucidated. Neuro-Behçet disease (NBD), one of the most severe manifestations of BD, is significantly rarer in pediatric populations and lacks definitive biochemical markers for early detection [1]. Consequently, neuroimaging is crucial for the timely identification of NBD. This paper presents a case study of a 13-year-old patient with Behçet's disease (BD) who initially presented with atypical symptoms and imaging findings. The disease progression was closely tracked over five years of follow-up."

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Case presentation

A 13-year-old male was admitted to the hospital with with a one-week history of neck pain accompanied by bilateral ocular discomfort, photophobia, and fever, without relevant family history. Physical examination indicated pharyngeal congestion. Laboratory tests indicated elevated leukocyte and neutrophil counts, as well as markedly elevated levels of high-sensitivity C-reactive protein (CRP) and calcitonin, while hepatic and cardiac profiles remained within normal limits. Empirical antibiotic therapy was initiated for suspected bacterial infection. On hospital day 3, multiple oral mucosal ulcerations emerged. Subsequently, brain MRI demonstrated patchy prolonged T1/T2 signal abnormalities in the left posterior horn of the lateral ventricle (Fig. 1), Clinicians attributed these changes to the underlying infection. By day 11, the patient experienced dizziness and headaches, and D-dimer levels were elevated at 0.58 mg/L. After 15 days of treatment, the patient's leukocyte and neutrophil

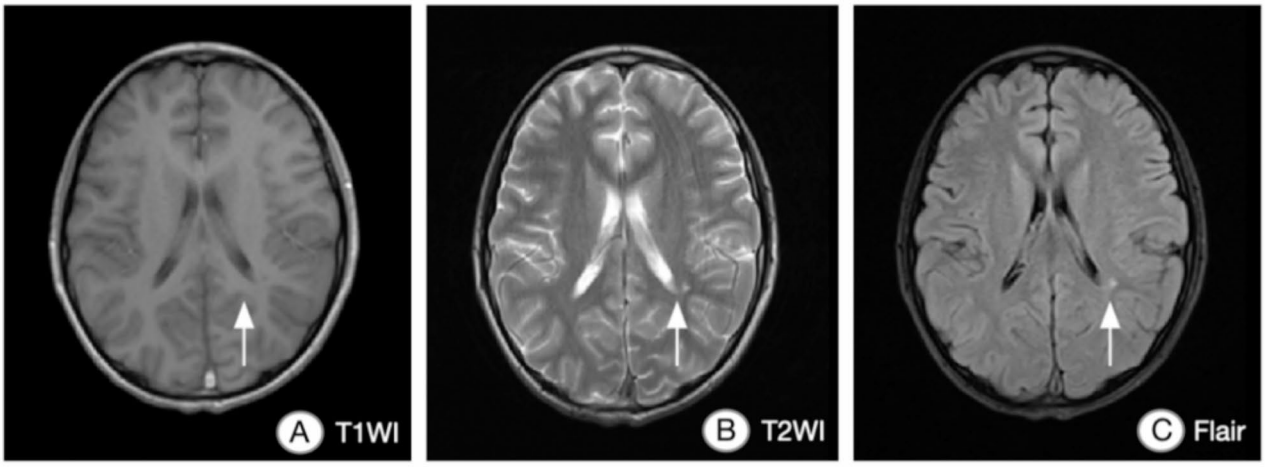

Fig. 1 Non-contrast MRI during the patient's initial admission demonstrates patchy prolonged T1/T2 signals in the white matter of the left posterior horn of the lateral ventricle (**A**, **B**), with corresponding FLAIR hyperintensity (**C**)

counts, along with CRP levels, decreased but remained above normal ranges. Ultimately, the patient's symptoms stabilized, and he was discharged from the hospital.

Between the ages of 13 and 18 years, the patient experienced developed recurrent oral and genital ulcerations accompanied by progressively manifesting left visual field deficits, fulfilling the 2014 International Criteria for Bechet's Disease [2]. At the age of 18, the patient was readmitted to the hospital due to right-sided limb weakness and ocular symptoms. Laboratory tests indicated persistent elevation of leukocytes and CRP, suggesting ongoing inflammation. Treatment with ceftriaxone-tazobactam showed no clinical improvement. The contrast-enhanced MRI revealed patchy long T1 and long T2 signals in the brainstem-thalamus-basal ganglia region (Figs. 2 and 3), with Fluid-Attenuated Inversion Recovery (FLAIR) hyperintensity, Restricted diffusion on Diffusion-Weighted Imaging (DWI), as well as hemorrhage in the left basal ganglia, indicating NBD. Additionally, cerebral venous thrombosis was identified on the Magnetic Resonance Venogram (MRV) (Fig. 2). Anticoagulation therapy was contraindicated due to suspected intracranial hemorrhage. The treatment regimen included glycerol fructose and mannitol for cerebral edema reduction, alongside intravenous methylprednisolone for anti-inflammatory management. After two weeks of treatment, the patient's clinical condition stabilized with improved right-sided limb mobility and was discharged.

Discussion

BD is characterized by systemic vasculitis with perivascular inflammatory infiltration [3]. The diagnosis of BD relies on clinical manifestations [2, 4], primarily including recurrent genital ulcers, oral aphthosis, and ocular inflammation. The pathergy test serves as a critical ancillary diagnostic tool. Approximately 10% of BD patients develop neurological involvement, termed NBD, which can lead to severe or permanent neurological deficits and significantly impair quality of life, representing a severe progression of BD [5]. NBD is exceptionally rare in pediatric populations. Early-stage NBD often lacks observable cardinal symptoms, making it prone to parental neglect. Compared to adults, pediatric BD patients exhibit a shorter interval between disease onset and NBD development [6]. While the pathergy test exhibits high specificity but low sensitivity in NBD [2]. Consequently, definitive diagnosis of NBD requires both established BD criteria and corroborative neuroimaging or cerebrospinal fluid abnormalities [1].

NBD is clinically and radiologically classified into two principal subtypes: parenchymal NBD and extraparenchymal neurovascular involvement. The parenchymal subtype predominates, characterized by headache, fever, and motor dysfunction [1]. Although the pathogenic mechanisms of NBD remain debated, the prevailing hypothesis centers on venocentric vasculitis, primarily affecting small venous walls in diencephalic and brainstem regions. Histopathological hallmarks include perivascular inflammatory infiltrates with focal neuronal loss and demyelinated areas, particularly in the brainstem [7]. Extraparenchymal neurovascular involvement typically manifests as cerebral venous sinus thrombosis (CVST) [8], which may result from inflammatory disruption of dural venous and vascular vaso vasorum or arise secondary to hypercoagulable states and impaired fibrinolysis. These mechanisms frequently coexist, creating a synergistic pathogenic loop [9]. Notably, CVST represents the most common neurovascular complication in pediatric NBD, whereas parenchymal lesions dominate in adult-onset cases [10].

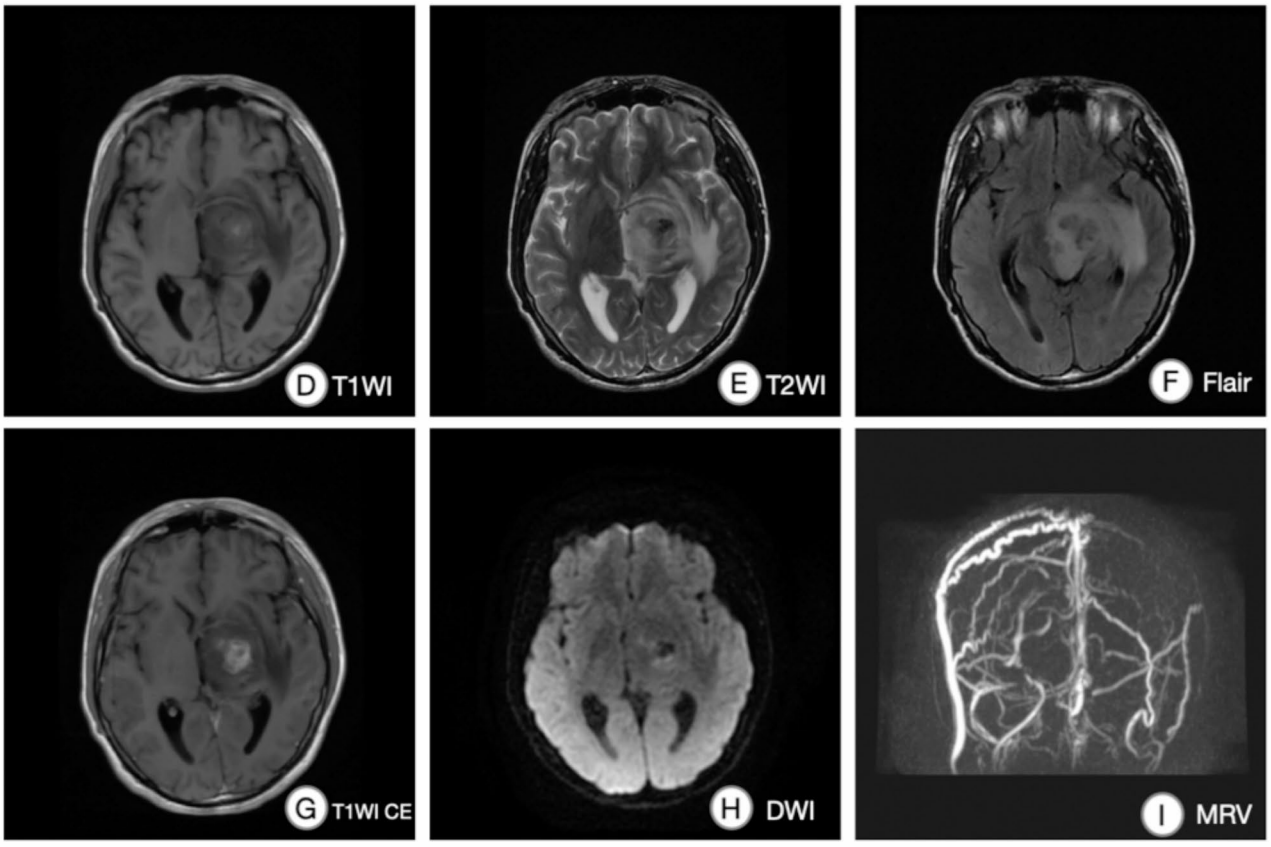

Fig. 2 Contrast-enhanced MRI at age 18 demonstrates Patchy prolonged T1/T2 signals are observed in the left basal ganglia and thalamus. Intracerebral hemorrhage is present in the basal ganglia. (**D**, **E**). Lesions show FLAIR hyperintensity(**F**). Heterogeneous ring-like enhancement on post-contrast(**G**) Restricted diffusion on DWI(H). MRV reveals bilateral transverse and sigmoid sinus stenosis, left transverse sinusthrombosis, and dilated right cortical veins draining into the superior sagittal sinus(**I**)

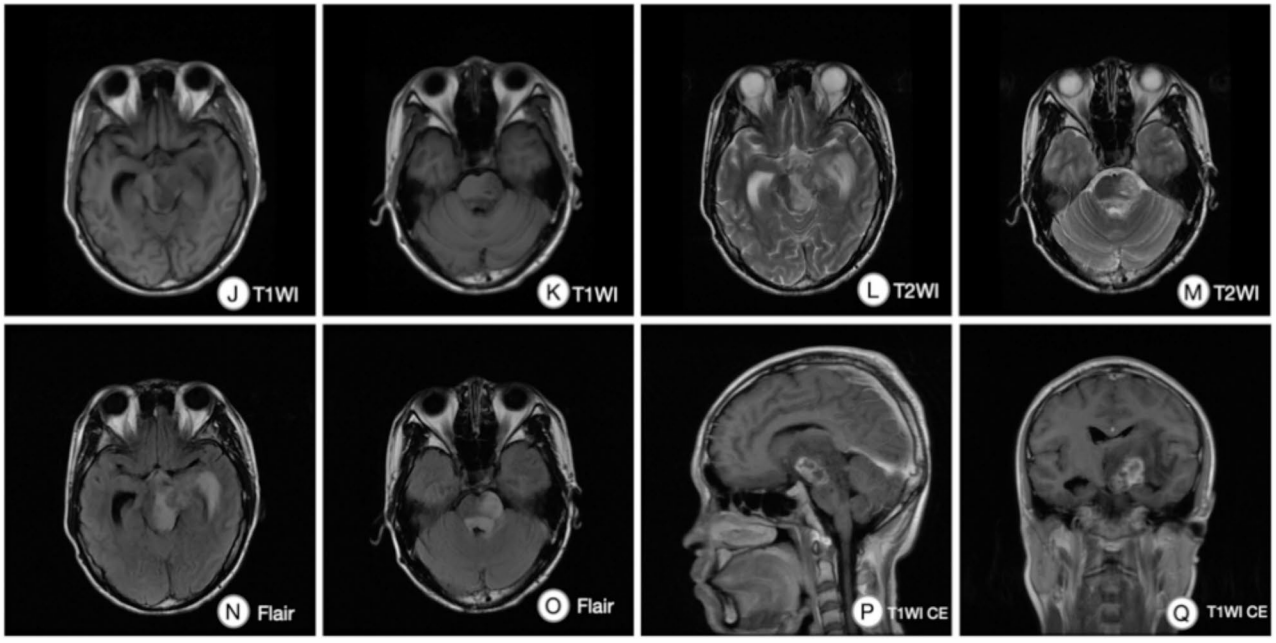

Fig. 3 Non-contrast MRI shows patchy prolonged T1/T2 signals in the left diencephalon, pons, and fourth ventricle periphery (**J**-**M**), with FLAIR hyperintensity (**N**, **O**) and heterogeneous ring-like enhancement post-contrast. Coronal imaging reveals the "cascade sign" (**P**, **Q**)

Consequently, neuroimaging plays a pivotal role in the early diagnosis of NBD. The brainstem represents the most characteristic neuroanatomical predilection site in NBD [5], with potential rostral extension to the midbrain, basal ganglia, and diencephalon. Coronal MRI may demonstrate a " the cascade sign " extending from the midbrain to thalamic regions [11]. Lesions typically exhibit hypointense to isointense signals on T1-weighted imaging (T1WI), with hyperintensity on T2-weighted imaging (T2WI) and FLAIR sequences [12]. With therapeutic intervention and disease progression, lesions may regress with diminished contrast enhancement, ultimately resolving on T2-weighted MRI while leaving focal atrophy at previous lesion sites [13]. MRV or computed tomography venography (CTV) can detect CVST formation [14]. Although parenchymal and extraparenchymal manifestations rarely coexist, this case demonstrates concurrent involvement. Notably, early-stage imaging in our patient revealed isolated abnormalities in the posterior horns of the lateral ventricles. This pattern may reflect inflammatory-mediated axonal injury and myelin breakdown, where perivascular neutrophil infiltration induces neuronal apoptosis and neurogliosis, ultimately manifesting as focal posterior horn lesions [13]. Progressive disease leads to gliosis, atrophic changes, and occasionally meningeal thickening with fibrosis [15]. A prior case report [16] documented analogous imaging findings in a 13-month-old female patient with NBD, demonstrating symmetric thalamic lesions extending to periventricular white matter and the right optic tract without contrast enhancement, mirroring the early-stage presentation observed here.

In this case, the patient initially presented with neurological involvement accompanied by leukocytosis, leading to an early misdiagnosis of bacterial infection. As the disease progressed, BD-specific manifestations subsequently emerged alongside parenchymal brain lesions and CVST. After rigorous exclusion of alternative etiologies, the diagnosis of NBD was confirmed. Pediatric NBD is characterized by an insidious onset with nonspecific initial symptoms. Published evidence indicates that neurological manifestations may precede systemic involvement in BD [17], while some cases never develop characteristic mucocutaneous lesions [18], significantly increasing diagnostic ambiguity. Due to the lack of specific laboratory biomarkers for definitive diagnosis in pediatric NBD patients, early implementation of multimodal cranial MRI– including advanced sequences such as FLAIR, DWI, and MR venography– is crucial for timely diagnosis.

The limitations of this case report include the initial testing restricted to inflammatory markers, lacking immunologic tests such as HLA-B51 and antinuclear antibodies, as well as cerebrospinal fluid analysis. Although the clinical manifestations and imaging findings were sufficient to confirm the diagnosis, the assessment process remained incomplete. Additionally, over the five-year disease progression period, the patient did not receive hospitalization, resulting in a lack of longitudinal data on changes in serological, imaging, and clinical parameters.

Conclusion

For pediatric patients with unexplained neurological symptoms and MRI findings showing lateral ventricular lesions or brainstem-thalamus-basal ganglia abnormalities, monitor closely for BD signs: recurrent oral and genital ulcers, skin lesions, and eye involvement. Assess coagulation function to detect cerebral venous thrombosis. Include BD in differential diagnosis when evaluating Central Nervous System infections or inflammation. Timely adjustment of treatment regimens is essential to improve the quality of life in patients, particularly children.

Abbreviations

BD	Bechet's syndrome
NBD	Neuro-Behçet
MRI	Syndrome magnetic resonance imaging
CRP	C-reactive protein
FLAIR	Fluid-attenuated inversion recovery
DWI	Diffusion-weighted imaging
MRV	Magnetic resonance venogram
CVST	Cerebral venous sinus thrombosis
T1WI	T1-weighted imaging
T2WI	T2-weighted imaging
CTV	Computed tomography venography

Acknowledgements

We thank the patient for granting permission to publish this information.

Author contributions

Concept and design: Wangsi Han and Shan Qi; data collection and analysis: Jiawen Tang and Jinyang Zhao; drafting of the article: Sihan Wang, Fan Xia; critical revision of the article for important intellectual content: Xiaohong Lyu; study supervision: Xiaohong Lyu. All the authors approved the final article.

Funding

This work was supported by the Liaoning Province Natural Science Doctoral Initiation Fund under Grant 2023-BSBA-141, and Research Project on Online Education Teaching for Postgraduate Students in Medical and Pharmaceutical Sciences in China under Grant B_YXC2024-02-01_59.

Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

Written informed consent to participate in this study was provided by the participants' legal guardian/next of kin. Written informed consent was obtained from the individual(s) and minor(s)' legal guardian/next of kin for the publication of any potentially identifiable images or data included in this article.

Consent for publication

Written informed consent has been obtained from the parents of the patient for the publication of clinical details and clinical images.

Competing interests
The authors declare no competing interests.

Received: 27 October 2024 / Accepted: 21 March 2025
Published online: 05 April 2025

References

1. Kalra S, Silman A, Akman-Demir G, Bohlega S, Borhani-Haghighi A, et al. Diagnosis and management of Neuro-Behçet's disease: international consensus recommendations. J Neurol. 2014;261(9):1662–76.
2. International Team for the Revision of the International Criteria for Behçet's Disease (ITR-ICBD). The international criteria for Behçet's disease (ICBD): a collaborative study of 27 countries on the sensitivity and specificity of the new criteria. J Eur Acad Dermatology Venereology: JEADV. 2014;28(3):338–47.
3. Yazici H, Seyahi E, Hatemi G, Yazici Y. Behçet syndrome: a contemporary view. Nat Rev Rheumatol. 2018;14(2):107–19.
4. Criteria for diagnosis of Behçet's disease. International study group for Behçet's disease. Lancet (London England). 1990;335(8697):1078–80.
5. Akman-Demir G, Serdaroglu P, Tasçi B. Clinical patterns of neurological involvement in Behçet's disease: evaluation of 200 patients. The Neuro-Behçet study group. Brain. 1999;122(Pt 11):2171–82.
6. Uluduz D, Kürtüncü M, Yapıcı Z, Seyahi E, Kasapçopur Ö, et al. Clinical characteristics of pediatric-onset neuro-Behçet disease. Neurology. 2011;77(21):1900–5.
7. Mnif N, Rajhi H, Mlika N, et al. Aspect En IRM du neuro-behcet. J Neuroradiol. 2006;33(4):250–4.
8. Uygunoglu U, Siva A. An uncommon disease included commonly in the differential diagnosis of neurological diseases: Neuro-Behçet's syndrome. J Neurol Sci. 2021;426:117436.
9. Al-Fahad SA, Al-Araji AH. Neuro-Behcet's disease in Iraq: a study of 40 patients. J Neurol Sci. 1999;170(2):105–11.
10. Mora P, Menozzi C, Orsoni JG, Rubino P, Ruffini L, et al. Neuro-Behçet's disease in childhood: a focus on the neuro-ophthalmolgical features. Orphanet J Rare Dis. 2013;8:18.
11. Cleaver J, Morrison H, Renowden SA, et al. An important diagnostic clue for neuro-Behçet's disease: the 'cascade sign'. Rheumatology. 2022;61(5):e130–1.
12. Borhani-Haghighi A, Kardeh B, Banerjee S, et al. Neuro-Behcet's disease: an update on diagnosis, differential diagnoses, and treatment. Multiple Scler Relat Disorders. 2020;39:101906.
13. Kidd DP. Neurological complications of Behçet's syndrome. J Neurol. 2017;264(10):2178–83.
14. Oliveira IM, Duarte JÁ, Dalaqua M, et al. Cerebral venous thrombosis: imaging patterns. Radiologia Brasileira. 2022;55(1):54.
15. Lee SH, Yoon PH, Park SJ, Kim DI. MRI findings in neuro-behçet's disease. Clin Radiol. 2001;56(6):485–94.
16. Pozzato M, Dilena R, Rogani G, et al. Can early-onset acquired demyelinating syndrome (ADS) Hide pediatric Behcet's disease? A case report. Front Pead. 2023;11:1175584.
17. Al-Araji A, Kidd DP. Neuro-Behçet's disease: epidemiology, clinical characteristics, and management. Lancet Neurol. 2009;8(2):192–204.
18. Lueck CJ, Pires M, McCartney AC, et al. Ocular and neurological Behçet's disease without orogenital ulceration? J Neurol Neurosurg Psychiatry. 1993;56(5):505–8.

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