Isolated sixth nerve palsy as a presentation of neuro-Behçet's disease

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ABSTRACT

Introduction: This study aims to report a case of neuro-Behçet's disease (NBD) presenting as isolated sixth nerve palsy in a patient with a history of inactive ocular Behçet's disease (BD), emphasizing the need for understanding diverse NBD manifestations.

Case Report: it is an observational case report. A 21-year-old BD patient, non-compliant with medications, presented with fever, headache, and double vision. Clinical, laboratory, and radiological assessments, including CT, MRI, and lumbar puncture, were conducted to diagnose and characterize NBD. Treatment involved prednisolone, azathioprine, and adalimumab. Neurological examination revealed sixth nerve palsy, supported by imaging findings indicative of vasculitis in the brainstem—a unique radiological presentation in the literature. Treatment led to symptom resolution in a 6-month follow-up.

Conclusion: This case reveals the diverse spectrum of NBD presentations, highlighting the importance of treatment compliance and contributing to a deeper understanding of the disease for refining therapeutic approaches.

CASE REPORT

INTRODUCTION

Behçet's disease (BD) is a chronic multisystemic inflammatory autoimmune disease of unknown origin. It is characterized by recurrent oral and genital ulcers, as well as uveitis with hypopyon [1]. Various tissues may be affected by inflammatory lesions, manifested as vascular, cutaneous, renal, gastrointestinal, articular, or neurological involvement [2]. Diagnostic criteria for this condition have been suggested by the International Study Group (ISG) for BD [3].

Ocular involvement occurs in 70% of BD patients, with anterior uveitis and posterior occlusive vasculitis being the most common manifestations of ocular BD. Recurrent attacks and inflammation may lead to the loss of useful vision early in the course of the disease [4].

Nervous system involvement is seen in about 5-10% of cases and commonly involves the brainstem [5]. Patients with neuro-Behçet's disease (NBD) exhibit numerous neurological symptoms, encompassing cerebral venous sinus thrombosis, central nervous system (CNS) parenchymal involvement, and others [6].

We report a rare case of isolated sixth nerve palsy as a presentation of NBD in a patient with a history of inactive ocular BD.

CASE DESCRIPTION

A 21-year-old man with a known history of BD diagnosed five years prior to presentation due to recurrent oral and genital aphthae and panuveitis. He was on a regimen of 500 mg mycophenolate mofetil and 5 mg prednisolone daily, and 40 mg adalimumab injections every 2 weeks, as well as prednisolone acetate drops once daily for both eyes. Unfortunately, he was non-adherent with his prescribed medications.

The patient sought care at our Emergency Department because of a four-day history of fever, headache, and double vision. . Vital signs were stable and within normal limits aside from a temperature of 39°C. Slit lamp examination showed normal anterior and posterior segments, with normal disc margins. Visual acuity was 20/20 with full color vision and normal visual fields in both eyes. Pupils were reactive to light and near with no afferent pupillary defect. Neurological examination revealed a right abduction deficit (Figure 1) with the patient reporting binocular diplopia on right gaze. Remaining

cranial nerves were intact, as were cerebellar, motor, and sensory functions. Gait examination was also within normal limits. CBC indicated polymorphonuclear leukocytosis and elevated ESR (51 mm/hr, normal range 0-15 mm/hr) and CRP (250 mg/L, normal range <5.0mg/L) levels were noted. Rheumatological workup for ANA, dsDNA, ANCA, RF, and complement were negative. Viral and other possible infectious etiologies such as malaria were also negative. Lumbar puncture yielded glucose of 3.50 mmol/L, and 0.40 g/L proteins which were within normal limits. WBC count was 240 uL, with 90% lymphocytes and 10% polymorphonuclear leukocytes. HSV PCR was negative. CT brain and CT venogram was unremarkable. MRI brain at the level of the tentorium and brainstem showed punctate foci of diffusion restriction, along the left uncus and dorsal pons. The lesions showed corresponding intermediate T2 signal intensity and low T1 signal. These features were suggestive of ischemic changes. (Figures 2,3). MRV was unremarkable. Both CSF and blood cultures were negative. To address the NBD flare, treatment was initiated with 40 mg prednisolone for two weeks followed by a 5 mg weekly tapering schedule, and 150 mg azathioprine daily, along with a 40 mg adalimumab injection every 2 weeks.

At a 6-month follow-up, the patient reported improved diplopia with resolution of symptoms.

DISCUSSION

BD is a chronic inflammatory condition of unknown origin, predominantly affecting individuals in the 20-40 age range. Its hallmark features include recurrent oral and genital ulcers, coupled with ocular inflammation [1].

Neurological involvement can occur in 5-10% of BD cases, with parenchymal disease making up 80% of the neurological presentations [5]. NBD frequently affects the brainstem and basal ganglia as a result of meningoencephalitis or vasculitis [6].

Neuro-ophthalmic manifestations of NBD are uncommon, with presentations such as optic neuritis, papillitis, oculomotor nerve palsies, and optic atrophy being reported in the literature.

A retrospective single-center review by Alghamdi et al. studied neuro-ophthalmic manifestations of BD in 217 patients and found neuro-ophthalmic manifestations to be present in 13% of NBD patients [7].

In the same study by Alghamdi et al., two patients were found to have third nerve palsy, while six were noted to have sixth nerve palsy. Similarly, a review by Benamour reported that the most common oculomotor palsy in NBD was sixth nerve palsy [8]. Interestingly, all sixth nerve palsies in these two reviews were attributed to high intracranial pressure (ICP). In contrast, our case presents a unique occurrence—an isolated sixth nerve palsy attributed to vascular occlusion in the brainstem, with normal ICP. A similar presentation was reported in two patients by Lew et al. [9]

Contrasting with the two cases reported by Lew et al. [9], which showed normal imaging, MRI findings for our patient were representative of ischemia due to vasculitis in the brainstem and left uncus, which is consistent with parenchymal disease. To our knowledge, this is the first case in the literature with this radiological presentation.

Our patient, with a history of ocular BD spanning several years, experienced a period of quiescence without systemic involvement. However, non-compliance with medications heightened the risk of a flare, which occurred as isolated NBD without concurrent ocular symptoms. Subsequent follow-up revealed a stable examination without any flare or progression.

In summary, our case highlights the diverse spectrum of NBD presentations and underscores the importance of compliance with treatment regimens. Unraveling such unique cases contributes to a deeper understanding of this complex disease and aids in refining therapeutic approaches.

TEACHING POINT

Presentations of NBD can be variable, such as isolated sixth nerve palsy. Recognizing atypical presentations and ensuring treatment compliance are crucial for effective management. Comprehensive diagnostics and tailored treatments are essential for symptom resolution and better patient outcomes.

Patient consent: An informed consent was obtained from the patient for the anonymous use and publication of data and images.

Competing interests: The Authors declare that there is no conflict of interest.

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FIGURES

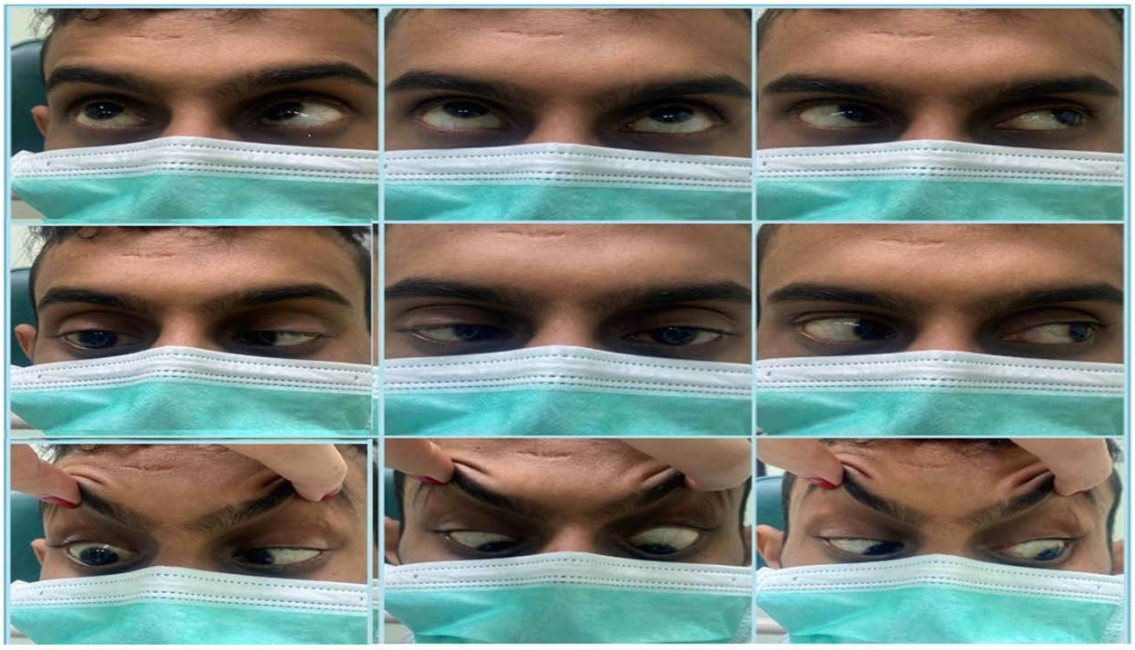

Figure 1: Eye movements in nine cardinal positions of gaze demonstrating a right esotropia in the primary position and a limitation of abduction in the right gaze.

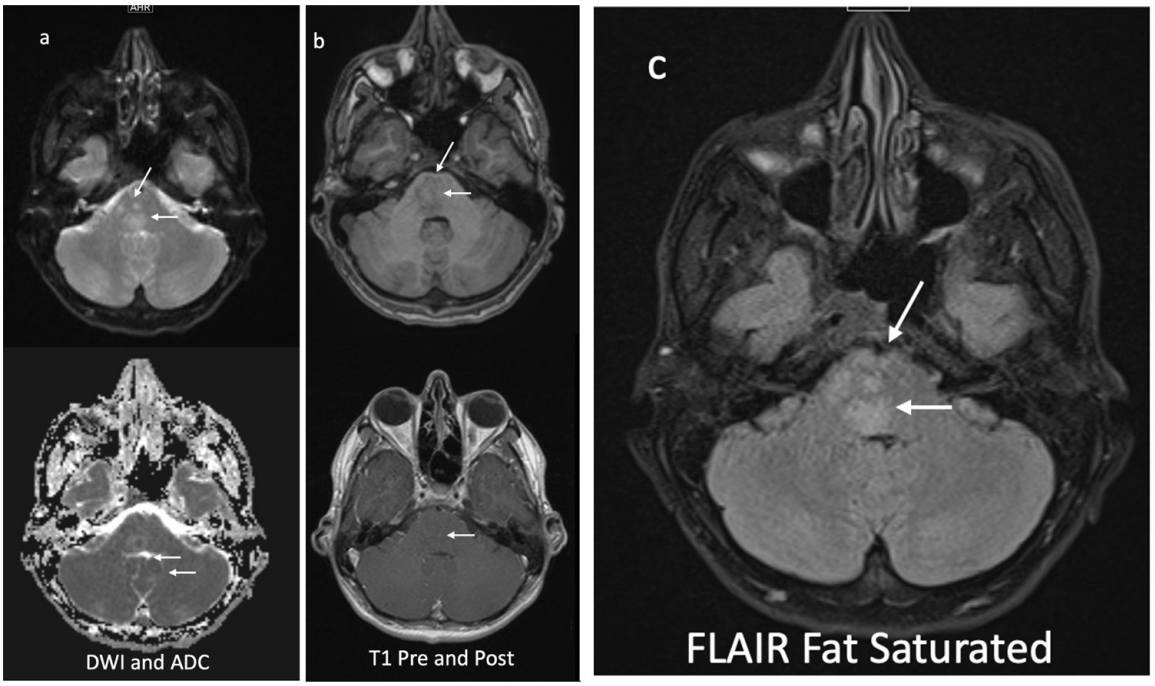

Figure 2: Selected axial DWI and ADC (Figure 2a) maps at the level of pons and middle cerebral peduncle, showing well defined punctate foci of diffusion restriction within the dorsal aspect of the right hemipons within the lateral lemniscus and superior olivary nuclei. (White arrows). Additional T1 pre and post contrast images (Figure 2b) show corresponding low T1 signal without contrast enhancement. On T2/FLAIR weighted sequence (Figure 2c), the same lesion show intermediate signal intensity without surrounding edema

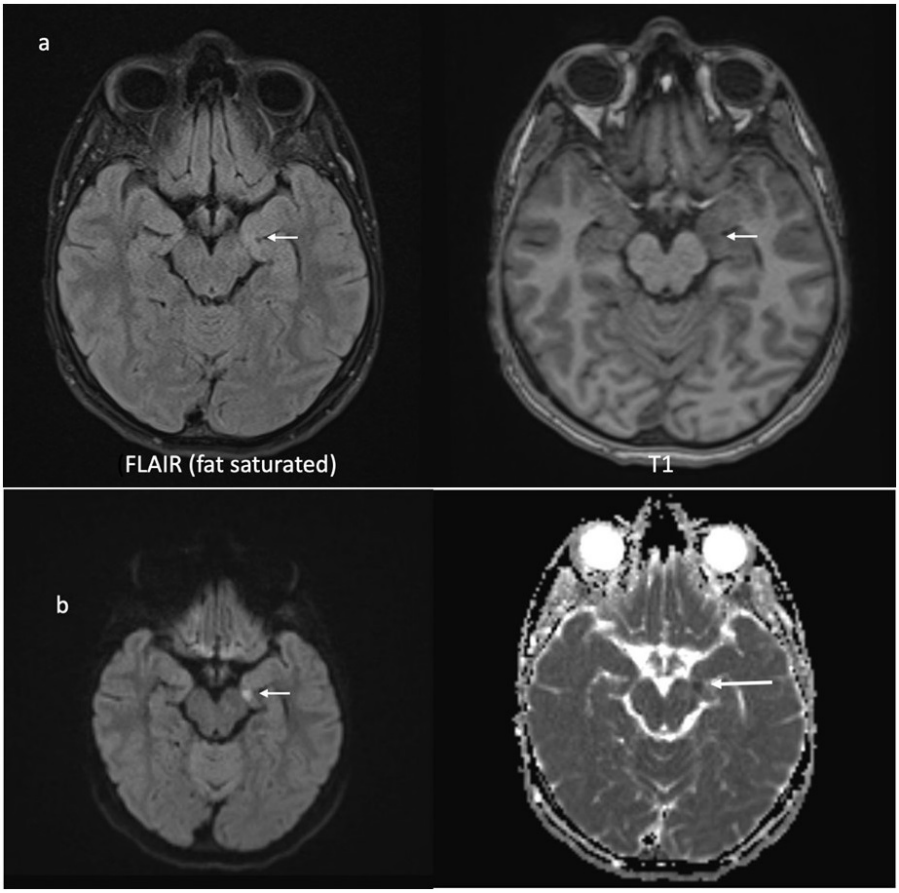

Figure 3: Additional focus of intermediate T2/FLAIR and low T1 signal intensity (Figure 3a) centered within the left uncus (White arrows), a corresponding diffusion restriction was also noted (Figure 3b, White arrows), eliciting similar signal characteristics as the aforementioned foci.

KEYWORDS

Bahcet disease; Abducens nerve palsy; Neuro-behcet.