Case Report

Pan Uveitis in a Patient Suspected of Multiple Sclerosis or Neuro-Behcet's Syndrome: A Case Report

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Abstract

Purpose: To report a case of pan uveitis in a patient initially suspected of having multiple sclerosis and later diagnosed with neuro-Behçet's syndrome.
Case Report: We present the case of a 35-year-old pregnant Iranian woman with a history of recurrent pan uveitis and sensory symptoms (Quadriparesis) who was initially treated based on a diagnosis of multiple sclerosis (MS). Upon reevaluation of the clinical and paraclinical findings, the condition of the patient was reclassified as neuro-Behçet's syndrome and treatment with corticosteroids and immunosuppressive agents was initiated.
Conclusion: The visual and neurologic manifestations of neuro-Behçet's syndrome can closely mimic those of multiple sclerosis, potentially leading to misdiagnosis. Accurate diagnosis in such cases is critical for achieving optimal treatment outcomes.

Keywords: Neuro-Behçet's Syndrome; Uveitis; Magnetic Resonance Imaging; Multiple Sclerosis; Differential Diagnosis.

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Introduction

Behçet's disease (BD) is a systemic vasculitis which might cause recurrent oral and genital aphthous ulcerations and uveitis [1]. Neuro-Behçet's disease (NBD), which involves neurological findings, is rare but challenging to diagnose and other conditions with similar clinical presentations can complicate its differential diagnosis [1]. Accurate diagnosis of NBD is essential for its treatment and to prevent long-term mortality and disability [1,2].

We report the case of a pregnant woman with pan uveitis who was initially misdiagnosed with multiple sclerosis (MS) and ultimately diagnosed and treated for NBD.

Case Report

All the procedures performed in the present study were approved by the Research Ethics Committee of the Shahid Beheshti University of Medical Sciences and were in accordance with the ethical standards of the institutional Human Research Review Committee (No. IR.SBMU.RETECH.REC.1402.667) and the 1964 Helsinki declaration and its later amendments. Also informed consent was obtained from the patient reported in this manuscript.

A 35-year-old pregnant Iranian woman (8 weeks, G2P1Ab0) was referred by an ophthalmologist to our hospital due to decreased vision. She had a history of recurrent bilateral pan uveitis at the age of 17, initially treated with 50 mg prednisolone and methotrexate, followed by a regimen of 10 mg prednisolone and methotrexate for two years. Upon recurrence of visual symptoms, azathioprine was added. However, during her first pregnancy, only prednisolone was continued, and the pregnancy concluded successfully.

Six years ago, she experienced dysarthria and sensory symptoms in her left upper limb. Four years later, she developed skin lesions consistent with erythema nodosum, although no oral or genital ulcers were detected. Treatment continued with 10 mg prednisolone and 50 mg azathioprine. Three years ago, she presented with quadriparesis and was diagnosed with multiple sclerosis (MS) based on compatible brain MRI findings. At that time, prednisolone and methotrexate were discontinued, and MS-specific treatment was initiated.

At this visit, she was referred to our hospital for reevaluation due to the progression of visual symptoms (uveitis). High-dose corticosteroids and mycophenolate mofetil (1 gram) were prescribed but were unsuccessful. Rheumatologic tests were negative, but skin aphthous lesions were noted. Azathioprine was discontinued due to her second pregnancy (8 weeks). She received several intraocular injections of triamcinolone. A pathergy test performed for the patient was positive.

At 14 weeks of pregnancy, her neurological symptoms flared, including decreased lower limb strength, paresis, and knee and hip pain. High-dose corticosteroids, 100 mg azathioprine, and 80 mg aspirin were initiated and later tapered to 10 mg prednisolone.

On physical examination, her blood pressure was 110/90 mmHg, pulse rate 90 beats per minute, respiratory rate 20 breaths per minute, oral temperature 36.5 °C, and oxygen saturation in room air 96 %. Laboratory tests revealed white blood cells (WBC) count of 10.7×10^3 /μL, lymphocytes 19 %, neutrophils 71 %, hemoglobin 11.2 g/dL, and platelets 288×10^3 /μL. C-reactive protein (CRP) was elevated at 8.3 mg/L, and erythrocyte sedimentation rate (ESR) was 53 mm/hr. Liver function tests, creatinine (0.5 mg/dL), creatine

kinase (CK), lactate dehydrogenase (LDH), and electrolytes were normal. Urinalysis showed 1+ blood, WBC count of 25–30, and Candida albicans in culture. Antinuclear antibodies (ANA), anti-double stranded DNA (Anti-dsDNA) antibodies, antiphospholipid syndrome (APS) antibodies, Anti-Ro, Anti-La, and Anti-neutrophil cytoplasm (ANCA) antibodies were negative. Complement levels, protein C, and protein S were normal. HLA-B5 was positive. IgG in cerebrospinal fluid (CSF) was slightly elevated at 10 mg/dL (normal ≤ 8), but CSF smear, oligoclonal bands, and neuromyelitis optica (NMO) antibodies were negative.

Neurologic examination revealed normal cranial nerve function. Muscle strength was 3/5 in both lower extremities (proximal and distal) and 5/5 in both upper extremities. Deep tendon reflexes were brisk (3 +), and plantar reflexes were positive. Brain MRI findings included generalized brain and cerebellar atrophy, temporal horns, corpus callosum agenesis with associated colpocephaly (racing car sign), T2 hyperintense foci in periventricular regions (confluent), U-fiber lesions, and a black hole on T1. A hyperintense lesion in the medulla was noted on diffusion-weighted magnetic resonance imaging (DWI). Lesions in the cerebellum, right middle cerebellar peduncle (MCP), and temporal regions were compatible with MS. However, the cervical MRI was inconsistent with MS lesions. The lumbar spine showed normal alignment and signal intensity without canal stenosis or cord abnormalities.

Ophthalmologic evaluation revealed no Marcus Gunn pupil and normal eye movements. Active uveitis was present in the left eye. Visual acuity was 3/10 in the right eye and hand motion in the left eye.

Altogether, clinical findings, imaging results, and a positive pathergy test supported the diagnosis of NBD. The patient was treated with 1 gram of corticosteroid pulse therapy for three days, leading to significant symptomatic relief. She was discharged on 40 mg prednisolone and 80 mg aspirin. Prednisolone was tapered to 30 mg over ten days, and 100 mg azathioprine was added.

The patient continued treatment with 15 mg prednisolone and 100 mg azathioprine until the end of her pregnancy, which concluded with preterm labor at 35 weeks. After delivery, she received three cyclophosphamide pulses, along with low-dose corticosteroids and azathioprine maintenance therapy. She has remained attack-free during one year of follow-up.

Discussion

Behçet's disease (BD) is a systemic vasculitis with various neurological manifestations. Although neurological complications are less frequent, they can cause severe disabilities [3].

There are several methods for differentiating NBD from conditions such as cerebrovascular accidents, multiple sclerosis (MS), and neuro-infections. Differentiating NBD from MS is particularly important. While MS is more common in women, NBD is generally seen in men. However, the onset age for both diseases is similar, typically between 20 and 40 years. Despite some overlapping clinical presentations, the frequency of these symptoms differs between the two diseases [4].

Optic neuritis, sensory symptoms, cerebellar symptoms such as dysarthria or ataxia, spinal pseudobulbar speech, headache, and cognitive-behavioral changes are more common in NBD [5]. Brainstem lesions that may extend to the diencephalic structures and basal ganglia are characteristic of NBD, whereas MS lesions

typically involve the corpus callosum and periventricular areas. A distinguishing feature of NBD is the disappearance or migration of lesions over time [6].

Previous case reports have highlighted the challenges of diagnosing NBD [2,3]. Tsalta-Mladenov et al., [3] reported a 25-year-old woman who presented with sudden onset of quadriparesis, general weakness, swallowing disorders, and slurred speech. Initially, she was diagnosed with cerebral infarction; however, the final diagnosis was NBD, and she was successfully treated with immunosuppressive and corticosteroid therapy. Similarly, Mantero et al., [7] described a 25-year-old woman who presented with decreased vision and sensory symptoms. She was initially treated for NBD, but further evaluation and characteristic brain lesions on imaging led to a revised diagnosis of MS, and her treatment was adjusted accordingly.

In our case, due to the presence of bilateral pan uveitis, skin aphthous lesions, erythema nodosum, a positive pathergy test, HLA-B5 positivity, and MRI findings suggestive of Behçet's disease, the patient was treated with cyclophosphamide and corticosteroids. Fortunately, the patient has not experienced any further attacks to date.

In summary, when a patient presents with neurological symptoms, it is crucial to evaluate all potential differential diagnoses thoroughly to reach an accurate diagnosis. Anti-tumor necrosis factor (anti-TNF) agents are one of the therapeutic options for NBD, although they may sometimes be associated with the progression of MS [8].

Conclusion

The visual and neurologic manifestations of neuro-Behçet's syndrome can closely resemble those of multiple sclerosis, potentially leading to misdiagnosis. Accurate diagnosis in such cases is critical for achieving optimal treatment outcomes.

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Footnotes and Financial Disclosures

Conflict of interest:

The authors have no conflict of interest with the subject matter of the present manuscript.