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Arti A. Patel

Amelia Stutman

Prarak Patel

Madhavi Capoccia

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# Elusive diagnosis of Behcet's disease in setting of painless vision loss

**Arti A Patel[1], Amelia Stutman[1], Prarak Patel[2] and Madhavi Capoccia[1]**

**Abstract**

Behcet's disease is a chronic, idiopathic vasculitis with multisystem involvement commonly characterized by the classic triad of oral lesions, genital ulcerations, and uveitis. We discuss the case of a 22-year-old woman with a long-standing history of oral ulcers and vulvovaginal burning who presented with acute painless uveitis. With this presentation, there was an initial concern for infectious retinitis for which she was started on systemic antiviral therapy. Subsequent infectious disease workup was ultimately negative. Given her medical history and current presentation, she was diagnosed and treated for an acute inflammatory episode of ocular Behcet's disease. The patient's vision returned to baseline prior to discharge after treatment with systemic glucocorticoids. The diagnosis of Behcet's disease in the setting of painless vision loss can oftentimes be elusive. However, it is important for clinicians to keep this condition as a differential diagnosis in patients presenting with acute onset uveitis as the progression of Behcet's disease can lead to severe vision loss and blindness without prompt and adequate treatment.

**Keywords**

Behcet's disease, uveitis, vision loss, ulcerations, steroids

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## Introduction

Behcet's disease (BD) is a rare, chronic, multisystem, relapsing vasculitis involving both arteries and veins.[1] As described by Dr. Hulusi Behcet in 1937, the classic triad of BD consists of recurrent oral lesions, genital ulcerations, and uveitis.[1] Ocular involvement is typically characterized by nongranulomatous inflammation with symptoms of photophobia, floaters, and visual loss.[2] In addition, patients can present with cardiac, pulmonary, musculoskeletal, and neurological manifestations.[1] Herein, we present the case of a young adult female with a history of recurrent oral ulcers and vulvovaginal burning who presented with acute on chronic onset of significant painless visual impairment.

## Case report

A 22-year-old female with a history of recurrent oral ulcers and vulvovaginal burning presented to the emergency department with painless bilateral uveitis of 8 months duration which acutely worsened in the past 2 weeks. Of note, the patient was seen outpatient by ophthalmology 5 days prior to presentation whereby a dilated eye exam demonstrated intraretinal hemorrhage in the right eye, dot-blot hemorrhage in the left eye, and bilateral vitritis with concern for cytomegalovirus retinitis (CMV) retinitis. Recommendations included 1% prednisolone eye drops twice a day, 1% cyclopentolate eye drops twice a day, and valganciclovir twice a day. However, the patient was only compliant with the cyclopentolate drops due to financial hardship. On presentation, she also endorsed floaters, blurry vision, and flashes of light. She denied any acute oral or genital symptoms; however, she reported a history of recurrent "canker sores" and burning in the vulvovaginal area that began approximately 5 years ago. She denied a history of genital ulcers. Physical exam was

[1]Department of Family Medicine, Lehigh Valley Health Network, Allentown, PA, USA
[2]College of Osteopathic Medicine, Kansas City University of Medicine and Biosciences, Kansas City, MO, USA

**Corresponding Author:**
Arti A Patel, Department of Family Medicine, Lehigh Valley Health Network, 1200 S Cedar Crest Blvd., Allentown, PA 18103, USA.
Email: Arti.Patel@lvhn.org

remarkable for photophobia and visual acuity of 20/50 in the right eye and 20/80 in the left eye with correction. Intraocular pressure in each eye was 16 mmHg by tonometry and pupils were equal, round, and reactive to light bilaterally. Extraocular movements were intact bilaterally. A slit lamp eye examination showed a flat macula and similar hemorrhages as seen in the ophthalmology office 5 days prior to presentation. Laboratory evaluation revealed an elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Extensive infectious disease testing for CMV, Herpes simplex virus (HSV), HIV, Varicella-zoster virus (VZV), syphilis, toxoplasmosis, tuberculosis, Hepatitis B and C was undergone, all of which resulted negative. Likewise, rheumatologic workup including testing for antinuclear antibody (ANA), anti-neutrophil cytoplasmic antibody (ANCA), angiotensin-converting enzyme (ACE), and human leukocyte antigen B27 (HLA-B27) was negative.

With concern for infectious retinitis, the patient was started on a course of IV acyclovir 10 mg/kg every 8 h. Her hospital course was complicated by acute kidney injury (AKI) secondary to acyclovir-induced nephrotoxicity; the medication was immediately discontinued, and she was given IV hydration to resolve the AKI. After discussion with ophthalmology about the patient's clinical course, presentation, and laboratory data, there was concern for BD as she met clinical diagnostic criteria. She was started on IV methylprednisolone 250 mg every 6 h for 3 days. The patient experienced significant improvement of photophobia and complete resolution of blurry vision and flashes of light.

At this point with the patient's negative infectious workup, it was felt a more detailed rheumatologic workup was needed. The patient was instructed to follow-up with outpatient rheumatology for skin pathergy testing to provide additional support to the diagnosis of BD and for long-term management. In that time, she was discharged home with a 2-week oral prednisone taper (50 mg daily for 7 days and 25 mg daily for the next 7 days) and valacyclovir to provide coverage for possible underlying retinitis.

Unfortunately due to financial hardship, this patient was unable to follow-up with the outpatient rheumatologist. Less than 3 months later she presented to the hospital with similar ocular symptoms and was readmitted for painless panuveitis. Of note, the patient reported she developed a painful ulcer on her labia a month prior that resolved spontaneously. She was given high-dose intravenous steroids for 3 days. Pathergy testing performed by rheumatology during this hospitalization was negative. Her vision returned to baseline and the patient was discharged with a scheduled outpatient rheumatology follow-up to determine long-term therapy.

## Discussion

BD remains largely a clinical diagnosis based on criteria from the International Criteria for Behcet's Disease (ICBD) as there has been no census on specific laboratory values for BD. The presence of ocular lesions, oral aphthous ulcers, and genital ulcers are each assigned 2 points.[3] Skin lesions, central nervous system involvement, and vascular manifestations are each assigned 1 point with an additional point for a positive skin pathergy test.[3] A total score of 4 or high is indicative of BD (Table 1). The above patient had oral ulcers and ocular lesions for a score of 4. On subsequent presentation she reported genital ulcers, bringing her score to a 6.

**Table 1.** International Criteria for Behcet's Disease.

| Symptoms | Points |
|---|---|
| Genital aphthosis | 2 |
| Ocular lesions | 2 |
| Oral aphthosis | 2 |
| Skin lesions | 1 |
| Neurologic manifestations | 1 |
| Vascular manifestations | 1 |
| Positive pathergy test | 1 |

Table 1: A pathergy test is optional and is not required for diagnosis. If performed and is positive, an additional point is assigned.

BD usually presents in individuals between the ages of 30–40 years old with an equal predilection for both sexes, though males typically have a more severe disease course.[4] Approximately 70% of people with BD will have some form of ocular involvement during their disease course, and 80% of those people will present with bilateral signs and symptoms.[4] Ocular BD is characterized by recurrent inflammatory episodes of uveitis flare-ups.[2] Patients who have inflammation of the anterior chamber of the eye will usually experience photophobia.[4] Those with posterior chamber inflammation will present with floaters and visual loss with retinal involvement which can ultimately progress to blindness, warranting prompt identification and treatment of the condition.[4] Despite aggressive management, many patients will still develop moderate or severe visual impairment.[2]

Treatment of BD involves management of the acute flare along with initiation of long-term maintenance therapy with the goal of reducing and preventing recurrences of BD. The acute inflammatory stage of ocular BD is typically treated with topical mydriatics and topical and systemic corticosteroids.[1] In a previously published case report, a 31-year-old man with chronic oral and genital lesions who was repeatedly misdiagnosed was eventually diagnosed with BD and immediately started on high-dose corticosteroids; after approximately a week of treatment, his lesions resolved.[5] However, practitioners should exercise caution for long-term usage of systemic steroids as they can result in significant adverse side effects. Traditionally steroid sparing, immunosuppressive agents have been used for long-term management of BD which includes azathioprine or cyclosporine.[6] Recently, biologic agents such as tumor necrosis

factor (TNF)—alpha inhibitors (infliximab) or anti-CD20 agents (rituximab) have been shown to induce remission when patients are resistant to the traditional agents.[6] Currently, there is no evidence that supports the use of antiviral therapy in the management of ocular BD.[7]

This case highlights the importance of broadening the differential in the setting of acute on chronic uveitis for proper management of the disease. In a published case series of 14 patients, researchers found the mean time to diagnosis of BD was 5.5 years.[8] Early in this patient's presentation, there was high suspicion for infectious retinitis, and the patient was started on IV acyclovir; however, this should have been discontinued earlier to prevent iatrogenic injury as the infectious disease workup was negative.[9] Furthermore, early identification and adequate treatment of her acute and chronic BD greatly reduced the severity of her ocular symptoms and prevented blindness, preserving sight in an otherwise healthy young female.

## Conclusion

BD is a chronic, multisystem, relapsing inflammatory condition that can affect multiple body systems. The classic symptom triad of BD is oral ulcers, genital lesions, and ocular involvement. This patient, who had a history of oral ulcers and genital burning, presented with painless bilateral uveitis and was ultimately found to have acute BD. The importance of having BD on the differential when a patient presents with uveitis cannot be understated. Adequate treatment of the acute inflammatory stage of ocular BD led to close resolution of her symptoms and prevented blindness.

### Author contributions

All authors listed have contributed sufficiently to the project to be included as authors. All authors read and approved the final manuscript.

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### ORCID iD

Arti A Patel https://orcid.org/0000-0002-6518-2103

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