Spontaneous resolution of unilateral Behcet’s associated neuroretinitis

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ABSTRACT

Purpose: Behcet’s disease is an immune-mediated condition which can commonly have ocular involvement. We present a case of Behcet’s associated neuroretinitis, which is a rare ocular manifestation of Behcet’s disease.

Observations: The patient experienced significant improvement in her neuroretinitis without initiation of treatment. After 28 days of observation there was spontaneous resolution of exam findings and return of Snellen visual acuity to 20/20.

Conclusion and Importance: There are only two other cases reported in the literature of Behcet’s associated neuroretinitis. Both cases report bilateral involvement with concomitant frosted branch angiitis. This is also the only reported case with spontaneous resolution of abnormal exam findings with return to 20/20 visual acuity. This case highlights the complexity of ocular involvement in Behcet’s disease.

1. Introduction

Behcet’s disease is an inflammatory disorder with highest prevalence along the silk road extending from east Asia to the Mediterranean basin. It is characterized by oral aphthous ulcers, genital ulcers, skin lesions and ocular inflammatory disease. Nongranulomatous inflammation and vaso-occlusive retinal vasculitis are among the most common ocular manifestations of Behcet disease. Neuroretinitis is a rare manifestation of Behcet’s disease in which only a few cases are reported in the literature. We present a unique case of unilateral neuroretinitis in a patient with a new diagnosis of Behcet’s disease.

2. Case report

A 26-year-old female presented with one week of blurry vision and eye pain in her left eye. Best corrected visual acuity was 20/20 in the right eye and 20/400 in the left eye. Intracocular pressure was 14 mm Hg in both eyes. No relative afferent pupillary defect was noted in either eye. On examination, the right eye showed no abnormal findings. Slit lamp examination of the left eye revealed trace cell and flare in the anterior chamber and 3+ anterior vitreous cells with 1+ vitreous haze according to the NEI scale. Fundoscopic evaluation revealed engorged venules, macular edema with associated exudates in a star-like configuration, optic disc edema and mild nasal peripapillary retinal hemorrhages in the left eye (Fig. 1). Optical coherence tomography of the left eye showed subretinal fluid and intra-retinal fluid, as well as hyper-reflective lesions consistent with the exudates seen clinically (Fig. 2).

A diagnosis of neuroretinitis was made. Of note, the patient had a recent history significant for recurrent oral ulcers, genital ulcers and erythema nodosum. The patient had not seen a primary care physician for these symptoms and had no history of being on immunomodulatory therapy. She had a previous history of an episode of acute unilateral non-granulomatous anterior uveitis in the left eye one-year prior, which was treated at a different ophthalmology practice and resolved with topical prednisolone acetate 1% eye drops. Laboratory work-up was initiated, and the results were as follows: negative Borrelia burgdorferi serum ELISA testing, negative rapid plasma reagen (RPR) and fluorescent treponemal antibody absorption test (FTA-Abs), negative Quantiferon gold assay (testing for Mycobacterium tuberculosis), and negative serum IgM/IgG for both Toxoplasma gondii and Bartonella henselae. Complete blood count and complete metabolic panel were within normal limits. Human Leukocyte Antigen typing was positive for HLA-B51. The patient was referred to rheumatology upon her initial visit for evaluation for systemic immunomodulatory therapy. Five days after initial presentation, the patient showed spontaneous improvement of visual acuity to 20/40 without intervention. On exam, there was significant improvement in macular edema as well as the amount of vitreous inflammation.
without initiation of pharmacotherapy. At this time, the results of her infectious laboratory workup were still pending. Given her significant clinical improvement, oral steroid therapy was deferred at this juncture. Eleven days after initial presentation, the patient had resolution of disc edema and macular edema with improvement in visual acuity to 20/30 without any treatment, although some mild subretinal fluid was seen on OCT at this time (Fig. 3). At 28 days following initial presentation, the patient’s Snellen visual acuity had improved to 20/20 without correction and the OCT had largely normalized, with just several hyperreflective spots seen within the outer nuclear layer, likely representing intraretinal exudate in the process of resorption (Fig. 3).

The patient was referred to rheumatology for initiation of definitive systemic immunosuppressive therapy for Behcet’s disease and for further evaluation of potential systemic involvement.

3. Discussion

We present a patient with the uncommon ocular manifestation of Behcet’s-associated neuroretinitis, which resolved spontaneously in this particular instance. While Behcet’s-associated neuroretinitis has been reported in the literature, there are no reports of spontaneous resolution of neuroretinitis. Our patient showed improvement in disc swelling, macular edema and visual acuity without acute treatment with corticosteroids or immunomodulatory therapy. Interestingly, two cases of neuroretinitis due to Behcet’s disease in the literature had concomitant frosted branch angiitis. This finding was not seen in this individual. Treatment of ocular Behcet’s typically involves oral corticosteroids in the acute phase with transition to other long term systemic immunosuppressants. A well-known marker for Behcet’s disease is human leukocyte antigen testing. HLA-B51 and/or HLA-B5 are carried by up to two thirds of patients with Behcet’s disease and increases the risk of development of Behcet’s Disease six-fold. Our patient was positive for HLA-B51.

According to Razzak et al. the most common ocular manifestation in Behcet’s disease is panuveitis. Other common manifestations include retinal vasculitis, posterior uveitis and cataract. Disc edema was the second least common manifestation with the least common being retinal ischemia. The most common extraocular manifestations were oral aphthous ulcers (100%), genital aphthous ulcers (41.3%), osteoarticular involvement (24%), and skin lesions (20.6%). The International Study Group for Behcet’s Disease developed the most widely used diagnostic criteria for Behcet’s Disease which simplified many of the published diagnostic criteria into a simpler system which requires the presence of oral ulceration plus any two of: genital ulceration, typical defined eye lesions, typical defined skin lesions or a positive pathergy test for diagnosis.

4. Conclusion

This case of a patient with unilateral neuroretinitis represents a rare finding in Behcet’s Disease. Two of the other cases reports found in the literature were of patients that had bilateral neuroretinitis with concomitant frosted branch angiitis. To our knowledge, this is the only reported case of unilateral neuroretinitis, and is a rare reported case of neuroretinitis without frosted branch angiitis. This unique case helps illustrate the complexity of ocular involvement of Behcet’s Disease.

Patient consent

Consent to publish this case report was not obtained. This report does not contain any personal information that may lead to the identification of the patient.

Funding

No funding or grant support.

Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Declaration of competing interest

The following authors have no financial disclosures: GS, SP, BD.
Acknowledgements

None.

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