A Brushfire in the Eye

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Presentation:

This month's case concerns a 75-year-old male who presented to us with two weeks of blurry vision and distortion in the left eye with persistent floaters. His medical history includes a history of tuberculosis infection treated 25 years ago. He also had been hospitalized recently for urinary tract infection and had placement of intraurethral catheter, recently discontinued. His ocular history involves a long standing atrophic macular scar in the right eye. He presented with visual acuity of 20/400 in both eyes, with intraocular pressures of 21 and 17. Pupils were equally round and reactive, and he had decreased temporal visual fields in the left eye. Exam of the right eye demonstrated inferior pavingstone and an old chronic central macular scar. (Figure 1A) Anterior examination in the left eye demonstrated rare cell and pigment in the anterior chamber with fine keratic precipitates. Posterior examination in the left eye had 2+ hazy view with vitreous cells and inferior pavingstone with a nasal patch of retinal whitening. (Figure 1B)

Narrowing the Differential:

The differential diagnosis of posterior uveitis is wide, and a thorough review of systems and medical history must be adequately performed to narrow the differential. In any patient with anterior uveitis, it is extremely important to dilate the patient to rule out posterior involvement. In this case, the most likely diagnosis and most vison threatening is the presence of viral acute retinal necrosis. Other possibilities include endogenous endophthalmitis from recent catheterization, or a granuloma from reactivated tuberculosis as indicated by his past medical history. Other less likely etiologies to consider would be sarcoidosis, syphilis, Bechet's syndrome, Toxoplasmosis, and lymphoma.

Plan and Outcome:

Due to the high suspicion of acute retinal necrosis, the patient underwent a diagnostic anterior chamber tap which was sent for viral PCR. Uveitic work up for

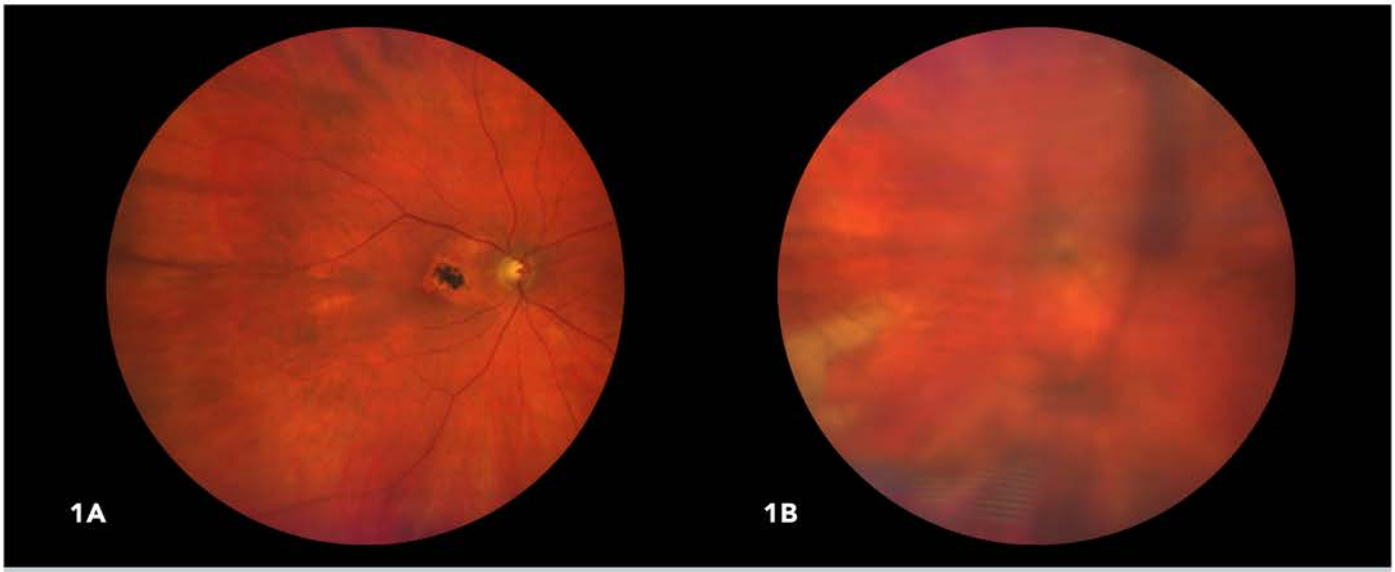

Figure 1. Fundus photograph of the right eye (A) demonstrated central macular scar. Left eye (B) demonstrated vitreous haze and debris with inferonasal retinal whitening.

syphilis and sarcoid were found to be negative. Additionally, the patient was sent for infectious disease consultation and they ruled out any active tuberculosis infection. The patient underwent treatment with valacyclovir 1g three times daily. He was monitored every few days and the lesion showed signs of improvement and the margins began to atrophy and become inactive. (Figure 2). Visual acuity remained stable at 20/400 due to the presence of vitreous debris. The diagnostic tap PCR eventually returned positive for VZV, and negative for other viral etiologies. It is important to not delay treatment while waiting for results.

Discussion:

Acute retinal necrosis is an inflammatory viral disease that can progress rapidly and result in devastating vision loss from retinal necrosis if diagnosis and treatment are not initiated promptly and accurately. Although it is associated with an immunocompromised state, it has been shown to be virulent in otherwise healthy immunocompetent individuals regardless of age and gender. Herpes simplex, herpes zoster, and cytomegalovirus are the usual culprits. Pathophysiology is thought to be due to reactivation a latent virus. Infection occurs in two stages.

The initial acute herpetic stage begins with the infiltration of the virus into the retinal tissues, generally starting peripherally and working towards the posterior pole circumferentially. The exam typically demonstrates a classic triad of retinal whitening with intraretinal hemorrhages, retinal or choroidal vasculitis, and a moderate vitritis. Left untreated this can progress rapidly and involve the entire retina resulting in necrosis and permanent vision loss. Treatment during this phase involves arresting the progress by the initiation of systemic acyclovir or valacyclovir. Research has shown that oral medications lead to similar concentrations in the vitreous cavity when compared to IV medications, although IV may be considered in cases of immunocompromised patients. Renal function must be monitored during treatment. Intravitreal therapy foscarnet may be considered in those cases with resistant disease or fovea threatening disease. Steroids are controversial but it has been suggested they can improve treatment if initiated after 48 hours, but they should never be used without antivirals. This phase generally lasts approximately 4-6 weeks. Some studies have advocated the use of aspirin for prevention of occlusive vasculitis but strong evidence is lacking.

The second cicatricial stage of the disease occurs as a consequence of resolving inflammation. As the acute phase ends, scar tissue in the form of tractional contractile membranes develops at the vitreoretinal interface. These membranes pull on the now weakened atrophic retina which can result in the formation of isolated or combined tractional and rhegmatogenous detachments which can be very challenging to repair. Reports in the literature estimate the incidence of retinal detachment to be as high as 46.9%. Visual prognosis is guarded once detachment occurs. The data to support the role of

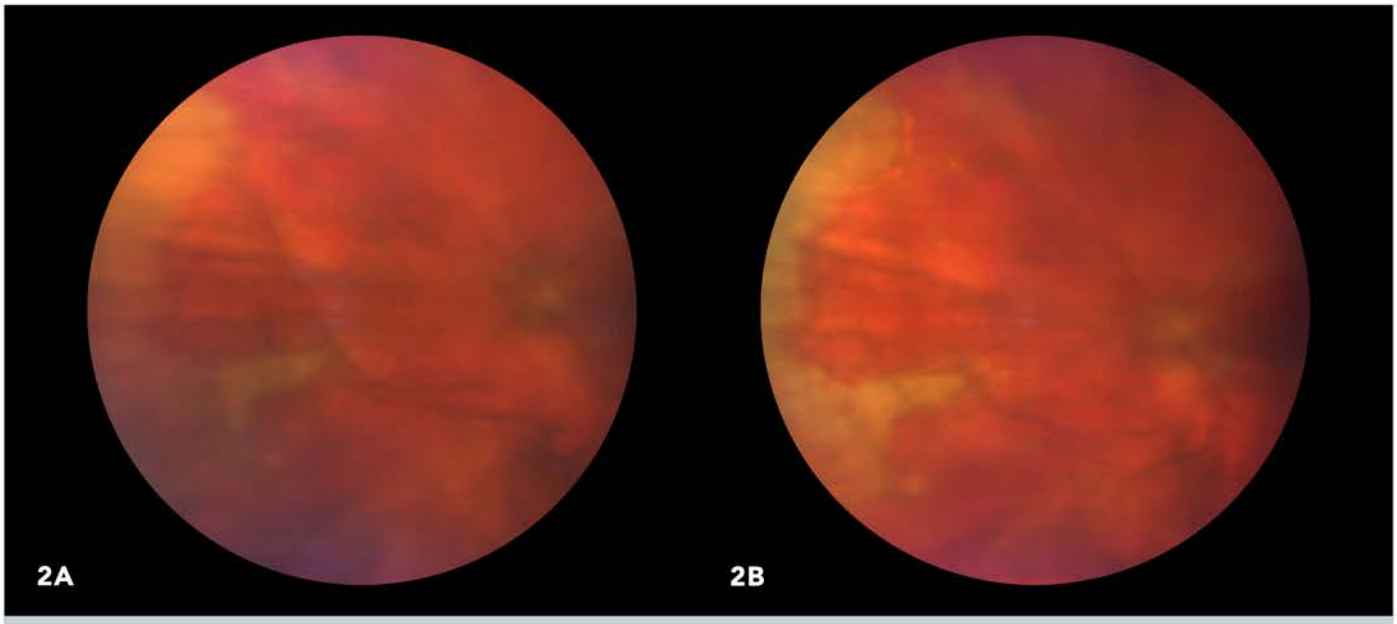

Figure 2. Day 6 (A) and day 13 (B) after initiation of treatment with valacyclovir demonstrated resolving retinitis with development of atrophic borders demonstrating resolving infection. Visual acuity was limited by the presence of vitreous debris although color fungus photography showed clearance of visual axis.

prophylactic laser retinopexy or vitrectomy prior to detachment is inconclusive.

Overall, a good prognosis depends on the sparing of the macula during the infectious and cicatricial stages. Visual acuity may be limited by vitreous debris and although tempting, removal through vitrectomy should be avoided until resolution of both stages. Most importantly, the non-involved eye must be monitored carefully for life, as contralateral involvement has been shown to occur, even decades after the initial episode. Although the duration of prophylaxis is not standardized, most studies advocate lifelong prophylaxis with valacyclovir 1 gram once daily to prevent recurrence.

References:

1. Kopplin LJ, Thomas AS, Cramer S, Kim YH, Yeh S, Lauer AK, Flaxel CJ. Long-Term Surgical Outcomes of Retinal Detachment Associated With Acute Retinal Necrosis. Ophthalmic Surg Lasers Imaging Retina. 2016 Jul 1;47(7):660-4.

2. Baltinas J, Lightman S, Tomkins-Netzer O. Comparing Treatment of Acute Retinal Necrosis With Either Oral Valacyclovir or Intravenous Acyclovir. Am J Ophthalmol. 2018 Apr;188:173-180.

3. Donovan CP, Levison AL, Lowder CY, Martin DF, Srivastava SK. Delayed recurrence of acute retinal necrosis (ARN): A case series.J Clin Virol. 2016 Jul;80:68-71.

4. Risseeuw S, de Boer JH, Ten Dam-van Loon NH, van Leeuwen R. Risk of Rhegmatogenous Retinal Detachment in Acute Retinal Necrosis With and Without Prophylactic Intervention. Am J Ophthalmol. 2019 Oct;206:140-148.

5. Tibbetts , Shah CP, Young LH, Duker JS, Maguire JI, Morley MG. Treatment of acute retinal necrosis. Ophthalmology. 2010 Apr;117(4):818-24.

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