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Monroe
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Acute Retinal Necrosis

Case presentation:

A 34 year old female with no past medical history presented with a one week history of left eye redness and blurry vision. Her visual acuity was 20/20 in the right eye and 20/30 in the left eye. Her pupils, extraocular movements, and intraocular pressures were within normal limits. Slit lamp exam was normal in the right eye but revealed 1+ conjunctival injection and 2+ anterior chamber cell in the left eye.

Dilated fundus exam was normal in the right eye. In the left eye, the exam revealed 1+ vitreous haze, disc edema, diffuse retinal hemorrhages, vascular sheathing, and peripheral retinal whitening (Figure 1). Optical coherence tomography (OCT) of the left eye showed hyperreflectivity of the inner retinal layers as well as peripapillary subretinal fluid consistent with the disc edema (Figure 2).

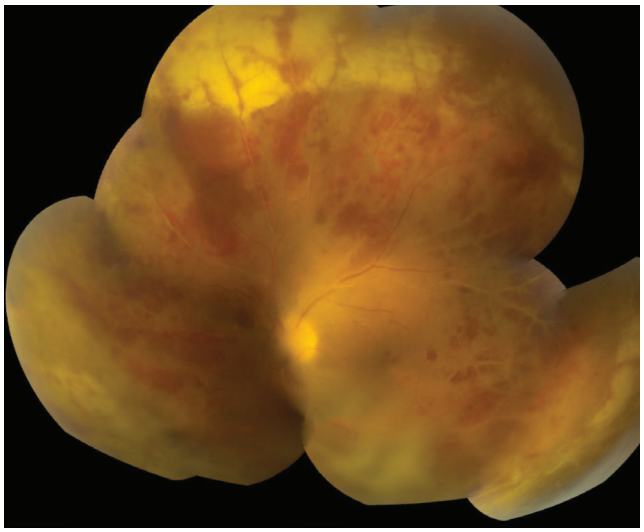

Figure 1: Color fundus photo of left eye at presentation. Exam shows vitreous haze, disc edema, vascular sheathing, retinal hemorrhages, and peripheral retinal whitening.

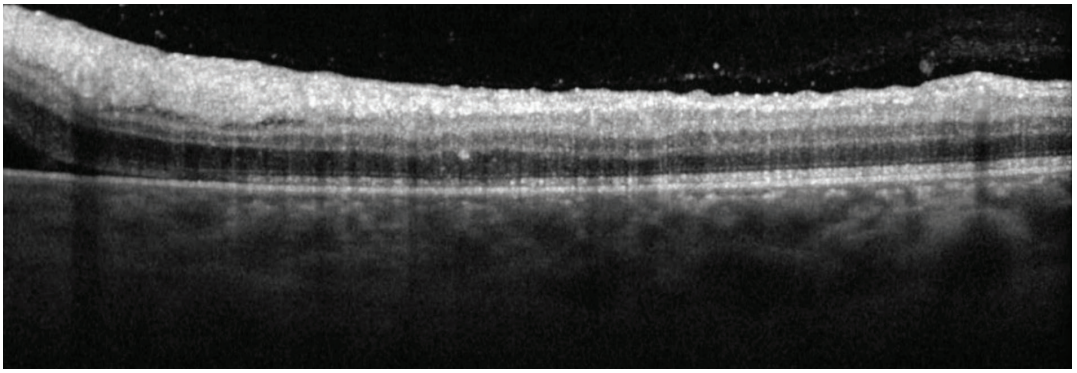

Figure 2 Optical coherence tomography image of left inferior macula at presentation. Inner retinal hyperreflectivity can be seen due to retinitis, and peripapillary subretinal fluid is present due to disc edema. Vitreous cells can be seen as well.

Due to the high suspicion for viral retinitis, the patient was sent to the hospital for further testing and management. Upon arrival, she was started on intravenous acyclovir. Anterior chamber tap of the left eye was performed, and the fluid was sent for viral polymerase chain reaction (PCR) testing. Subsequently, intravitreal injection of ganciclovir was performed. The PCR test was positive for varicella zoster virus (VZV). Despite treatment, progression of the disease caused her vision to decrease to 20/100 in the left eye. She continued to receive twice weekly intravitreal injections of ganciclovir. Her intravenous acyclovir was transitioned to oral valacyclovir 2 grams three times daily, and she was started on oral prednisone 30 milligrams to control the intraocular inflammation.

Two weeks after diagnosis, the patient unfortunately developed a rhegmatogenous retinal detachment that extended 360 degrees in the periphery. She underwent retinal detachment repair with scleral buckle, pars plana vitrectomy, endolaser, and silicone oil. As her retinitis resolved, the oral prednisone was tapered, and the valacyclovir was decreased to a prophylactic dose of 1 gram daily. The retina remained attached, and the silicone oil was successfully removed several months after the initial surgery. Her visual acuity in the left eye at her final follow-up was 20/250.

Discussion:

Acute retinal necrosis (ARN) is a potentially blinding necrotizing retinopathy caused by members of the Herpes virus family, and most commonly varicella zoster virus[1-7]. ARN is rare, with an estimated incidence of one case per 2 million people per year [8-9]. ARN often affects otherwise healthy adults with no racial or sex-based predilection. However, there is evidence that suggests an underlying genetic component, in particular with certain human leukocyteantigen (HLA) expressions such as HLA-DQw7 and DR4 in Caucasian patients and HLA-Aw33, B44, and DRw6 antigens in Japanese patients[10-11].

Most cases of ARN are unilateral, but up to 30% of cases can result in bilateral ARN [12]. Therefore, when ARN is suspected, it is important to examine the fellow eye carefully for any involvement. Examination findings can include anterior chamber and vitreous inflammation, retinal vasculitis, retinal hemorrhages, and multifocal, peripheral, and confluent areas of retinal whitening consistent with retinitis. Optic disc edema can be seen in cases of optic nerve involvement. The peripheral lesions can rapidly progress to the posterior pole [13]. In later stages of the disease, vitreous traction and retinal atrophy can commonly lead to retinal detachment, with prior studies reporting rates from 47% to 66% [14-17]. Other complications of ARN include hypotony, phthisis, proliferative vitreoretinpathy, epiretinal membrane, macular edema, and optic nerve atrophy[13].

Diagnosis of ARN is made clinically as defined by the American Uveitis Society criteria: 1) at least one focus of peripheral retinal necrosis with well-defined borders, 2) rapid circumferential progression when antiviral therapy is not instituted, 3) occlusive vasculopathy (with arteritis), and 4) prominent vitreous and anterior chamber inflammation[18]. Differential diagnosis includes progressive outer retinal necrosis, cytomegalovirus (CMV) retinitis, syphilis, toxoplasmosis, tuberculosis, endophthalmitis, Behcet syndrome, and intraocular lymphoma.
Testing for HIV/AIDS is also important to assess for immunocompromised status. More recently, PCR testing has become a useful tool to establish a diagnosis. The testing can be performed using small samples of either aqueous and vitreous humor, although aqeous is usually preferable because it is safer to obtain[13].

Treatment of ARN should be initiated shortly after suspected diagnosis due to the rapid progression of untreated disease. Systemic antivirals are important in treating the retinitis and reducing the risk of fellow eye involvement. For active retinitis, treatment can be initiated with intravenous acyclovir 10 mg/kg three times per day or oral valacyclovir 2 grams three or four times per day. Both drugs have similar bioavailability, and either medication can be used to manage ARN. After induction therapy, standard practice is to maintain patients on a prophylactic dose (usually valacyclovir 1 gram daily) for at least six months[19].

Intravitreal agents are commonly used in combination with systemic antivirals. Intravitreal injection of foscarnet or ganciclovir can be used to achieve immediate therapeutic levels of intraocular antiviral drug [19]. Topical and oral steroids are also commonly used in conjunction with antivirals in order to decrease inflammation[13]. Oral steroids are typically started at least 24-48 hours after initiation of antiviral therapy.

Laser retinopexy has been used as a possible method to prevent retinal detachment. Early pars plana vitrectomy has also been proposed to remove intraocular inflammation and reduce risk of retinal detachment by releasing areas of traction. Some small case series have been performed, but there currently no clear evidence that either treatment is effective in the prophlyaxis of retinal detachment.

When rhegmatogenous retinal detachment occurs, surgical techniques for repair include pars plana vitrectomy, scleral buckle, endolaser, long-acting gas, and/or silicone oil tamponade. Unfortunately, even in some cases of anatomic success, visual outcome can be poor especially when the infection involves the macula or optic nerve[20].

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Michael Park, MD

NJRetina Welcomes
Michael Park, MD to our medical staff

What is your philosophy of care?
Treat patients as I would treat my family.

Why did you pursue a career in ophthalmology?

As a medical student, I had the opportunity to observe an ophthalmologist perform cataract surgery on a patient. I saw how the 10-minute surgery drastically improved the patient's vision and quality of life and knew I wanted to take part in the mission to give sight back to my patients.

What made you choose Retina as your specialty area?
The retina gives us the opportunity to diagnose and assess underlying systemic health issues. It allows us to become involved in our patients' medical care not only as their ophthalmologist but also as their physician. I also appreciate the diversity and complexity of the surgical cases in our field.

Are there any unique cases you have experienced?
During my fellowship, I performed an autologous retinal transplant on a patient with a large macular hole. It was quite unique in that I counseled the patient to remain face up as opposed to face down during the immediate postoperative course.

Why did you choose to join NJRetina?
I grew up in New Jersey and have family here, so I was motivated to return after fellowship training. NJRetina has a large presence in New Jersey, and I knew I wanted to join a well-run and well-established practice where I have the means to provide the highest quality of care to my patients. The physicians at NJRetina also have a strong reputation of being the innovators and leaders in the field, and are pioneering many of the clinical research trials that I am interested in taking part in. I am excited to join such an esteemed group!

What are some of your personal interests?
Playing tennis and golf, traveling the world, and learning about different cultures.

Are you fluent in any language aside from English?
Korean and Spanish

At the forefront of clinical research

NJRetina continuously conducts clinical trials at key locations. Our clinical research coordinators will be happy to discuss the inclusion/ exclusion criteria or any other aspect of these studies with you or your patients. If you have any questions, please feel free to contact:

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Enrolling Studies:

Wet AMD

Edison
Opthea Coast: A Phase 3, Multicenter, Double-masked, Randomized Study to Evaluate the Efficacy and Safety of Intravitreal OPT-302 in Combination with Aflibercept,
Compared with Aflibercept Alone, in Participants with Neovascular Age-related Macular Degeneration (nAMD)

Toms River
Opthea ShORe: A Phase 3, Multicenter, Double-masked, Randomized Study to Evaluate the Efficacy and Safety of Intravitreal OPT-302 in Combination with Ranibizumab,
Compared with Ranibizumab Alone, in Participants with Neovascular Age-related Macular Degeneration (nAMD)

Teaneck
Luna: A Multi-Center, Randomized, Double-Masked Phase 2 Study to Assess Safety and Efficacy of ADVM-022 (AAV.7m8-aflibercept) in Anti-VEGF Treatment Experienced Patients with Neovascular (Wet) Age related Macular Degeneration

Diabetic Retinopathy

Teaneck
Ocuterra: A Phase 2 Randomized, Double-Masked, Vehicle Controlled, Multicenter Study to Evaluate the Safety and Efficacy of OTT166 Ophthalmic Solution in the Treatment of Diabetic Retinopathy (DR)

RVO

Teaneck, Toms River, Edison
Bayer Study: A Randomized, Double-Masked, Active-Controlled, Phase 3 Study of the Efficacy and Safety of Aflibercept 8 mg With Macula Edema due to Retinal Vein Occlusion

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