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Case Report

Reversal of Retinal Ischemia from Idiopathic Obliterative Vasculitis with Treatment

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Abstract

Obliterative vasculitis is an inflammatory process affecting the retinal vessels that can lead to permanent vision loss due to inadequate blood supply. The management of idiopathic obliterative vasculitis can be tricky with few options including topical steroids, hyperbaric oxygen, and balloon angioplasty. Here we report the case of profound retinal reperfusion following a course of systemic steroids in an elderly gentleman with idiopathic obliterative vasculitis and a history of retinal ischemia.

Keywords: *Retinal Ischemia; Idiopathic Obliterative Vasculitis*

Introduction

Obliterative vasculitis is a rare but significant disease process which can affect the retinal vessels leading to inflammation including vitritis, retinal edema, retinal and vitreous hemorrhage, retinal neovascularization, neovascular glaucoma, and in severe cases, irreversible ischemia [1]. Fundoscopic exam and fluorescein angiography (FA) are essential for early diagnosis and management of obliterative vasculitis [1]. We present a patient with severe obliterative vasculitis who exhibited significant ischemia, which reversed after treatment.

Case Report

A 72-year-old male with history of well controlled B-cell Chronic Lymphocytic Leukemia (CLL) on rituximab, hypertension, cataract surgery with intracameral moxifloxacin OS 9 months and OD 4 months prior to presentation, and poor visual acuity in the right eye which had been previously attributed to vitreous hemorrhage presumably from a retinal vascular occlusion 2 - 3 months prior to presentation and was being followed by an outside retina specialist presented to our clinic with worsening vision in the left eye. The visual acuity in the right eye was counting fingers and left eye had declined from 20/70 to counting fingers. Clinical findings showed neovascular glaucoma and vitreous hemorrhage in the right eye without a view to the fundus. The left eye had vitritis with retinal whitening, intraretinal hemorrhages, and vasculitis with several perivascular white lesions in the retina. Full systemic physical exam was otherwise unremarkable. FA of the left eye revealed nonperfusion of the major vessels with significant retinal ischemia in the macula and midperiphery, as well as staining of the optic nerve and vessels (Figure 1). Given the acute vitritis, retinal whitening, and his relative immune compromise with CLL on rituximab, viral retinitis was suspected and inpatient management with intravenous acyclovir, anterior chamber paracentesis sent for viral PCR, and intravitreal foscarnet and ganciclovir OS were administered. Twenty-four hours after systemic and local treatment with anti-virals, high dose steroids (1 mg/kg) were also initiated to cover possible inflammatory etiologies. Lab testing for syphilis, tuberculosis

and toxoplasma gondii was negative. Following a negative PCR for viral etiologies, intravenous and oral acyclovir were discontinued. High dose oral steroids were continued while further testing was pending. Serologic studies, blood culture, and lumbar puncture were negative for any infectious or neoplastic causes. Inflammatory workup was unremarkable including normal ESR and CRP. MRI/MRA of the brain, transthoracic echocardiogram, and carotid ultrasound were unrevealing.

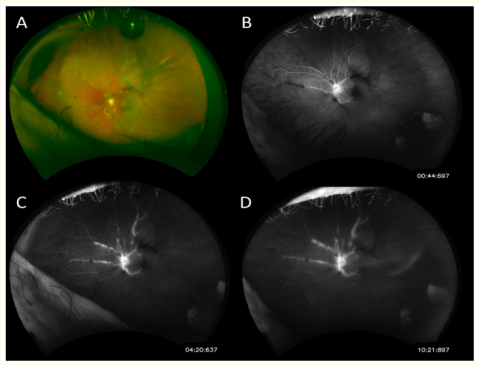

Figure 1: *Pretreatment fundus photo and fluorescein angiography.*
(A) Pretreatment fundus photo showing significant retinal whitening, intraretinal hemorrhage, and several perivascular white lesions in the left eye. (B, C, D) Pretreatment early, mid, and late phase fluorescein angiography of the left eye revealed significant retinal ischemia, diffuse capillary drop out, perivascular sheathing, and vascular staining with poor perfusion of the superior-temporal and inferior-temporal arcades.

During the hospital course, the patient developed NVI OS which was treated with intravitreal Avastin on day 6 followed by panretinal photocoagulation OS. Vision OS improved to 20/100 with reversal of the ischemia seen on FA after 15 days of steroid treatment (Figure 2). A vitrectomy with panretinal photocoagulation was performed for non-clearing vitreous hemorrhage in the right eye with minimal improvement in vision due to widespread retinal ischemia.

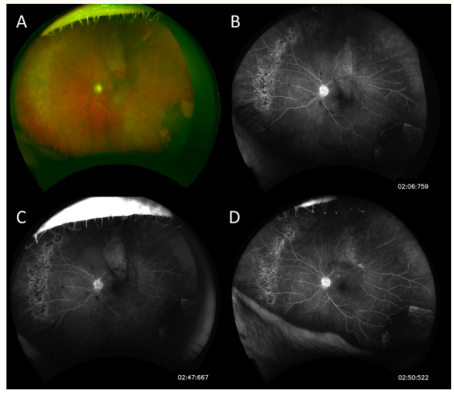

Figure 2: *Post treatment fundus photo and fluorescein angiography.*
(A) Fundus photo 15 days post treatment initiation showing significant reversal of the retinal whitening, improvement of the intraretinal hemorrhages and perivascular white lesions, and PRP laser scars nasally in the left eye. (B) Fluorescein angiography 15 days, (C) 26 days and (D) 2.5 months post treatment showing reversal of retinal ischemia, perivascular sheathing, vascular staining and improvement of the capillary drop out and perfusion of the superior and inferior arcades.

Discussion

There are several reports in the literature presenting reversal of retinal ischemia using various treatments including balloon angioplasty for stenotic ophthalmic artery (Hwang), carotid angioplasty and stenting for ocular ischemic syndrome (Marx), and a case of transient reversal of macular ischemia secondary to radiation retinopathy with intravitreal Ozurdex implant (Verma) [2-4]. Hadanny., *et al.* showed in a retrospective study that patients with Central Retinal Artery Occlusion had reversibility of vision loss with hyperbaric oxygen therapy in the absence of cherry red spot formation [5].

Reversal of retinal ischemia after treatment is unusual but has been previously reported following local steroid treatment [4]. A case of CLL presenting as both optic disc edema and branch retinal artery occlusion was shown to respond well to a combination of intrathecal chemotherapy and IV steroids [6]. Optic neuropathy due to CLL has also been reported and was found to be responsive to systemic steroids with worsening vision upon dose reduction [7].

Our patient demonstrated an idiopathic obliterative vasculitis with a robust response to systemic steroids with exceptional improvement as visualized on FA. This patient showed remarkable reperfusion as shown in figure 2. This is highly unusual given his history of retinal ischemia. It is also important to note that oncology thought the patient's CLL was well under control and a dosage increase in the rituximab did not relieve ophthalmic symptoms.

Conclusion

With the excellent response to oral steroids seen in this patient with idiopathic obliterative vasculitis and retinal ischemia, further investigation should be performed to elucidate the efficacy of oral steroids on patients with idiopathic obliterative vasculitis.

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