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Case Report

Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis: A Case of Extensive Macular Edema

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Abstract

Idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) is a rare retinal vasculitis that can occasionally present with macular edema. A treatment paradigm has been proposed to treat its neovascular sequalae but only case reports have described treatment of IRVAN-induced macular edema. We present a case of a middle-aged woman with macular edema secondary to IRVAN which was unresponsive to a single intravitreal injection of ranibizumab followed by a sub-tenon's capsule injection of Kenalog.

Keywords: Retinal Vasculitis; Aneurysms; Macular Edema

Introduction

Idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) was first characterized by Chang et al in 1995 in a case series of 10 patients [1], although there are earlier reported cases describing the same pathology [2]. It is an aptly named condition consisting of retinal vasculitis, branch point arterial aneurysms, neuroretinitis, and retinal exudation [3]. Peripheral retinal non-perfusion is typically present as well [1,3] although there are cases without this feature, especially in children [4,5]. As a result of the retinal ischemia, neovascularization and its sequelae can occur [1,3,6]. This disorder is not associated with any systemic vasculidities [3,7].

IRVAN is a purely ophthalmic disease which has a female preponderance and typically affects patients in their fourth and fifth decades of life [3]. Patients may be asymptomatic and diagnosed incidentally or present with decreased vision or flashes and floaters. However, while previously thought to be a self-limited disease, recent cases with neovascular complications have caused poor vision or no light perception [3].

A grading system, based on intravenous fluorescein angiography (IVFA) has been proposed by Samuel et al as follows: Stage 1 – neuroretinitis, retinal vasculitis, macroaneurysms, exudation, Stage 2 – capillary nonperfusion, Stage 3 – posterior segment neovascularization, Stage 4 – anterior segment neovascularization, Stage 5 – neovascular glaucoma [3].

IRVAN is an idiopathic inflammatory condition [6] which upon development of capillary non-perfusion may follow the course of any other ischemic retinal condition with neovascularization and its sequelae of neovascular glaucoma, vitreous hemorrhage, and tractional retinal detachment [3].

Many different forms of therapy have been utilized to treat IRVAN. The earlier inflammatory stages have been treated with periocular [8], intravitreal [8,9], and oral steroids [10], steroid-sparing immunomodulators [6,11], and very recently intravitreal anti-vascular endothelial growth factor injections (anti-VEGF) [12] all with varying success. The later ischemic stages (stage 3 or greater or stage 2 with widespread capillary nonperfusion) are treated with photocoagulation and/or anti-VEGF [13-15].

Case Report

Clinical History: A 54 year-old Hispanic female with past medical history of well-controlled type 2 diabetes mellitus on metformin presented with decreased visual acuity for one month. No history of recent travel, trauma, insect bites, or changes in medication. She denied any flashes or floaters. Her review of systems was negative.

Exam

Her best-corrected visual acuity at presentation was 20/20 in the right eye and 20/400 in the left eye. Her pupils were equal, round, and reactive and without relative afferent pupillary defect.

Confrontational visual fields were full and her intraocular pressure was within normal limits. Anterior segment examination was normal and without neovascularization of the iris. Vitreous was clear with sharp and hyperemic nerves in both eyes. Retinal vein sheathing was present with branch-point arterial aneurysms, massive perivascular exudation in the posterior pole, and perivascular intraretinal hemorrhages. Her macula was normal in the right eye with center-involving exudates in the left. Her peripheral retinal examination showed hard exudates, edema, and areas of intraretinal hemorrhages (Figure 1).

Figure 1: Fundus photo, left eye. Black arrows, macular and perivascular hard exudates. White solid arrow, venous sheathing. White dashed arrows, arterial branch-point aneurysms.

Imaging

OCT macula revealed retinal thickening with edema and hard exudates in both eyes; the edema was center involving in the left eye (Figure 2). Fluorescein angiography shows branch point arterial aneurysms with associated arteritis, capillary leakage, disc leakage, and areas of peripheral capillary nonperfusion (Figure 3).

Laboratory workup revealed

WBC wnl, platelets wnl, cholesterol 243 (high), triglycerides 75 (normal) LDL 155 (high), HDL 73, (high), Vitamin D 22.1 (low), A1C 6.5 (elevated), ESR 23 (wnl), CRP 3.0 (wnl), RPR nonreactive, Myeloperoxidase Ab (p-ANCA) 2.1 (wnl), Serine protease 3 Ab (c-ANCA) 3.0 (wnl), ACE 31 (wnl).

Figure 2: OCT Macula, left eye. A, At initial presentation. B, Three months following intravitreal injection of ranibizumab. C, Three months following subtenon injection of kenalog.

Figure 3: Fluorescein angiogram, left eye. Note the branch point aneurysms, peripheral capillary non-perfusion, perivascular leakage.

Treatment

The patient was treated with a single intravitreal injection of ranibizumab 0.5 mg in the left eye. 10 weeks later she was given a sub-tenon's capsule injection of Kenalog 40 mg/ml (STK).

Outcome

The patient's left eye showed minimal, if any, improvement in the amount of retinal edema over ten-week follow-up (Figure 2b) which prompted an STK injection. This resulted in a moderate decrease in paracentral fluid on OCT macula (Figure 2c).

Discussion

IRVAN is a rare retinal vasculitis and has been increasingly diagnosed within the last decade. The largest case series, by Samuel et al., included 22 patients. Optimal treatment options are still largely unknown.

The presentation of this patient is unique in a number of ways. First in terms of her age, being 54 years old. IRVAN typically affects patients in their 30's and 40's [1,3] although there are numerous reports of patients in the first, second, and third decade of life [3,4,8-10]. One patient in the large case series by Samuel et al was 60 at presentation with the rest being age 47 or younger (mean 31.5 years) [3]. Additionally, while there are a handful of cases of patients presenting later in life, with the oldest being 80, the majority of these older patients have a unilateral disease [5,6,12,15-18].

The literature contains an abundance of reports of anti-VEGF being used successfully for neovascular, stage 3 or greater, IRVAN [13-15], however, to our knowledge, only Massicotte et all used anti-VEGF purely for the purpose of treating macular exudation in patients with stage 2 IRVAN [12]. Marín-Lambíes also successfully resolved macular edema with the use of anti-VEGF, however her primary use of the drug was to treat neovascularization [13]. Our patient with stage 2 IRVAN received intravitreal ranibizumab with the goal of resolving macular subretinal fluid due to retinal ischemia and also STK as an anti-inflammatory agent for vasculitis. If there is no IOP increase, one can consider a combined treatment of intravitreal steroids and anti-VEGF therapy.

This raises the question of what causes macular edema in patients with IRVAN. While leakage from neovascularization likely plays a role in some patients, the presence of edema in our patient with Stage 2 IRVAN proves that it cannot be the sole cause. An alternate hypothesis is that the inflammatory nature of IRVAN results in the upregulation of various cytokines and inflammatory mediators, likely including but not limited to VEGF, which augment the permeability of the blood-retina barrier [19]. If VEGF were the sole-offender the edema would have been responsive to anti-VEGF injections, however this was not seen in our case.

Conclusion

Our report adds an additional IRVAN case to the ophthalmic literature. This patient is unique in both her age and the severity of her macular edema relative to stage 2 IRVAN.

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