Aflibercept for the Treatment of Macular Edema Secondary to Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis Syndrome

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Introduction

Idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) syndrome is a rare clinical entity of unknown etiology.[1,2] IRVAN syndrome typically affects healthy female individuals in their youth or middle age.[1] This disease can be diagnosed by three major criteria: retinal vasculitis, retinal microaneurysms, neuroretinitis, and three minor criteria: peripheral capillary nonperfusion, retinal neovascularization, or hyperemia of the optic disk with macular exudation.[1]

The pathogenesis of IRVAN is unclear. However, it is widely accepted that is a consequence of intraocular inflammation due to the presence of mild intraocular inflammation, vasculitis, and the formation of epiretinal membranes.[3] Some authors propose that IRVAN is associated with hypersensitivity reactions to tubercular antigens reporting a link between IRVAN and systemic disease.[4] Literature reports that treatment outcomes for IRVAN syndrome are significantly influenced by early diagnosis.[1,5] Treatment options include pan-retinal photocoagulation, vitrectomy, corticosteroids, and cryotherapy.[1,5] We report a case of idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) syndrome in a patient whose cystoid macular edema was successfully treated with aflibercept and pan-retinal photocoagulation.

Case Report

A 56-year-old male was sent for a consultation to our uveitis service for further evaluation after an intravitreal angiogram that revealed symmetric retinal ischemia for 360 degrees in both eyes. Fundus examination revealed an aneurysm, neuroretinitis, and occlusive vasculitis consistent with a diagnosis of IRVAN syndrome (Figure 1). OCT examination revealed cystoid macular edema (CME) of the left eye. Chest x-ray revealed minimal prominence of interstitial markings. The patient had a positive QuantiFERON Tb-Gold test and was treated for Tb with a one-year course of isoniazid and pyrimethamine. Further work-up for other infectious and autoimmune etiologies was negative. The initial treatment consisted of bilateral pan-retinal photocoagulation to the areas of peripheral ischemia, which was provided in a fragmented fashion over the course of eight months. Soon after the diagnosis, he received treatment with two intravitreal aflibercept (2 mg/0.5 mL) injections, one month apart, to the left eye. Subsequently, at four months following presentation, he developed CME in the right eye, which was treated with a single intravitreal aflibercept (2 mg/0.5 mL) injection. At his last follow-up visit, four years after initial presentation, the patient remained asymptomatic, with 20/20 visual acuity in both eyes, and no evidence of CME recurrence.

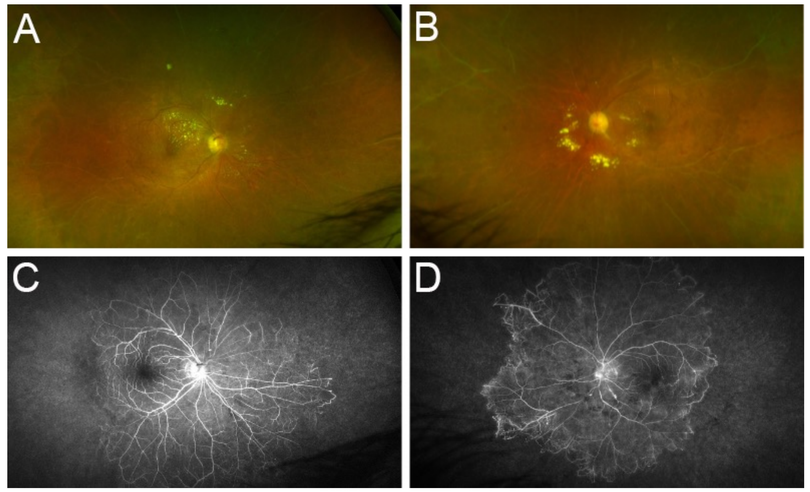

Figure 1. Fundus imaging at initial presentation. Right (A) and left (B) color photographs revealed aneurysm, neuroretinitis, and occlusive vasculitis. Right (C) and left (D) intravitreal fluorescein angiogram revealed diffuse leakage in the xchoroidal watershed zone and severe marked capillary drop-out for 360 degrees OU.

Discussion

Our case suggests that aflibercept may serve as an adjuvant to the standard treatment with pan-retinal photocoagulation, especially in cases that are present with associated macular edema. Despite our successful response to the described treatment, prospective studies are needed to assess its effectiveness and safety.

References

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