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Case report

The fluorescein angiographic photodiagnosis of idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) syndrome: Outcome of combined therapy

Buğra Karasu*, Derya Özkan, Gürkan Erdoğan, Özgür Artunay

University of Health Sciences, Beyoglu Eye Training and Research Hospital, Istanbul, Turkey

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ABSTRACT

Idiopathic retinal vasculitis, aneurysms and neuroretinitis (IRVAN) syndrome is a disease characterized by multiple retinal macroanurysms, neuroretinitis and ischemia in peripheral vessels, which are difficult to diagnose and cause visual loss if delayed. It consists of 5 stages and causes irreversible vision loss with severe complications after stage 2. In this report, photodiagnosis and combined treatment are defined in a Turkish patient with IRVAN syndrome during 6 months of follow-up. Fundus fluorescein angiography (FFA) showed that bilateral aneurysms in retinal vessels as well as aneurysms and ischemic regions were observed in the inferotemporal retina of the left eye. Argon laser photocoagulation was performed to ischemic regions at the left eye. 2 months later best corrected visual acuities were 20/20 and counting fingers from 1 m in the right and left eyes, respectively. Fundus and OCT images showed that an increase in exudations was observed at the left eye and intravitreal injection of dexametasone implant was considered. Three months after initial presentation, best corrected visual acuities were 20/20 and counting fingers from 2 m in right and left eyes, consequetively. In the last fundus and OCT images, the exudations decreased and disappeared on left eye.

In this patient, we could not detect an increase in vision due to damage of photoreceptor cells because of subretinal exudation. In the shed-light of this case, the combination therapy seems to improved the anatomical and functional outcomes in IRVAN syndrome however close follow-up and frequent examinations should be prioritized.

1. Introduction

Idiopathic retinal vasculitis, aneurysms and neuroretinitis (IRVAN) syndrome was first defined in 1983 by Kincaid and Schatz. It is a retinal disorder of unknown etiology. IRVAN is diagnosed with three major fundus findings such as multiple aneurysmal dilatations, retinal vasculitis and neuroretinitis in arterial bifurcation as well as three minor fundus findings including peripheral capillary perfusion disorder, retinal neovascularization and macular exudation [1].

Idiopathic retinal vasculitis, aneurysms and neuroretinitis (IRVAN) syndrome is a retinal vasculitic disease that is frequently seen in young women. The staging of IRVAN syndrome based on ocular findings was defined by Samuel et al. (Table 1) [1,2]. Visual deterioration usually results from severe complications such as retinal ischemia, neovascular glaucoma or macular exudation. Therefore, there is no common approach in the management of treatment in these patients.

There are several approaches to advocating different treatment protocols in this disease such as photocoagulation, intraocular steroids [3], cryotherapy, intravitreal bevacizumab [4], or intravitreal ranibizumab [5]. However, the progression of the disease can be stable in some patients and following without treatment [6].

This is a case report in which demonstrating the efficacy of intravitreal dexametasone implant and photocoagulation therapy in a patient with IRVAN syndrome.

2. Case report

A 60-year-old male patient was admitted to the retina clinic of the Beyoglu Eye Training and Research Hospital with visual impairment and metamorphopsia on his left eye. On passing through the patient's documents, he had no history of any systemic disease. In ophthalmologic assessment, best-corrected visual acuities were counting 30/100

* Corresponding author at: University of Health Sciences, Beyoglu Eye Training and Research Hospital, Bereketzade Mah, Bereketzade Sok. No: 2, Beyoğlu, İstanbul, Turkey.
E-mail address: bugra_karasu@hotmail.com (B. Karasu).

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Table 1
Stages of IRVAN syndrome.

Stage Ocular findings	Suggested Theraphy
Macroaneurysms, exudation, neuroretinitis, retinal vasculitis	High-dose prednisolone, photocoagulation, local steroid injections
Capillary nonperfusion (angiographic evidence)	High-dose prednisolone, photocoagulation, local steroid injections
Posterior segment neovascularization of disc or elsewhere and/or vitreous hemorrhage	High-dose prednisolone, local steroid injections, photocoagulation, anti-VEGF agents, cryotherapy
Anterior segment neovascularization (rubeosis iridis)	Surgery, anti-VEGF agents, cryotherapy
Neovascular glaucoma	Surgery and treatment for glaucoma

Initially, the induction process followed by sequelae of retinal ischemia leading to neovascularization is followed. Fibrotic complications are observed in the 4th and 5th stages and cause permanent vision loss. Surgery may require vitrectomy due to pars planitis.

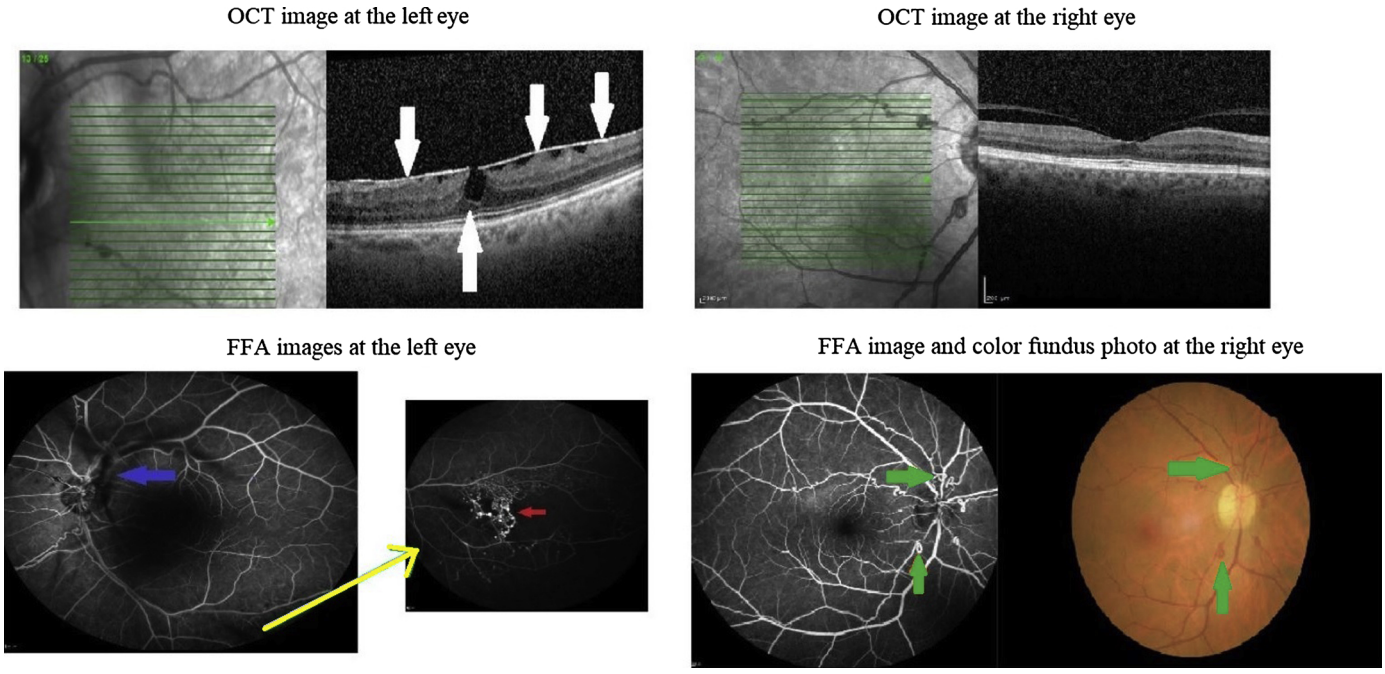


Fig. 1. White arrows indicate epiretinal membrane and lamellar hole at the left eye. OCT image of the right eye. Blue arrow shows increase in vitreous condensation. Red arrow shows aneurysmal dilatation areas with ischemic region. The green arrows show the aneurysms around the optic disc.

and 20/20, in the left and right eyes, respectively. There were no significant findings in the anterior segment examination in both eyes. Intraocular pressure measurement using the Goldmann Applanation tonometer was 16- and 15-mm Hg in the right and left eye, respectively. Fundus examination revealed that an epiretinal membrane with a lamellar hole on the left eye. Fundus fluorescein angiography showed that bilateral aneurysms in retinal vessels located at optic disc as well as aneurysms and ischemic regions which were observed at the inferotemporal retina of the left eye and also increased density in the vitreous (Fig. 1).

No abnormality was found in the fundus fluorescein angiography except that retinal vessel aneurysms at the right eye located at the optic disc (Fig. 1). As a result, the patient's fundus findings were evaluated and IRVAN syndrome was diagnosed. Detailed research was considered. Biochemical blood markers were normal. There was no abnormality in cranial magnetic resonance imaging. Argon laser photocoagulation was performed to ischemic regions after fundus fluorescein angiography at the left eye.

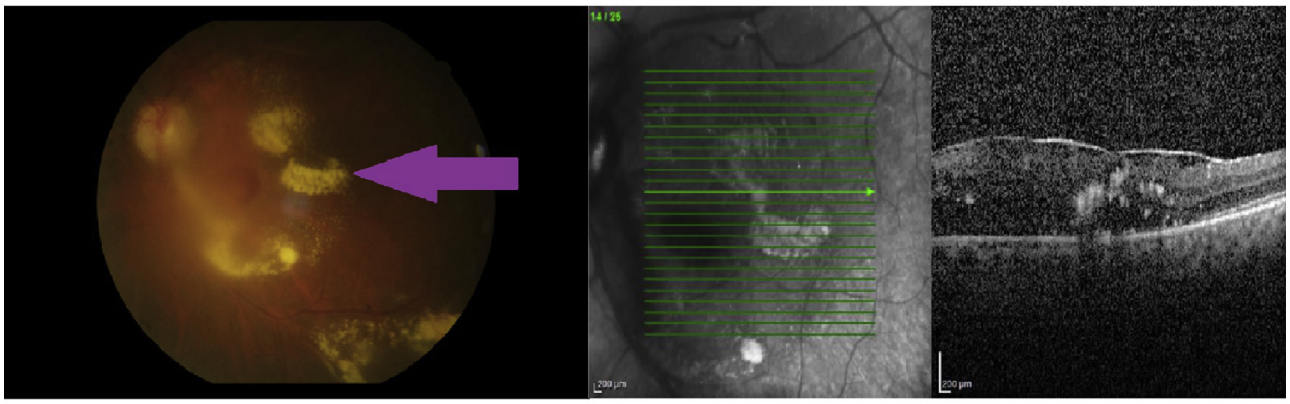
The patient's vision decreased to hand motion at 2 months later in the left eye. Fundus examination showed that extensive and dense exudations and aneurysms at the macula in the left eye (Fig. 2). Intravitreal dexametasone implant was decided to be performed because of sight-threatening of the left eye. After 1 month of intravitreal dexamethasone injection, best-corrected visual acuity increased to hand motion from counting fingers at 2 m with the exudations decreased and disappeared on the left eye (Fig. 2). There was no abnormality in intraocular pressure at the left eye after the intravitreal injection.

3. Discussion

Idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) syndrome is a rare retinal vasculitis characterized by a combination of neuroretinitis, exudation and aneurysms scattered to the posterior pole of the retina. IRVAN syndrome is extremely uncommon and shows characteristics of general vasculitis features such as retinal vasculitis, irregular vessel diameter, perivascular sheath, neovascularization, fibroplasia, hemorrhagic exudation and macular edema [6].

In current study, ischemic regions, macroaneurysms and retinal vasculitis were seen in fundus fluorescein angiography. Patient was included in stage 1–2 according to samuel's definition in this case-

The fundus and oct image of the left eye before the intravitreal dexametasone implant was performed



The fundus and oct image of the left eye after the intravitreal dexametasone implant was performed

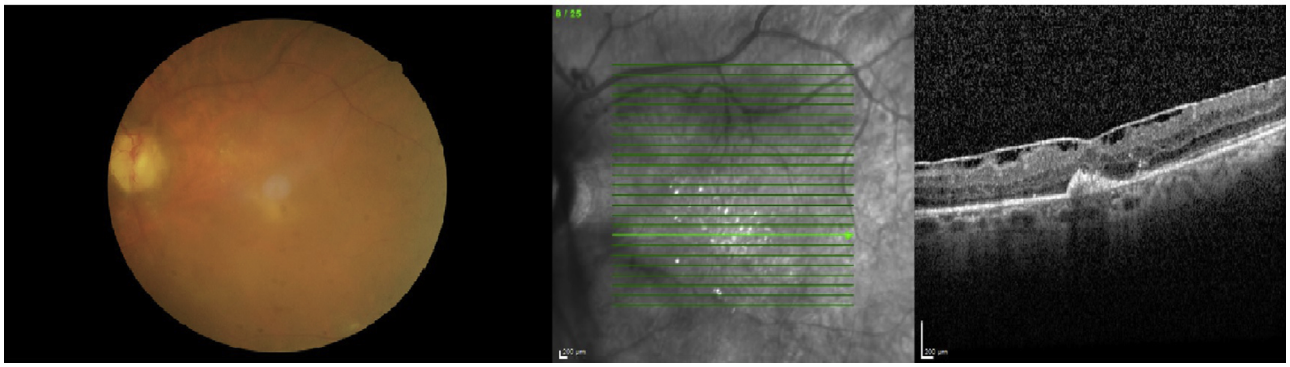


Fig. 2. The fundus and oct image of the left eye before the intravitreal dexametasone implant was applied. The purple arrow shows serious exudations in macula. The fundus and oct image of the left eye after the intravitreal dexametasone implant was performed.

report. Laser photocoagulation and intravitreal injection treatments for ischemia have been reported in the literature [3–5]. Laser photocoagulation, especially in the second phase and after, is one of the most effective treatments to control macroanurysm and ischemia [7,8].

Laser treatment was performed using 200-mm spots of 0.2 s duration with sufficient intensity to cause a 'greyish to white' response that extend just beyond the margins of the lesion on both sides. The argon laser photocoagulation system provides using blue (488 nm wavelength) and green (514 nm wavelength) light emission absorbed by hemoglobin and melanin.

Photocoagulation of the photoreceptors reduces the oxygen consumption of RPE and allows more oxygen to diffuse from the choroid to the inner retina, where the amount of oxygen increases and consequently hypoxia decreases [9].

There were 2 case reports of using intravitreal dexamethasone implant at studies until now. These were the case presentations of Empeslidis and Saatci. Empeslidis et al. reported that an increase in visual acuity and a reduction in macular edema in 4 weeks after performed to intravitreal dexametasone implant [3]. Saatci et al. showed that serous retinal detachment and peripapillary exudates gradually decreased in both eyes after using intravitreal dexametasone implant and hereby vision improved [7].

In the patient of present study, there was an increase in visual acuity after performing of intravitreal dexamethasone implant with the exudations decreased and disappeared.

We did not prefer intravitreal anti- vascular endothelial growth factor(VEGF) agents primarily because in this patient did not have intense ischemia on FFA and laser treatment was applied to the ischemic regions before performing intravitreal dexametasone implant according to Samuel stages [1,2]. The priority in the selection of intravitreal agents was the amount of ischemia and the intensity of exudations at the macula.

Herein, IRVAN syndrome in a case with epiretinal membrane and lamellar hole formation for the first time and showed that combined theraphy of intravitreal dexametasone implant and argon laser photocoagulation are influential at the treatment of severe and extensive exudations. Therefore it can be effective and preferable method at the treatment of severe, calcified and hard exudations in IRVAN syndrome. In this patient, we could not detect an increase in vision due to damage of photoreceptor cells because of subretinal exudation.

In the shed-light of this case, the combination therapy seems to improved the anatomical and functional outcomes in IRVAN syndrome however close follow-up and frequent examinations should be prioritized.

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Informed consent

Informed consent was obtained prior to every surgical procedure from all individual participants included in the study.

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