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Case Report

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Irvan syndrome: A case report and literature review

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Abstract

Purpose: Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis syndrome (IRVAN) is a rare disorder of unknown etiology. It is most commonly characterized by bilateral involvement with arterial bifurcation aneurysms, arterial vasculitis, exudative maculopathy, neuroretinitis, and peripheral retinal ischemia. The aim of this case report and a review of the literature is to highlight the clinical and therapeutic features of this syndrome.

Case report: We report the case of a 28-year-old patient with IRVAN syndrome. Clinical and angiofluographic semiology were characteristic. She was treated by retinal photocoagulation in peripheral non perfusion areas and subsequent intravitreal injections of bevacizumab.

Conclusion: Irvan syndrome is a rare but important condition to be aware of. Retinal photocoagulation is the first-line treatment.

Keywords: Irvan; Fluorescence angiography; Optical coherence tomography; Retinal photocoagulation; Bevacizumab.

Abbreviations: IRVAN: Idiopathic Retinal Vasculitis Aneurysms and Neuroretinitis; VEGF: Vascular Endothelial Growth Factor; OCT: Optical Coherence Tomography; IVT: Intravitreal Injection; NVG: Neovascular glaucoma.

Introduction

Idiopathic, Retinal Vasculitis, Aneurysms, and Neuroretinitis syndrome (IRVAN) is a rare disorder first described in 1995. It is a vasculitis that most often affects young women [1]. Involvement is often bilateral but unilateral case have been reported [2]. The diagnosis is based on major and minor criteria [3]. Prognosis may be severe with rapid vision loss due to retinal ischemia or macular exudation. Retinal photocoagulation is the first-line treatment which can be combined with anti-VEGF injection or adjuvanted corticosteroids [3]. We report a case of IRVAN syndrome treated with retinal photocoagulation and anti-VEGF intravitreal injections.

Case report

A 28-year-old melanoderm women, complained about vision loss with myodesopsias in the right eye over the past year. Her visual acuity was 20/40 in the right eye and 20/20 in the left eye. The anterior segment was normal and the intraocular pressure was 13 mmHg in both eyes. The fundus examination showed in both eyes: tortuosity of the peripapillary vessels, vasculitis with narrowed and sheathed arteries, aneurysms at the arterial bifurcations with nasal and peripapillary and macular exudation (Figure 1). In the right eye, Optical Coherence Tomography (OCT) showed retro-foveolar thickening with hyper-reflective dots in the retinal inner layers and epiretinal membrane. In the

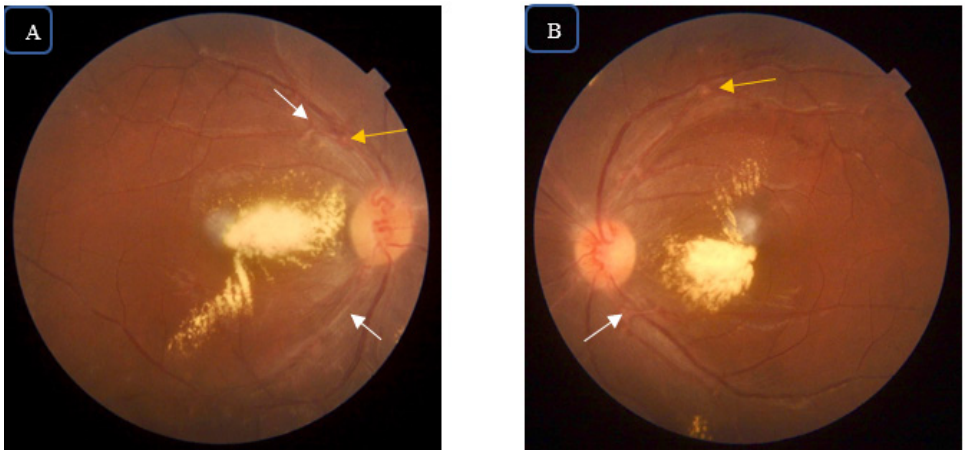

Figure 1: Color fundus picture of the right **(A)** and left **(B)** eyes showing exudations, vasculitis (white arrows), and arterial aneurysms (yellow arrows).

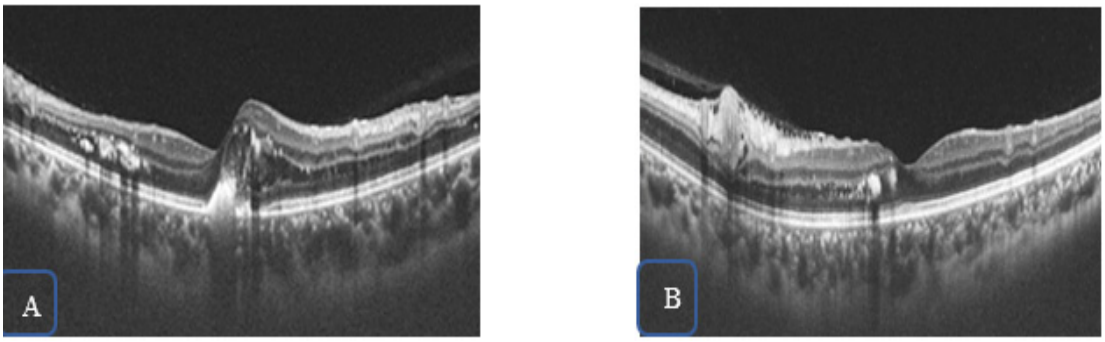

Figure 2: Macular OCT at the right eye **(A)** showed retro-foveolar thickening with hyper-reflective dots in the inner layers, and macular epiretinal membrane with juxta-foveolar hyper-reflective dots at the left eye **(B)**.

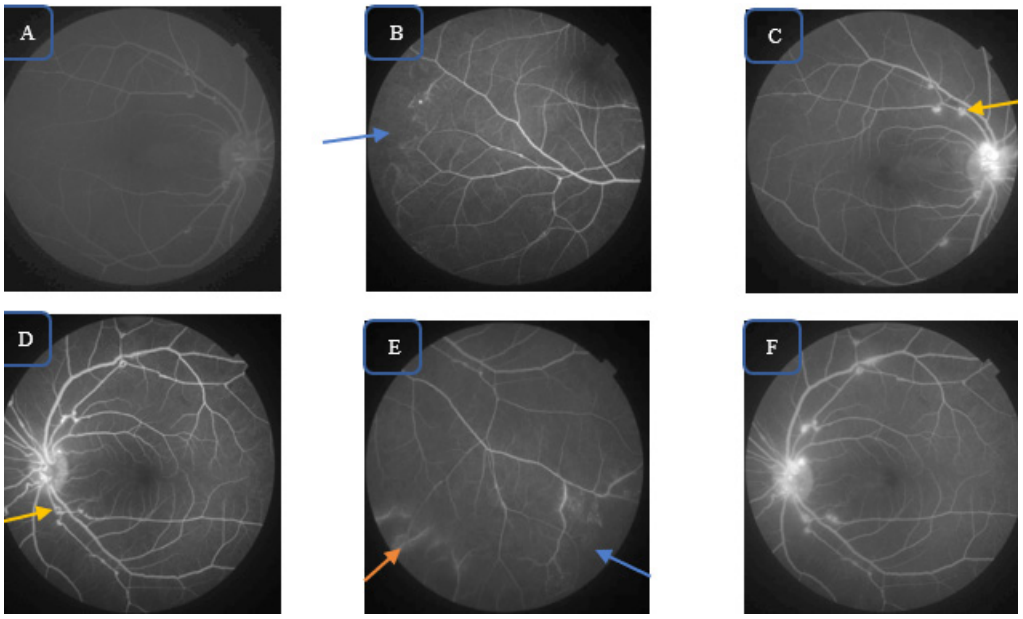

Figure 3: Angiographic images of the right eye **(A,B,C)** and left eye **(D,E,F)** showing macroaneurysms (yellow arrow), temporal peripheral ischemia (blue arrow), vasculitis (orange arrow).

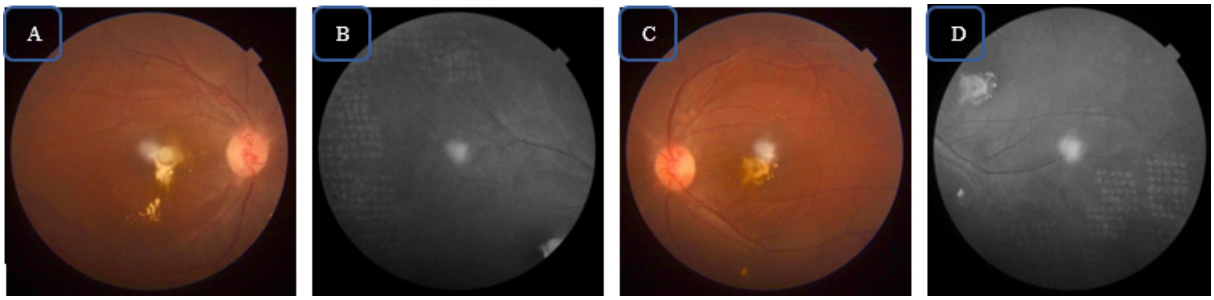

Figure 4: Bevacizumab post-IVT color fundus picture of the right **(A,B)** and left **(C,D)** showing a persistence of arterial aneurysms and a juxta-foveal scar fibrosis.

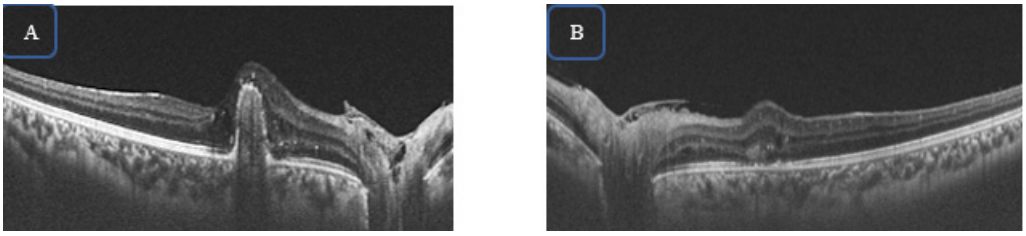

Figure 5: Post-IVT macular OCT of the right eye **(A)** and the left eye **(B)** revealing juxta-foveal dome uplift of the Pigment Epithelium (PE) more marked on the right and somes hyper-reflective dots.

left eye, para-foveolar thickening, epiretinal membrane with juxta-foveolar hyper-reflective dots was noted (Figure 2). Fluorescence angiography showed multiple macroaneurysms at arterial bifurcation sites, capillary non-perfusion areas and subtle papillary and para-macular diffusion at the late stage (Figure 3).

The cardiovascular evaluation was normal. IRVAN syndrome was the most likely diagnosis in this case. A photocoagulation of the ischemia areas was performed on both eyes, and she had three Intravitreal injections (IVT) of bevacizumab for the macula exudation. After one year of follow-up, the visual acuity was 20/40 in the right eye and 20/32 in the left eye. The fundus showed in both eyes a persistence of arterial aneurysms, a regression of retinal exudation and a juxta-foveal scar fibrosis (Figure 4). OCT control showed a juxta-foveal dome uplift of the Pigment Epithelium (PE) more visible on the right eye with a preservation of the systematization of the outer retinal layers (Figure 5).

Discussion

IRVAN syndrome is a condition of the young adult and female predominance has been reported in the majority of series [1,3]. Involvment is most often bilateral [3-5], but unilateral case has been described by Mossavi [2]. The diagnosis is based on criteria including three major (aneurysmal dilations in arterial bifurcations, retinal vasculitis and neuroretinitis) and three minors (retinal ischemia, exudative maculopathy and retinal neovascularization) [1]. In our patient, the clinical and angiofluographic presentation was typical and included all major criteria and 2 minor criteria. Aneurysms located in the arterial branches of the head of the optic nerve or first-order arteries, are the most characteristic signs of this syndrome. However, exudative maculopathy and peripheral retinal ischemia are most described [1,6]. Retinal vasculitis and neuroretinitis were described in all patients in the Chang series [1].

Samuel proposed a prognostic and therapeutic classification in 5 stages [3]. Our patient was classified stage 2. Exudative maculopathy and retinal ischemia, responsible of visual loss, are poor factors prognosis [3]. IRVAN syndrome is an idiopathic pathology, however an inflammatory component has been described in its pathophysiology [7]. An association with p-ANCA vasculitis and anti-phospholipid antibody syndrome was described [8-10]. It is important to differentiate this condition with other pathologies such as macroaneurysms of high blood pressure, Coats' disease and Eales' disease [7]. In adult Coats' disease, saccular dilations are both arterial and venous. Eales' disease, which primarily affects the peripheral retina, is not associated with arterial aneurysms or vascular tortuosity [7]. Photocoagulation of ischemic areas performed in our patient, is the first line treatment admitted in the literature allowing prevention of neovascularization [3]. Contrary to our case, photocoagulation allowed macular edema regression in Mansour's IRVAN syndrome case reported [6]. Anti-VEGF is a therapeutic alternative and is thus indicated in case of neovascular complications [11,12]. Our patient had 3 injections of bevacizumab with a regression of macular edema. Using systemic or intravitreal of corticosteroids is discussed. Saatci [4] reported a case successfully treated with a dexamethasone implant but according to Lemaitre [13] their efficiency would be short.

After 12 months follow-up, our patient presented a bilateral macular scar fibrosis with near-acuity conservation, justified by the integrity of the outer retinal layers. Indeed, this complication was also reported by Nabil in 16.7% [5]. The prognosis for our patient is unpredictable, but according to Samuel early treatment with photocoagulation helps maintain good vision and prevent neovascularization [3].

Conclusion

IRVAN syndrome is a rare idiopathic entity, mainly affecting young women. The search for an associated systemic disease is appropriate. Visual prognosis depends on ischemic involvement and exudative maculopathy. Photocoagulation is the reference treatment, supplemented by anti-VEGF.

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