IRVAN syndrome : Case report

Abstact :

Idiopathic retinal vasculitis, arteriolar macroaneurysms, and neuroretinitis (IRVAN) is a rare condition predominantly affecting young, healthy females without systemic disease. We present the case of a 35-year-old female she presented with a 3-month history of decreased visual acuity without associated symptoms. Ophthalmological examination revealed reduced visual acuity in both eyes, papillary edema, stellate macular edema, and hemorrhages bilaterally. Retinal imaging confirmed IRVAN syndrome . Treatment included panretinal photocoagulation (PRP) and intravitreal bevacizumab injections, resulting in macular edema regression.

Key Words : Vasculitis- Arteriolar macroaneurysms- Neuroretinitis - Anti VEGF- PRP

Introduction :

Idiopathic retinal vasculitis, arteriolar macroaneurysms, and neuroretinitis (IRVAN) is a rare clinical entity typically observed bilaterally in a young, healthy female without any systemic disease.[1]Retinal vessel inflammation can occur in conjunction with different ocular inflammations and systemic vascular diseases, or it may be of unknown origin. Phlebitis is typically more prevalent than arteritis. Arterial involvement is frequent in IRVAN,a condition linked with multiple aneurysmal dilatations of the retinal arterioles and the optic nerve head. Peripheral capillary non-perfusion, retinal neovascularization, and macular exudation are additional characteristics of this condition. Failure to treat it promptly can result in severe vision-threatening complications.[1–3]

Numerous treatement have been used to treat the IRVAN syndrome such as intavitreal injection of anti vascular endothelial grought factor (VEGF)[4,5]and panretinal photocoagulation (PRP) for ischemic areas.

Our case highlights the clinical presentation, diagnostic workup, and management challenges of IRVAN, emphasizing the importance of continued monitoring and adherence to treatment protocols.

Case presentation :

We present the case of a 35-year-old female who was admitted to our department with a three-month history of decreased visual acuity, without any other associated symptoms. Ophthalmological examination revealed a best-corrected visual acuity (BCVA) of 2/10 in the right eye and 3/10 in the left eye, with no abnormalities detected in the anterior segment examination. Fundus examination showed papillary edema with stellate macular edema and scattered hemorrhages in all four quadrants, along with pre-retinal

hemorrhages in both eyes (Fig 1).A comprehensive clinical examination of the other systems reveals no abnormalities. Fluorescein angiography (FFA) revealed aneurysmal dilatations along the arterial pathways in the right eye and peripheral ischemia in both eyes (Fig 2). Macular Optical Coherence Tomography (OCT) demonstrated bilateral macular edema (Fig 3). A comprehensive biological assessment, including serology and immunology, returned normal results. The patient underwent extensive panretinal photocoagulation (Fig 4) in addition to intravitreal injections of Bevacizumab in both eyes. The patient's course was characterized by regression of macular edema (Fig 5) but without improvement in her BCVA.

Discussion :

The IRVAN syndrome (Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis) was first defined in 1983 by Kincaid and Schatz [3]It is a retinal disorder of unknown etiology. The diagnosis of IRVAN is based on three major elements: multiple aneurysmal dilatations, retinal vasculitis, and neuroretinitis at the arterial bifurcation[6–8] .

Samuel et al [2] categorized the progression of the disease into five stages :Stage I : encompasses macroaneurysms, exudation neuroretinitis and retinal vasculitis . Stage 2 is characterized by capillary non-perfusion as evidenced by FFA . Stage 3 manifests as neovascularization in the posterior segment , either at the disc or elsewhere , and /or vitreous hemorrhage . In stage 4, there is the presence of anterior segment neovascularization specifically rubeosis iridis . Stage 5 is marked by neovascular glaucoma . Accordingly , our patient exhibit featues consistent with stage 2 of the desease

PRP stands as the singularly recognized treatment method in cases of peripheral ischemia or neovascularization, and its early implementation is crucial to ward off complications arising from ischemia. PRP commonly demonstrates effectiveness, particularly in stages 2 and 3 of the disease [9] . Rouvas et al [10], recommended delaying panretinal photocoagulation (PRP) when peripheral ischemia affects fewer than two quadrants of the retina.

Numerous treatments have been used for IRVAN syndrome, with varying degrees of efficacy. Intravitreal injections of anti-VEGF agents, bevacizumab and ranibizumab, have produced favorable results.[5,10,11]

Recently, Cheema et al[12] proposed that infliximab therapy could be beneficial in mitigating inflammation and leakage from the optic nerve, based on their observation in two cases of treatment-resistant IRVAN syndrome.

Sawhney et al. [13]utilized PRP in the regions of retinal ischemia and aneurysmal dilatations following three bevacizumab injections for a patient with stage 3 IRVAN syndrome. Over the subsequent 8 months, the patient received monthly treatment comprising seven additional bevacizumab injections, one intravitreal dexamethasone implant, and one periocular triamcinolone injection. Later, a pars plana vitrectomy was

performed to remove the epiretinal membrane and alleviate vitreomacular traction. The macular lipid exudation had completely resolved with a residual lamellar hole.

Eale's disease could be a differential diagnosis of IRVAN syndrome due to the presence of retinal vasculitis and peripheral nonperfusion features. Furthermore, Eale's disease is more likely to be found in the retinal veins instead of arterioles. Also, multiple aneurysms and optic nerve head vascular tortuosity distinguish IRVAN syndrome from Eale's disease [14]

Conclusion :

The IRVAN syndrome is thus a highly characteristic clinical picture due to its angiographic features, to be considered in any posterior uveitis that combines neuroretinitis with arterial vasculitis, especially in the case of a young woman. It is crucial to investigate any underlying inflammatory or vascular pathology to confirm the idiopathic nature of this condition.

Once the diagnosis is established, it is crucial to treat retinal periphery ischemia and to conduct regular follow-ups to ensure the absence of evolving complications that may arise during its course.

Images :

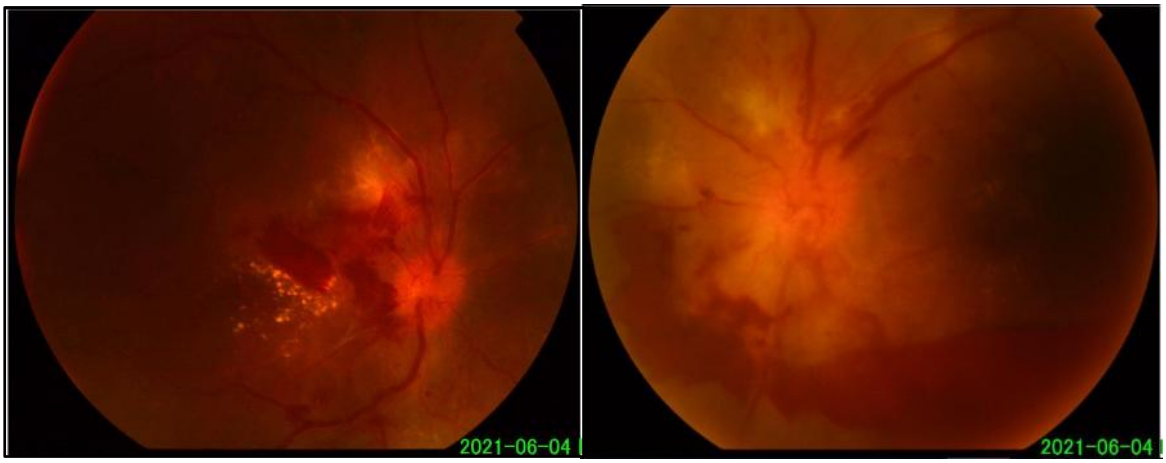
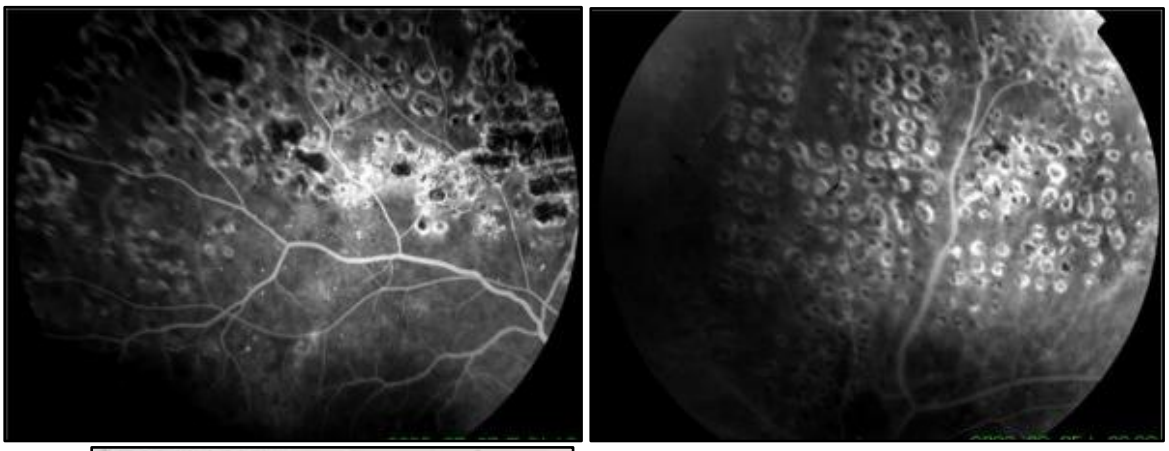
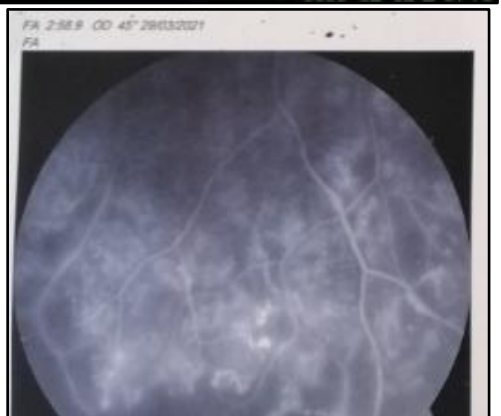


Figure 1**:** Color fundusphotograph of the rightand the lefteyeshowingneuroretinitiswithpre-retinalhemmoragias in botheyes

Figure 2 :
Fluoresceineangiographyshowinganeurysmaldialtationswithperipheralischemiain the right eye





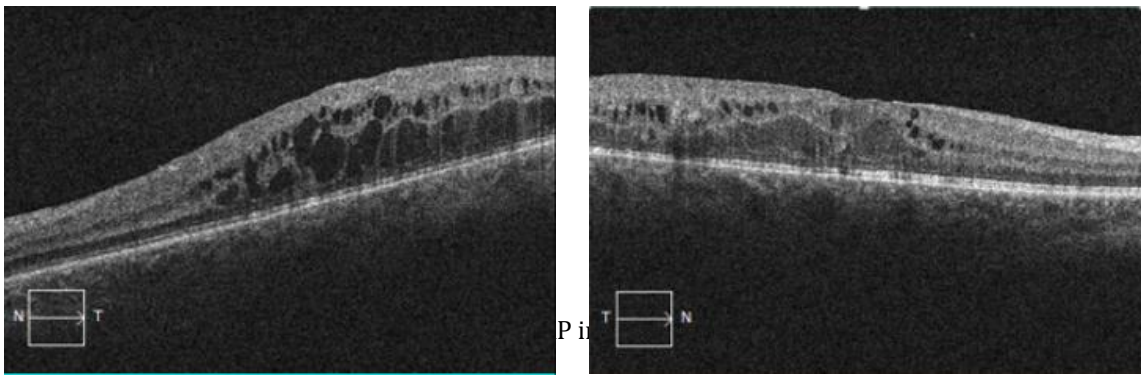


Figure 4 : The macular Optical CoherenceTomography (OCT) showeddiffusedretinalthikeningright eye and cystoidemacularedema of the lefteye

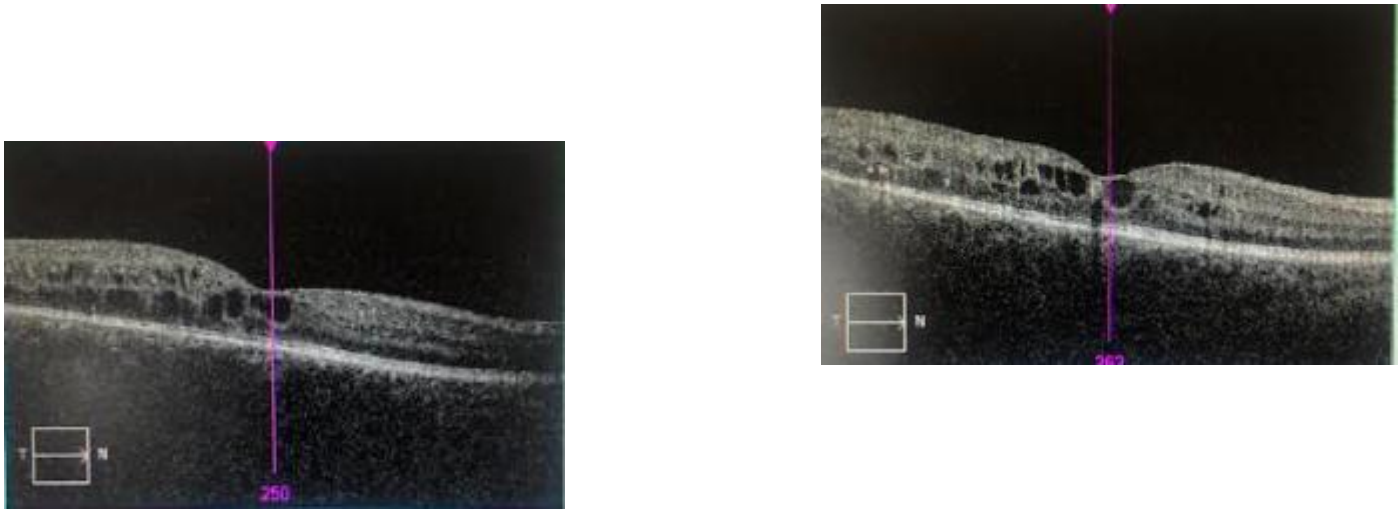


Figure 5 :Themacular Optical CoherenceTomography (OCT) post one cycle of intravitreal injections of Bevacizumab

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