Case report

IRVAN syndrome : Case report

Abstact :

Idiopathicretinalvasculitis, arteriolarmacroaneurysms, and neuroretinitis (IRVAN) is a rare condition predominantlyaffectingyoung, healthyfemaleswithoutsystemicdisease. Wepresent the case of a 35-year-old femaleshepresentedwith a 3-month history of decreasedvisualacuitywithoutassociatedsymptoms. Ophthalmologicalexaminationrevealedreducedvisualacuity in botheyes, papillaryedema, stellatemacularedema, and hemorrhagesbilaterally. Retinalimagingconfirmed IRVAN syndrome .Treatmentincludedpanretinalphotocoagulation and intravitrealbevacizumab injections, resulting in macularedemaregression.

Introduction :

Idiopathicretinalvasculitis, arteriolarmacroaneurysms, and neuroretinitis (IRVAN) is a rare clinicalentitytypicallyobservedbilaterally in a young, healthyfemalewithoutanysystemicdisease.[1]

Numeroustreatement have been used to treat the IRVAN syndrome such as intavitreal injection of anti vascularendothelialgrought factor (VEGF) and panretinalphotocoagulation (PRP)

Our case highlights the clinicalpresentation, diagnostic workup, and management challenges of IRVAN, emphasizing the importance of continued monitoring and adherence to treatmentprotocols.

Case presentation :

We report the case of a 35- year-oldfemalwasadmitedto ourdepartemnetwith a 3 monthhistory of decreasedvisualacuitywithoutotherassociatedsymptoms.

Ophtphtlmologicalexaminationrevealedbest correctedvisualacuity (BVCA) of 2 / 10 in the right eye and 3/10 in the lefteye ,theanterior segment examinationshowed no abnormalities

Fundus examinationrevealedpapillaryedemawithstellatemacularedema and spottedhemorrhagies in all four quadrantswithpre-retinalhemmoragias in botheyes(Fig 1)

fluorosceinangiography (FFA)wasperformeddemostratinganeurysmal dilatations along the arterielpathwaysin the right eyeand peripheric ischemiain botheyes(Fig 2)

The macular Optical CoherenceTomography (OCT) showedbilateralmacularedema(Fig 3).

A comprehensivebiologicalassessementincludingserology and immunolgyreturned normal

A heavyPanretinalphotocoagulationwasperformed (Fig 4)in conjunctionwithintavitreal injections of Bevacizumabin botheyes

the patient cours wasmarked by regression of macularedemabut withoutimprovementof her BVCA .

Discussion :

The IRVAN syndrome (IdiopathicRetinalVasculitis, Aneurysms, and Neuroretinitis) was first definedin 1983 by Kincaid and Schatz[2]It is a retinaldisorder of unknownetiology. The diagnosis of IRVAN isbased on three major elements: multiple aneurysmal dilatations, retinalvasculitis, and neuroretinitis at the arterial bifurcation[3–5] .

Samuel et al [6]categorized the progression of the diseaseinto five stages :Stage I :encompassesmacroaneurysms, exudationneuroretinitis and retinalvasculitis . Stage 2 ischaracterized by capillary non-perfusion as evidenced by AGF . Stage 3 manifests as neovascularization in the posteriorsegment ,either at the disc or elsewhere , and /or vitreoushemorrhage . In stage 4, thereis the presence of anterior segment neovascularizationspecificallyrubeosisiridis . Stage 5 ismarked by neovascularglaucoma .Accordingly ,our patient exhibitfeatues consistent with stage 2 of the desease

PRP stands as the singularlyrecognizedtreatmentmethod in cases of peripheralischemia or neovascularization, and itsearlyimplementationis crucial to ward off complications arisingfromischemia. PRP commonlydemonstrateseffectiveness, particularly in stages 2 and 3 of the disease.[7]

Numeroustreatments have been used for IRVAN syndrome, withvaryingdegrees of efficacy. Intravitreal injections of anti-VEGF agents, bevacizumab and ranibizumab, have produced favorable results.[8–10]

Recently, Cheema et al[11]proposedthat infliximab therapycouldbebeneficial in mitigating inflammation and leakagefrom the optic nerve, based on their observation in two cases of treatment-resistant IRVAN syndrome.

Sawhney et al. [12]utilized PRP in the regions of retinalischemia and aneurysmal dilatations followingthreebevacizumab injections for a patient with stage 3 IRVAN syndrome. Over the subsequent 8 months, the patient receivedmonthlytreatmentcomprisingsevenadditionalbevacizumab injections, one intravitrealdexamethasone implant, and one periocular triamcinolone injection. Later, a pars plana vitrectomywasperformed to remove the epiretinal membrane and

alleviatevitreomacular traction. The macularlipidexudationhadcompletelyresolvedwith a residuallamellarhole.

Eale'sdiseasecouldbe a differentialdiagnosis of IRVAN syndrome due to the presence of retinalvasculitis and peripheralnonperfusionfeatures. Furthermore, Eale'sdiseaseis more likely to befound in the retinalveinsinstead of arterioles. Also, multiple aneurysms and optic nerve headvasculartortuositydistinguish IRVAN syndrome fromEale'sdisease[13]

Conclusion :

The IRVAN syndrome isthus a highlycharacteristicclinicalpicture due to itsangiographicfeatures, to beconsidered in anyposterioruveitisthat combines neuroretinitiswitharterialvasculitis, especially in the case of a youngwoman. It is crucial to investigateanyunderlyinginflammatory or vascularpathology to confirm the idiopathic nature of this condition.

Once the diagnosisisestablished, itis crucial to treatretinalperipheryischemia and to conductregular follow-ups to ensure the absence of evolving complications thatmay arise duringits course.

Images :

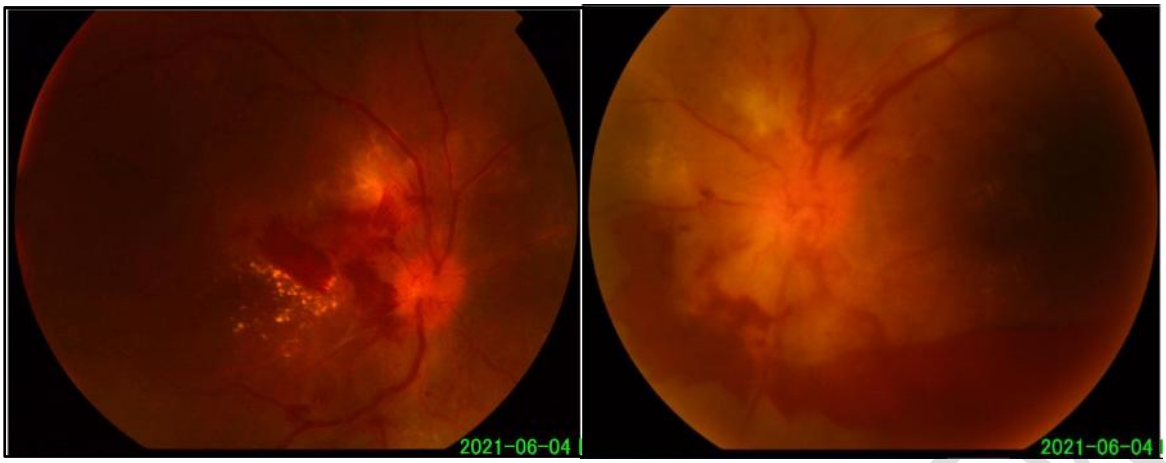
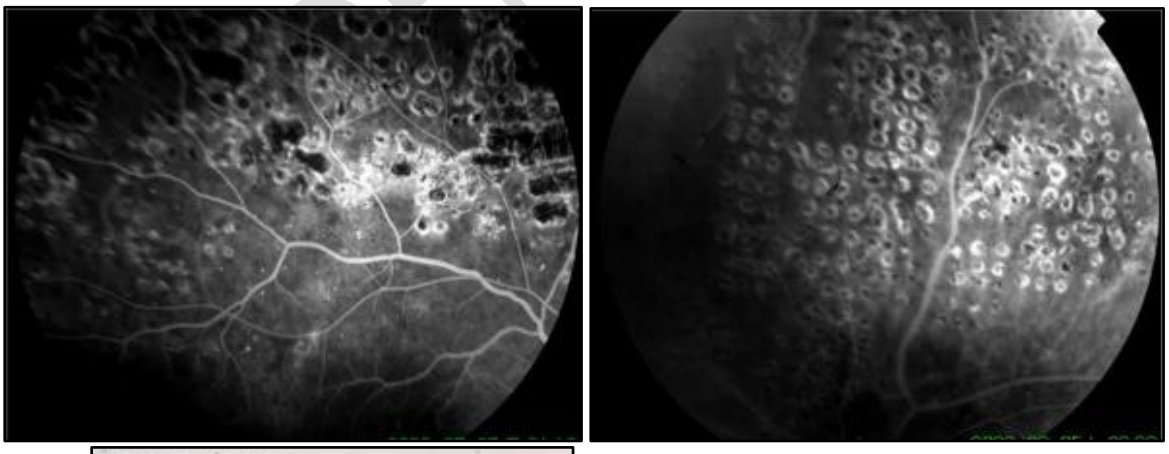
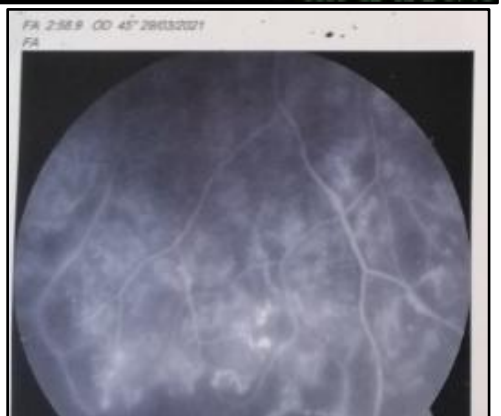


Figure 1**:**Color fundusphotograph of the rightand the lefteyeshowingneuroretinitiswithpre-retinalhemmoragias in botheyes

Figure 2 :
Fluoresceineangiographyshowingan eurysmaldialtationswithperipheralis chemia in the right eye





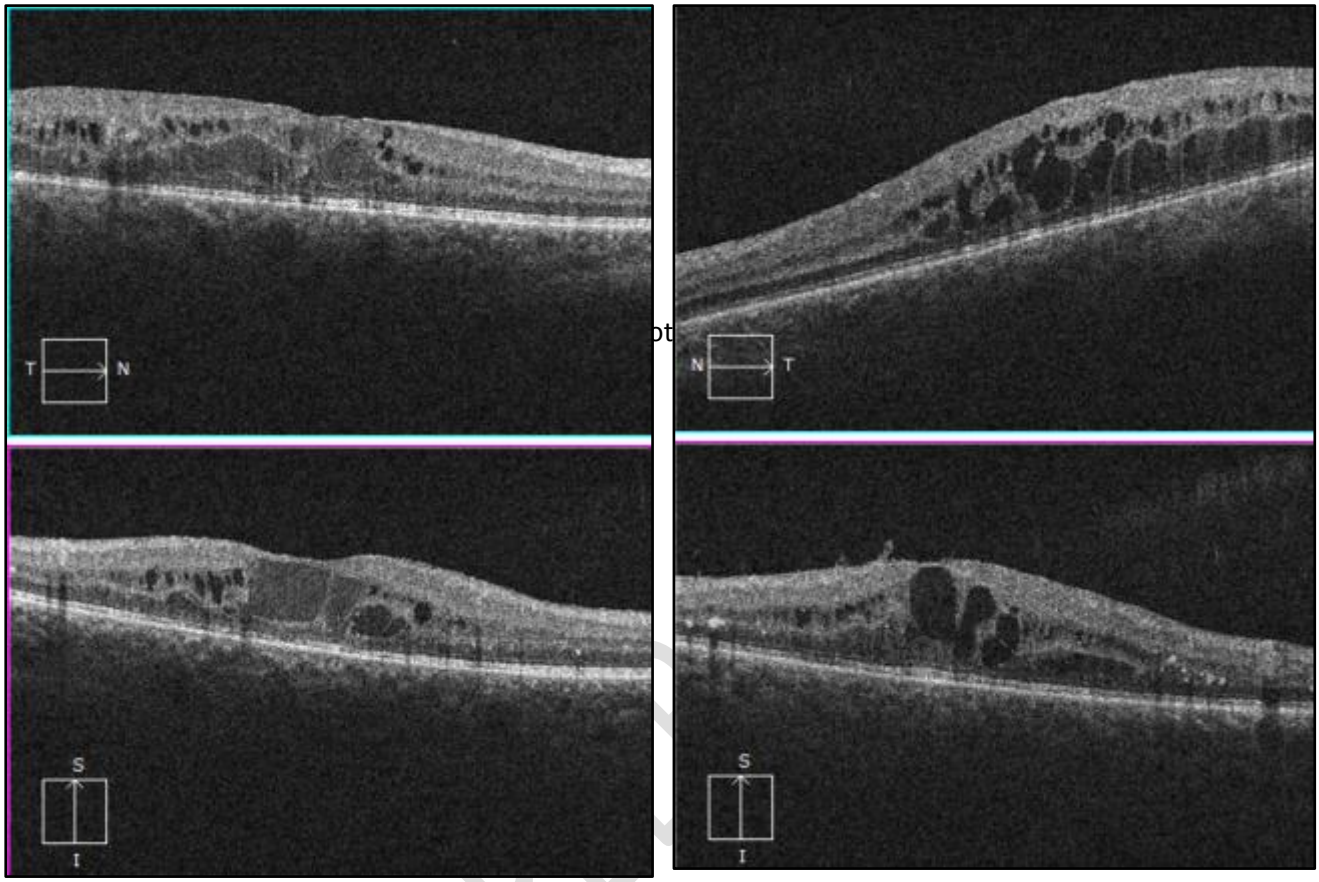


Figure 4 : The macular Optical CoherenceTomography (OCT) showedbilateralmacularedema

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