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Case Report

IRVAN syndrome: A case report and a literature review

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ABSTRACT

Introduction and Importance: Idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) syndrome is a rare retinal defect. It has a female predominance. The staging of IRVAN was defined depending on ocular criteria.
Case presentations: A 30-year-old female patient presented with floaters in the left eye. One year later, it was diagnosed with Fluorescein fundus angiography (FFA) at stage III of IRVAN syndrome. After one year, the right eye was affected, diagnosed at stage II, and successfully treated with heavy Pan-Retinal Photocoagulation (PRP).
Clinical discussion: IRVAN Syndrome is an extremely rare retinal defect which has female predominance. The etiology of this syndrome still idiopathic. The most encountered symptoms are blurred vision and vision loss. FFA is the best diagnostic investigation to reveal the retinal abnormalities. Many suggested protocols were mentioned to treat IRVAN Syndrome. Our experience suggests IRVAN Syndrome a differential diagnosis for patients with floaters, and assures that PRP is an affective curement for late stages of IRVAN Syndrome.
Conclusions: Heavy PRP was used successfully for stage II and III. The case emphasizes the importance of early diagnosis to assert the complications. Furthermore, IRVAN syndrome should be considered as a differential diagnosis in patients with floaters. Therefore, heavy PRP is highly recommended as a suitable treatment for IRVAN syndrome.

1. Introduction and importance

IRVAN syndrome is a rare retinal defect. IRVAN syndrome is usually seen in young females. The Etiology of IRVAN syndrome is still idiopathic [1]. The clinical presentations of IRVAN syndrome may vary widely. It may include non-symptomatic [2], painless blurred vision [3], exotropia, and a central scotoma [4]. To diagnose IRVAN syndrome, six criteria must be fulfilled, three significant criteria, and three minor criteria [1]. Samuel et al. have defined the staging of IRVAN syndrome depending on ocular criteria (Table .1) [1,5]. The most reliable diagnostic test is FFA. The main complications sum in retinopathy, retinal neovascularization, and peripheral capillary nonperfusion [6]. There are several modalities to treat those patients, such as panretinal laser photocoagulation, corticosteroids, and vitrectomy [7].

2. Case presentation

A 30-year-old female was admitted to the Department of Ophthalmology complaining of a floater in the left eye (LE). No medical and family history were recorded. On physical examination, her visual acuity was 20/20 in both eyes. Intraocular pressure measurement using the Goldmann Applanation tonometer was 8 mm Hg in both eyes. The right eye (RE) examination was within the normal limit. LE examination showed: anterior chamber +1 cells, fundus examination revealed retinal vasculitis and optic nerve hyperemia, FFA showed a wide peripheral capillary drop out and leaking from optic disc vessels and peripheral ischemia, LE macular optical coherence tomography (OCT) was within normal limits. Initially, the patient was diagnosed with posterior uveitis and retinal vasculitis, so she was treated with oral Prednisolone and mycophenolate mofetil (500 mg once a day) for 6 months with no response. One year later, the RE was still stable, whereas, visual acuity in the LE deteriorated dramatically and the patient developed mild

Abbreviations: IRVAN:, Idiopathic Retinal Vasculitis, Aneurysms, And Neuroretinitis; FFA:, Fluorescein Fundus Angiography; OCT:, Optical Coherence Tomography; PRP:, Pan-Retinal Photocoagulation; RPE:, Retinal Pigment Epithelium; FVP:, Fibrovascular Proliferation; RD:, Retinal detachment; ANA:, Antinuclear Antibodies; HLA-B51:, Human Leukocyte Antigen B51; LE:, Left Eye; RE:, Right Eye; AFS:, Allergic Fungal Sinusitis.

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Table 1
Describes the stages of idiopathic retinitis, vasculitis, aneurysms, and neuroretinitis (IRVAN) syndrome depending on the ocular findings.

Stage	Ocular Findings
1	Macroaneurysms, exudation, neuroretinitis, retinal vasculitis.
2	Capillary nonperfusion (angiographic evidence)
3	Posterior segment neovascularization of disc or and/or vitreous hemorrhage.
4	Anterior segment neovascularization (rubeosis iridis)
5	Neovascular glaucoma

vitreous hemorrhage. The LE FFA revealed wide and intensive leaking from the optic disc and hard exudates in a circular manner around the optic disc Fig. 1 (A). At that time, macroaneurysms became more obvious Fig. 2 (A, B) and OCT revealed macular edema. This confirmed the diagnosis of IRVAN syndrome. The patient was treated with infliximab, limited PRP, Bevacizumab, and repeated eylea intravitreal injection for 6 months with no response. Because of long-standing macular edema and retinal pigment epithelium (RPE) detachment in the LE, the patient developed a later RPE rip which was revealed in the OCT findings. Later she developed a macular scar Fig. 3 (A, B). One year later, the patient developed leaking in the RE in the inferior temporal retinal branch artery, and OCT showed edema in the macula Fig. 3 (E, F), that time the patient was treated with heavy PRP in both eyes Figs. (1. B, 4). OCT for the RE was repeated and the edema was significantly decreased Fig. 3 (C, D). All the retinal leaks are resolved. Within an 8-year follow-up, the patient was stable and visual acuity was 20/20 in the RE and counting fingers 1 m in the LE. Intraocular pressure measurement is still stable at 8 mm Hg in both eyes.

3. Discussion

IRVAN syndrome is an extremely rare retinal disease that was first characterized in 1983 by Kincaid and Schatz [1,8]. The epidemiology of the syndrome assured that the majority of patients were young females [1,5]. Our literature review included publications about cases of 10 females and 6 males, and the average age at the time of diagnosis was 28.8 years (range, 9–60 years) [2–4,9–17]. Whereas, Samuel et al. study determined the average age of diagnosis at 31.5 years (range, 9–60 years) in their study which involved 22 patients. Also, Samuel et al. reported that 3 patients were of Latin American descent, 2 were of Middle Eastern descent, 13 patients were Caucasian, 1 was East Indian, and 2 were African American [7]. According to our review, only 4 cases reported the origin of the patient, 1 was South Asian, 1 was Somalian, 1 was Afro-Caribbean and 1 was Indian [3,17]. The previous publications etiology sections generally described IRVAN syndrome as an idiopathic retinal disease that consists of neuroretinitis, exudates, and aneurysms [1,8]. According to a study, there was no association between IRVAN and any systemic disorder [6]. The majority of reviewed cases had a clear medical history and negative serum tests [2–4,10–15,17], in comparison to positive human leukocyte antigen B51 (HLA-B51) and antinuclear antibodies (ANA) at 1/50 titer in one case [9]. AM Abu El-Asrar et al. reported a case of association between Allergic fungal sinusitis (AFS) and IRVAN syndrome. Considering the rarity of both diseases, this association does not seem a coincidence. This may lead to suggesting that an immunologically mediated hypersensitivity reaction to the fungal antigens caused the retinal vasculitis and was involved in the development of IRVAN in a predisposed patient [18]. Samuel et al. determined the staging of this syndrome according to ocular findings (Table 1) [7]. Our patient was diagnosed at stage-3 and stage-2 for the LE and RE, respectively. Several symptoms were recorded in the literature such as progressive painless diminution of vision [10], bilateral blurred vision [12], sudden vision loss [13], exotropia, and central scotoma [4]. However, our patient was presented with unilateral floaters which were not reported before. We found in our review that the most common symptoms were blurred vision [3,12,16] and vision loss [13]. There are six criteria to diagnose IRVAN syndrome, three major criteria (numerous aneurysmal dilatations, neuroretinitis, and retinal vasculitis in arterial bifurcation), and three minor criteria (retinal neovascularization, macular exudation, and peripheral capillary perfusion defect) [1]. The most common features of IRVAN syndrome are exudative retinopathy and peripheral capillary nonperfusion. Also, these two features are considered the most visual threat. Another complication is neovascularization of the retina and anterior segment which causes destructive effects on visual acuity. As a diagnostic investigation, FFA clearly reveals pathological retinal changes. Moreover, FFA preceded conventional fundus photography in detecting retinal aneurysmal changes. Eale's disease could be a differential diagnosis of IRVAN syndrome due to the presence of retinal vasculitis and peripheral

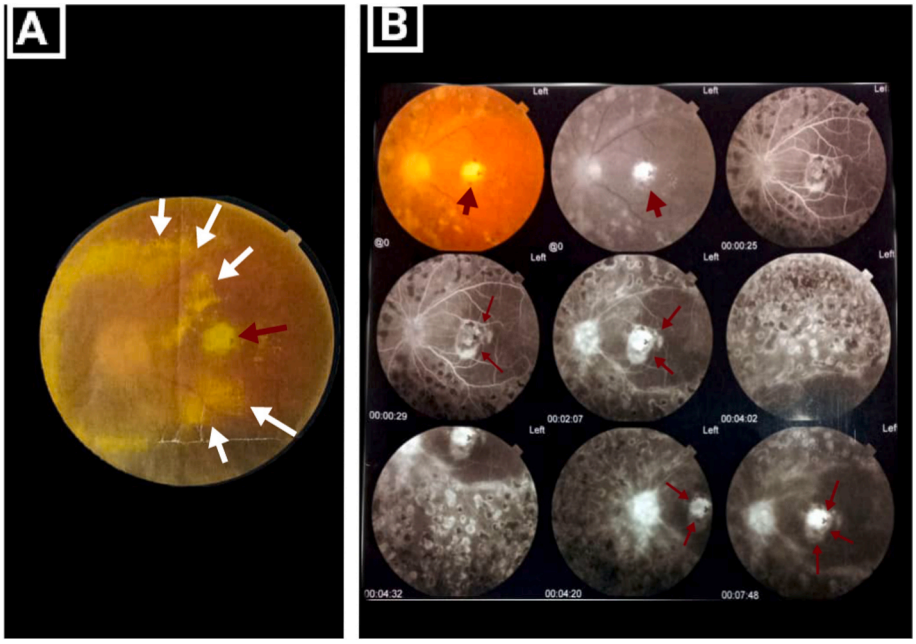

Fig. 1. (A, B): (A) Left eye FFA before heavy PRP, leaking in the optic disc (White Arrow), macula scar (Red Arrow). (B) Left eye FFA after heavy PRP, leaks totally decreased, macula scar (Red Arrow).

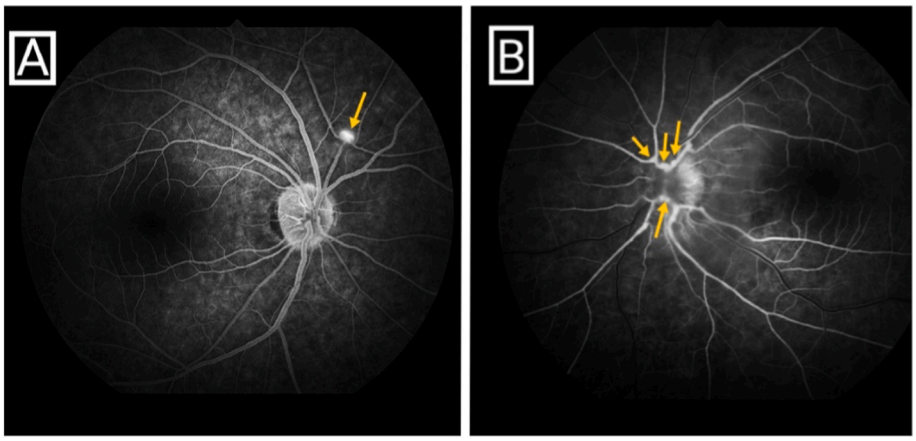

Fig. 2. (A, B): (A) Left eye FFA, Macroaneurysms (Yellow Arrow). (B) Right eye FFA, Macroaneurysms (Yellow Arrow).

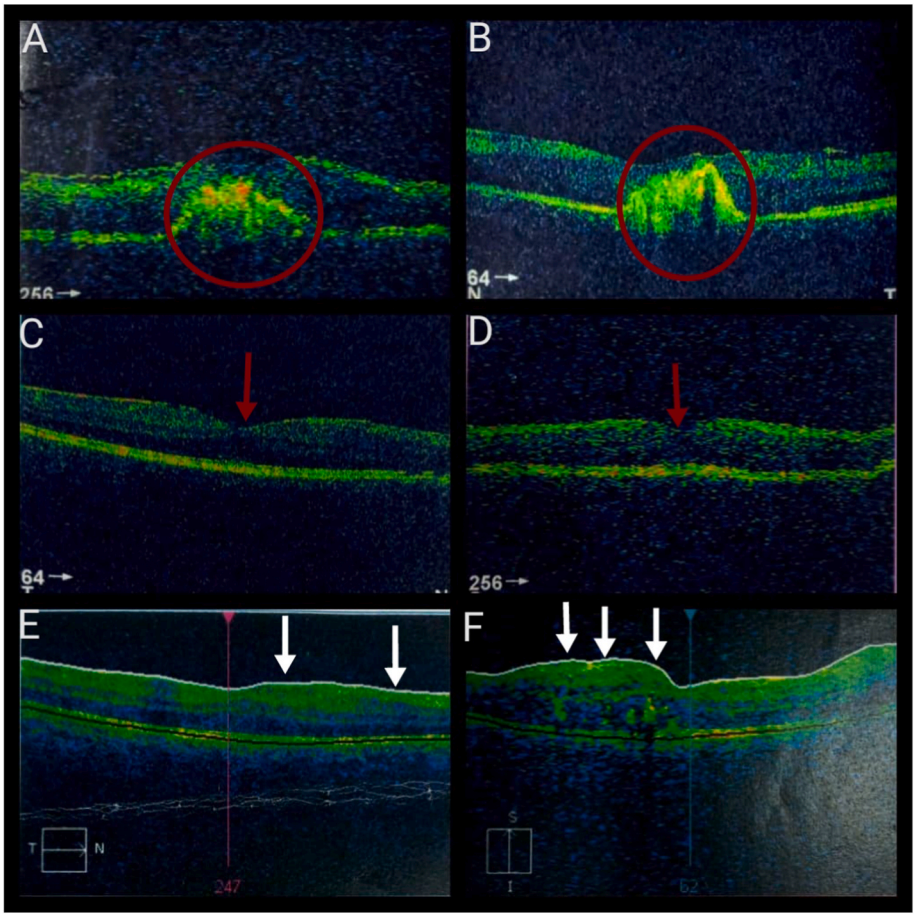

Fig. 3. (A, B, C, D, E, F): (A, B) Left eye, Macula OCT: Macular Scar (Red Circle). (C, D): Right eye after treating with heavy PRP, Macular OCT: Edema totally decreased (Red Arrow). (E, F): Right eye before treatment, Macula OCT: macular edema (White Arrow).

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 [6]. The prognosis of IRVAN syndrome depends on early initiation of treatment and visual impairment seems to be associated with disease progression [7]. Many medical protocols were reported to treat IRVAN syndrome. For stage one, a study recommended a protocol consisting of high-dose prednisone, steroid-sparing drugs (ex.: mycophenolate mofetil), photocoagulation, or local steroid injections. The authors assured that immunosuppression (ex.: Infliximab) is more likely to be effective in the early stages [2]. The most effective treatment for stage II and subsequent stages is PRP [7,19]. In other words, PRP may resolve retinal aneurysms. A study mentioned initiating with PRP at stage III may not stop the neovascular sequelae of retinal ischemia and cause a high rate of severe visual loss. Also, there is a recommendation to treat with PRP when angiographic evidence of widespread retinal nonperfusion is present, and before or shortly after the neovascular development. Moreover, Samuel et al. mentioned neovascular glaucoma as a contraindication to treat with PRP. In our case, LE was stage III and PRP succeeded in stopping the progression of retinal abnormalities without

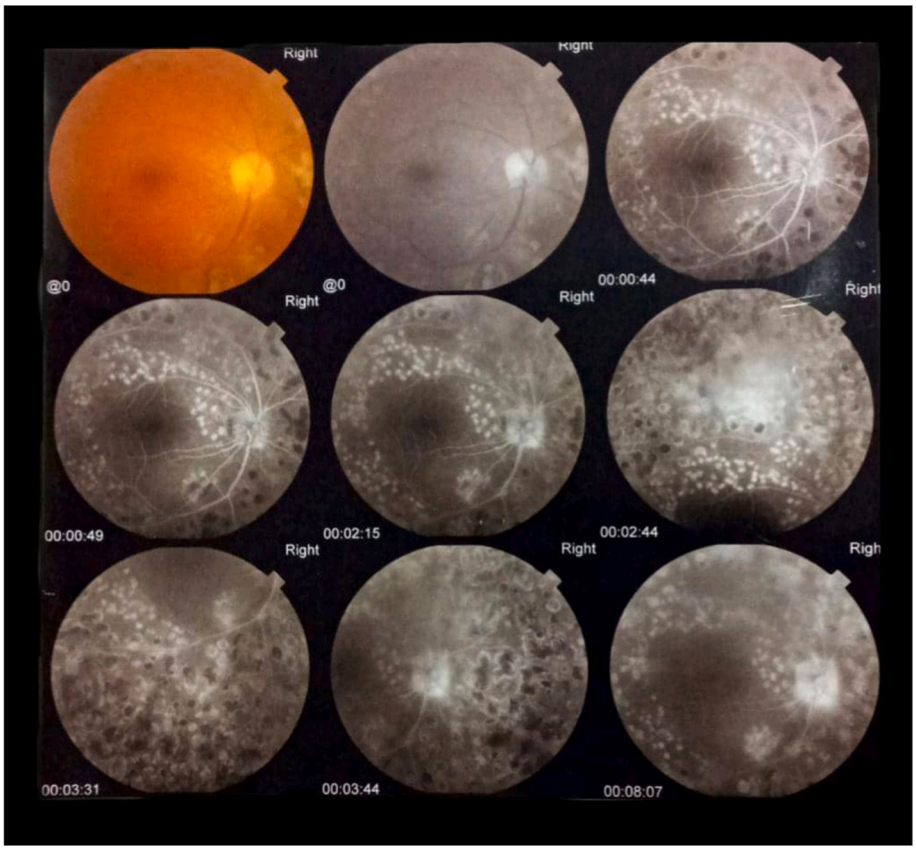

Fig. 4. Right eye FFA after heavy PRP.

any side effects. Also (stage III LE, stage II RE), we found heavy PRP as one of the most potentially curative treatments after a non-beneficial steroids treatment, immunosuppression, Bevacizumab, and Eylea intravitreal injections. Furthermore, Samuel et al. assured that the role of corticosteroids is still controversial [7]. Some studies mentioned that intravitreal dexamethasone implant had improved the visual acuities and decreased the macular edema in four weeks [20]. According to our review, five studies choose PRP as the first choice of treatment [3,4,10,13,15], one of them combined PRP with vitrectomy [10]. Three studies started their medical protocol with steroids [3,13,17]. Whereas there are five studies that performed a combined treatment of PRP and oral steroids as the first followed protocol [3,4,9,12,16], three of them involved other procedures in their protocol such as mycophenolate mophetil, intravitreal Bevacizumab, and Ranibizumab injections [2,12,16]. A study mentioned performing pars plana vitrectomy to treat their patient resulted in partly absorbed exudate and no observed neovascularization [13]. One last study reported serial intravitreal aflibercept injections as a successful curement [14]. Some of the reviewed articles advised PRP to prevent neovascularization and resolve retinal aneurysms and leaks [4,10,13]. The latest established article mentioned a first-time performed treatment which included Fluocinolone acetonide intravitreal implant. The authors assured that it was effective and they strongly recommended local long-standing steroids to control the progression of the retinal disorder [9]. PRP was very effective and stopped the progression of retinal ischemia and exudation sequelae in our case. Owing to the lack of clinical signs at first, the diagnosis was delayed. As a result, the treatment was less effective to improve visual acuity in the LE. Whereas, the RE significantly benefited from the treatment because of early treatment with PRP. On following-up, some cases which first treated with PRP needed additional PRP courses [3,13] or intravitreal Dexamethasone implant [15]. Whereas, one case was worsened with the combined treatment (steroids, PRP) and got better with intravitreal Fluocinolone acetonide injections [9]. Moreover, serial intravitreal aflibercept injections and pars plana vitrectomy were a sufficient curement each separately [13,14]. In our case, no subsequent procedures were needed after applying heavy PRP and visual acuity remained stable without any additional deterioration. This study strongly recommends using PRP to treat IRVAN syndrome, especially in the early stages and assures that early diagnosis may have a significant effect on the treatment outcomes and decrease the complications. Our review also suggests that IRVAN syndrome may have genetic etiologies and may be associated with HLA-B51 and ANA positivity (Table .2). Of note, some authors proposed adding vitreoretinal Fibrovascular Proliferation (FVP) and Retinal detachment (RD) to the staging system in their cohort study [11]. IRVAN progression might vary substantially despite different therapeutic approaches, therefore, several studies suggest treating patients on individual approach [3]. In the early stages of IRVAN, few cases showed previously unreported reversibility of arterial aneurysms with steroid treatment alone [17]. According to a meta-analysis study, the number and site of retinal aneurysms in IRVAN are related to the size of the retinal non-perfusion region and the type of neovascularization [21].

4. Conclusions

IRVAN syndrome is an extremely rare disease and has female predominance. IRVAN affects the retina causing ischemia with unknown etiologies. The correlation between autoimmune disease and IRVAN syndrome is still unknown. In this case, heavy PRP was used successfully, thus, it may represent an efficient treatment for stage II and III. This case emphasizes that early diagnosis has a considerable role in preventing major complications. Also to advise considering IRVAN syndrome as a differential diagnosis in patients with floaters. Further investigations should be conducted.

Table 2
Describes the therapeutic options to manage idiopathic retinitis, vasculitis, aneurysms, and neuroretinitis (IRVAN) Syndrome.

Paper NO.	First Author	Patient/s Age (Ys)/ sex	Chief complaint	Clinical Examination	FFA	Other Diagnostic tests	Treatment	Follow-Up
1	Lucía Moreno-Castro	12/F	N/A	BCVA: 20/20 in RE. 20/40 in LE. Intraocular pressure: 12 mm Hg in both eyes. Vitritis, vasculitis, papillitis, macular exudates. Hx: Bilateral neuroretinitis, vasculitis.	Peripheral ischemia, macroaneurysms	HLA-B51 + ANA 1/50 titer	Corticosteroids, PRP, Periocular Triamcinolone, Bevacizumab. Dexamethasone implant. Fluocinolone acetonide intravitreal implant. Localized perifoveal. Focal laser	At 2 y, BCVA: 20/30 and 20/60 in RE and LE -Intraocular pressure: 16 mm Hg in both eyes -LE Macular retinal thickening decreased after fluocinolone acetonide intravitreal implant. -Evolution of central macular thickness and VA 3 y before fluocinolone and 2 y after implant.
2	Xin Lin	39/M	Diminution of vision in RE, Sudden vitreous hemorrhage	Visual acuity: hand motion in RE, 20/20 in LE. RE reached 20/40, corrected to 20/20. Reached 20/100, corrected to 20/20 when discharged. RE: stage III, LE: stage II progressed to stage III after 1 month. Hx: superior temporal aneurysms	Multiple aneurysmal dilatations, peripheral retinal nonperfusion areas	N/A	Vitrectomy, retinal laser photocoagulation	At 18 months, Aneurysm disappeared. -LE: high signal of the superficial glomerular vascular membrane in OCTA decreased x3. Macular aneurysm: superficial OCTA -RE: optic disc membrane hyperplasia, old laser scars.
3	Meriem Ouederni	40/F	Bilateral blurred vision	Visual acuity: 8/10 in both eyes. 1+ vitreous cells in both eyes. Bilateral papillary loops with disc neovascularization RE, bilateral segmentary arteritis, periphlebitis, peripheral ghost vessels.	Bilateral staining of optic disc, massive dye leakage in RE, vascular sheathing, and large areas of capillary drop out in both eyes. Punctuate hyperfluorescence of tempo-superior artery in LE corresponds to an aneurysm.	Papillary aneurysm, neovascularization of optic nerve head in RE, papillary aneurysm in LE. Extensive areas of CNP in both eyes.	PRP in both eyes + Bevacizumab in RE. corticosteroids	Visual acuity: 8/10 No neovascularization recurrence at 10-month.
4	Yun Zhang	Pt1: 9/M Pt2: 21/F Pt3: 44/F	C1: Bilateral vision loss for 1 year C2: 1-month vision loss LE C3: sudden vision loss in RE and LE blurred vision for 3-month	C1: BCVA: 16/20 in RE, 5/20 in LE. Inflammatory cells in both anterior chamber and vitreous cavity. Disc swelling and extensive hard exudates in posterior pole. Multiple retinal aneurysmal dilatations, vascular white sheath. C2: BCVA: 20/20 in RE and 20/500 in LE. Inflammatory cells, VItreous hemorrhage in LE vitreous. Bilateral peripapillary hard exudate deposition, disc swelling. C3: BCVA was counting fingers in RE, 20/60 in LE. Inflammatory cells, Exudate in the macula in	C1: Bilateral multiple retinal arteriolar aneurysms, optic disc, and peripheral CNP in both eyes. C2: RE: Multiple retinal aneurysms, optic disc, and peripheral CNP in both eyes. C3: vascular dilatations with retinal aneurysms. Peripheral CNP in both eyes	N/A	C1: Peripheral photocoagulation bilaterally, Triamcinolone Retrobulbar injection, Acetonide, and Lucentis intravitreal injection in LE. C2: Corticosteroids C3: Vitrectomy on RE.	C1: At 7y, BCVA remained at 20/20 in RE, 12/20 in LE. Some aneurysms diminished while some new developed. C2: At 6y, bilateral BCVA: 20/20 Few hard exudates on RE superior retina, proliferative fibrovascular band floating in LE. Aneurysms vanished, residual aneurysms decreased without recurrence. C3: At 2y, BCVA: 20/30 in both eyes. No evidence of neovascularization in anterior/ posterior segment.

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Table 2 (*continued*)

Paper NO.	First Author	Patient/s Age (Ys)/ sex	Chief complaint	Clinical Examination	FFA	Other Diagnostic tests	Treatment	Follow-Up
				LE. RE vitreous hemorrhage.				
5	Buğra Karasu	60/M	Visual impairment, metamorphopsia on LE	BCVA was counting 30/100 and 20/20, in LE, RE. Intraocular pressure:16-,15-mm Hg in RE, LE. Epiretinal membrane with a lamellar hole on LE. **Stage 1-2**	Bilateral aneurysms at optic disc and ischemic at inferotemporal retina of LE. Vitreous increased density.	Biochemical blood markers, Cranial MRI	Argon laser photocoagulation	LE BCVA: hand motion at 2 months later. Extensive, dense exudations, aneurysms at LE macula. Intravitreal dexametasone implant in LE. At 1-month, BCVA increased to hand motion from counting fingers at 2 m with exudations decreased and disappeared on LE.
6	Yu Jeat Chong	Pt1: 18/F Pt2: 20/ M Pt3: 25/ M	C1: asymptomatic, C2: blurred vision in LE. C3: N/A	C1: visual acuity: 6/6 in both eyes. Mild bilateral disc swelling, neovascularization temporally in LE. Ishihara color plates were full in both eyes. **stage 2-3.** C2: BCVA: 6/9 on RE, 6/24 in LE. 1 + activity in the vitreous bilaterally. -Marked ischaemic changes in RE, with peripheral vascular sheathing, arteriolar closure, neovascularization, macular exudates optic disc swelling. -Extensive macular exudates on RE and significant macular edema on LE. Arteriolar closure, neovascularization of optic disc, and macular exudates in LE. **stage 3** C3: visual acuity: 6/5 in RE and 6/4 in LE. 0.5+ cells in anterior chamber bilaterally, + cells in vitreous in both eyes. Bilateral ghost vessels, periphlebitis, retinal edema.. **stage 3**. Hx: C1: IRVAN in 2010, Tx. bilateral pan-retinal photocoagulation C3: Non-specific fundal changes in 2010.	C1: Aneurysmal dilatation, peripheral drop out. C2: Extensive capillary dropout in both eyes, late staining of the optic disc, and peripheral neovascularization on RE C3: Bilateral neovascularization at optic disc, sausage-like aneurysmal changes, peripheral vascular occlusion.	C1: ANA, ENA, Rheumatoid factors, complement levels, Anti-DNA, Anti-Cardiolipin antibodies, Toxoplasma screen, T-spot test, syphilis antibodies, OCT. C2: OCT, +CMV IgG, and EBV antibodies. C3: Subsequent uveitis blood investigations N	C1: Bilateral PRP C2: Bilateral PRP + Corticosteroids C3: Corticosteroids	C1: BCVA remained 6/9 in both eyes. Asymptomatic. Evidence of increased hemorrhage. Recurrence peripheral capillary dropout. PRP rearranged. BCVA: 6/6 vision in both eyes for 7 y. C2: At 3-month, BCVA: 6/6 in RE, BCVA: 6/60 in LE. Pre-retinal hemorrhage treated with PRP in RE Subsequent intravitreal Ozurdex implantation in LE. BCVA 5-month post-intravitreal Ozurdex: 6/9 in RE and 3/60 in LE. Bilateral 1+ cells in anterior chamber, vitreous inflammation on LE treated with topical steroids. 1-month later, BCVA: 6/18 in RE and 1/60 in LE. FFA: New vessels in RE, and new areas of peripheral capillary dropout in LE. OCT: increased exudates and macular thickness in both eyes. C3: 3-month, vitreous hemorrhage in RE, treated with bilateral PRP. 6-month, BCVA: 6/18 in RE, 6/6 in LE. Methotrexate. At 8-month, Mycophenolate mofetil. Next 4-month, PRP to RE, LE. At 1y, BCVA: 6/60 in RE, 6/9 in LE. Bilateral vitreous hemorrhage treated with bevacizumab (Avastin) injections in RE, PRP in LE. Over next 3 y, persistent neovascularization bilaterally, recurrent vitreous hemorrhages in RE. Treated with PRP in RE and LE and

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Table 2 (*continued*)

Paper NO.	First Author	Patient/s Age (Ys)/ sex	Chief complaint	Clinical Examination	FFA	Other Diagnostic tests	Treatment	Follow-Up
								bevacizumab injections to RE. At 48-month, vitrectomy in view of recurrent hemorrhages. At 58-month, further vitreous hemorrhage in RE, gross retinal ischemia, subretinal fluid and neurosensory detachment in LE. Limited perfusion of central macula in LE managed by Immunosuppression, MMF, and steroids. At 72-month, BCVA: remained stable with MMF and steroids.
7	Chandrakumar Balaratnasingam	31/F	Bilateral blurred vision	BCVA: 20/30. Intraocular pressures: 15 mmHg. Multiple retinal aneurysmal dilatations and optic nerve arteries. Neuroretinitis, macular exudation and vascular sheathing in both eyes	Circumferential, peripheral CNP	Chest CT, carotid Doppler ultrasonographym, Laboratory tests.	Laser photocoagulation, Corticosteroids, Intravitreal injections Banibizumab	BCVA improved to 20/25. Vascular sheathing, aneurysmal dilatations, persistentmacular exudation, increased areas of peripheral nonperfusion, staining of retinal arteries and veins.
8	Álvaro Rodríguez	21/F	Exotropia, central scotoma in left eye	Visual acuity: 20/20 in RE, 20/100 in LE. Cicatricial closure. LE: noniatrogenic foveal scar, persistently diminished visual acuity. -N macula in RE, macular scar in LE with atrophy of retinal neuroepithelium, foveal retinal pigment epithelium hyperplasia in 2012. -Peripheral retinal areas of ischemia and neovascularization regressed in 2016, inferior and temporal peripheral vasculitis, and area of inferonasal and peripheral temporal ischemia in LE. Left optic disk aneurysm in involution.	Bilateral triangular aneurysms, macular stars, peripheral temporal retinal ischemia in LE, early peripheral retinal neovascularization.	Biomicroscopy	Panretinal argon laser photocoagulation, 2 in RE, 3 sessions in LE	Visual acuity: 20/20 in RE, 20/400 in LE. Retinal vasculitis and neuroretinitis with macular star bilaterally.
9	Devesh Kumawat	16/F	Vision Diminution in both eyes over 2-month	BCVA: 20/120 in RE and finger counting in LE. **Stage 2** Sluggishly reactive pupil in LE. A few vitreous cells in both eyes. Disk hyperemia, macular and peri-vascular hard exudates, multiple	Multiple arterial aneurysms. Neovascularization absence	Systemic and biochemical tests	Corticosteroids	At 16-month, BCVA: 20/40 in RE, 20/400 in LE. Improvement in both eyes. -Macular exudates, retinal hemorrhages, and peri-vascular exudation decreased. Resolution of retinal aneurysms and absence of disk leakage. -LE: minimal vascular leakage in superotemporal quadrant, RE: no

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Table 2 (*continued*)

Paper NO.	First Author	Patient/s Age (Ys)/ sex	Chief complaint	Clinical Examination	FFA	Other Diagnostic tests	Treatment	Follow-Up
				arteriolar dilatations, retinal hemorrhages in both eyes. OCT: severe intraretinal exudation with loss of normal foveal architecture.				vascular leakage. Foveal architecture disturbed in RE, persistent hard exudates, and fibrosis in LE.
10	Rohan Ameratunga	51/M	Asymptomatic	Visual acuity: 6/5 bilateral. **Stage 2** -Macular star, retinal aneurysm and nonaneurysm -Mild thickening of peripheral nasal macula, peripapillary retina. -A mild epiretinal membrane at right macula. -Large sectors of perivascular leakage, capillary dilatations, pruning of capillary networks, closure of capillary beds, adjacent vascular leakage.	Peripheral retinal capillary closure	Indocyanine green angiography, Peripheral angiography, CXR	Intravenous pulse methylprednisolone prednisone. Fluorescein Angiography–targeted peripheral retinal photocoagulation.	Rapid regression of aneurysms, retinal vasculitis. No recurrence of neovascularization **(Stage 2).** At 5-month, aneurysms, venous caliber, exudation reduced, and beading resolution. Normalization of retinal thickness. Leakage decreased and no neovascularization. No change in visual acuity.

Abbreviations: IRVAN: Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis; FFA: Fluorescein Fundus Angiography; OCT: Optical Coherence Tomography; PRP: Pan-Retinal Photocoagulation; RPE: Retinal Pigment Epithelium; ANA: Antinuclear Antibodies; HLA-B51: Human Leukocyte Antigen B51; LE: Left Eye; RE: Right Eye; Pt: Patient; F: Female; M: Male; Ys: Years; N: Normal; BCVA: Best Corrected Visual Acuity; NO.: Number; N/A: Not applicable; Hx: Medical History; CXR: Chest Radiography; MRI: Magnetic Resonance Imaging; CNP: Capillary Non Perfusion; CT: Computed Tomography; OCTA: Optical Coherence Tomography Angiography; C: Case.

Availability of data and materials

Not applicable. All data (of the patient) generated during this study are included in this published article and its supplementary information files.

Patient perspective

The patient was cooperative and happy by the procedures done.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Sources of funding

There were no sources of funding.

Ethical approval

Not required for this case report.

Consent

Written informed consent was obtained from the patient for publishing this case report and any accompanying/identifying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

All authors accepted and approved the final manuscript.

Marah Mansour: design of the study, data interpretation, and analysis, drafting, critical revision, approval of the final manuscript.

Basel Al-Ghotani: data collection, data interpretation, and analysis, critical revision, drafting, approval of the final manuscript.

Bana Abo-Shdeed: data collection, data interpretation, and analysis, critical revision, drafting, approval of the final manuscript.

Omaya Jannoud: The Supervisor, patient care, drafting, critical revision, approval of the final manuscript.

Registration of research studies

1. Name of the registry: this case report is not a first time of reporting, new device or surgical technique. So I would not need a Research Registry Unique identifying number (UIN).
2. Unique Identifying number or registration ID:N/A
3. Hyperlink to your specific registration (must be publicly accessible and will be checked):N/A

Guarantor

Omaya Jannoud.

Declaration of competing interest

All the authors declared that they have no conflicts of interest.

Acknowledgment

Not applicable.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2022.103725.

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