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Case Report

A rare case of idiopathic retinal vasculitis, aneurysms and neuroretinitis (IRVAN) syndrome

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exudation in macula causing vision impairment. Peripheral capillary non perfusion is another feature of this entity and predisposing factor for vision loss.^{1,3}

We report a case of a young male patient with bilateral vision loss and typical features of IRVAN syndrome who was managed successfully at our centre.

Case report

A 36 year old male patient was referred to our centre with history of diminution of vision in both eyes for last one month duration. Patient noticed painless diminution of vision which progressed over a period of few days. No history of any ocular trauma, redness, floaters or flashes of light. However, patient gave history of maxillary and frontal sinusitis two months back for which he was hospitalized and treated with intravenous antibiotics. Patient had recovered fully and was not on any medications when he developed diminution of vision. There was no other significant past history.

Ocular examination showed best corrected visual acuity (BCVA) in right eye was counting fingers close to face with accurate projection of rays in all quadrants. BCVA in left eye was 6/24. In both eyes pupils were sluggishly reacting but no relative afferent pupillary defect present. Anterior segment was within normal limits with open angles and no neo vascularization of iris or angle in both eyes.

On posterior segment examination grade 1 + vitreous cells were present in both eyes. Fundus examination right eye showed sheathing of vessels in temporal quadrant, retinal edema with multiple soft and hard exudates in macular area

Introduction

Idiopathic retinal vasculitis, aneurysms and neuroretinitis (IRVAN) syndrome is a rare but well recognized clinical entity. It is characterized by bilateral disease with retinal vasculitis, aneurysmal dilatations of arterioles, exudation in retina and neuroretinitis. Chang et al first recognized this new entity and proposed the acronym IRVAN.¹ Since then various reports of IRVAN syndrome have appeared in literature. Samuel et al conducted largest cohort study and gave detailed observation and proposed staging of this syndrome.² The disease characteristically involves young individuals who have no associated systemic illness.^{1–3} Involvement is typically bilateral with

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Fig. 1 – a) colour fundus photograph of right eye showing multiple soft and hard exudates in macular area (arrow) along with vascular sheathing superotemporally (line arrow) b) colour fundus photograph of left eye showing multiple soft exudates, aneurysmal dilatation of arterioles (arrow head) and macular star pattern (arrow).

(Fig. 1a). Left eye fundus examination showed multiple aneurysmal dilatations of retinal arterioles, soft exudates and hard exudates arranged in star shaped pattern in macula (Fig. 1b). Optical coherence tomography (OCT) done to assess macular status which showed macular edema and sub retinal fluid in both eyes (Fig. 2). Intra ocular pressure was 12 mm Hg in right eye and 14 mm Hg in left eye by applanation tonometry.

Fundus fluorescein angiography (FFA) was performed which revealed aneurysmal dilatation of retinal vasculature in both eyes with leakage in early phase and capillary non perfusion areas in periphery (Fig. 3).

Systemic examination was within normal limit (WNL). Routine blood and urine examination normal. HIV, HBsAg, TORCH titer and VDRL negative. Renal and liver function test WNL. Patient underwent MRI brain and orbit which showed normal study.

Initial clinical diagnosis of bilateral neuroretinitis was made due to bilateral macular exudates forming star shape but after FFA diagnosis was revised to IRVAN syndrome. Patient was

treated with intravenous Methylprednisolone 1 gm/day for three days and followed by oral prednisolone at 1 mg/kg with slow tapering. Patient showed good response with resolution of retinal edema and exudates in both eyes. At three weeks following initiation of treatment, BCVA 6/24 in right eye and 6/9 in left eye with no signs of neovascularisation (Fig. 4). Patient has been placed under regular follow up.

Discussion

The first reported association between bilateral retinal arteritis and multiple aneurysmal dilatations with neuroretinitis was provided by Kincaid et al.⁴ However, recognition of this new entity as IRVAN was established by Chang et al and Samuel et al after studying large cohort of patients, proposed the diagnostic criteria^{1,2} (Table 1). Our patient had all major as well as minor criteria fulfilled. The disease has no systemic

Fig. 2 – OCT macula showing macular edema and sub retinal fluid (arrow).

Fig. 3 – FFA showing aneurismal dilatation of retinal vasculature in both eyes with leakage (arrow) and capillary non perfusion areas in periphery (arrow heads).

Fig. 4 – colour fundus photograph showing partial resolution of exudates.

association. However, reports of positive P-ANCA tests in some cases have been reported.⁵

Based on clinical manifestations of progressive ischemia, IRVAN has been divided into 5 stages (Table 2).² Our patient at the time of diagnosis was in stage 2. The natural history and visual outcome of the disease vary. However, if left untreated it may lead to severe bilateral visual loss.^{2,3}

Over the years, many therapeutic regimens have been evaluated. Medical management includes use of steroids to control the inflammation and works in early stage of disease.³ Pan retinal photocoagulation (PRP) been suggested as the treatment of choice for IRVAN in the presence of widespread retinal ischemia so as to prevent the development of ocular neovascularization and vitreous hemorrhage.^{2,3} Initiating PRP before or shortly after the development of any neovascularization might reduce the likelihood of disease progression and loss of vision.³

Table 1 – Criteria for diagnosis.

Major criteria	Retinal vasculitis Aneurysmal dilations of arterioles Neuroretinitis
Minor criteria	Peripheral capillary nonperfusion Retinal neovascularization Macular exudation

Favorable results have been noted with use of Ranibizumab.⁶ Monoclonal antibodies such as Infliximab have been tried in treatment of IRVAN.⁷

Duration of disease is variable and may take many months to years to manifest.³ In our case presentation was acute and bilateral which prompted early initiation of treatment. Macular edema and exudates contributed to vision loss. Areas of CNP were seen in peripheral retina not associated with NVE. Patient is under regular follow up to detect sequels of non perfusion and intervention if required.

Table 2 – Stages of IRVAN syndrome.

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

Conflicts of interest

All authors have none to declare.

REFERENCES

1. Chang TS, Aylward GW, Davis JL, et al. Idiopathic retinal vasculitis, aneurysms, and neuro-retinitis. *Ophthalmology*. 1995;102:1089–1097.
2. Samuel MA, Equi RA, Chang TS, et al. Idiopathic retinitis, vasculitis, aneurysms, and neuroretinitis (IRVAN): new observations and a proposed staging system. *Ophthalmology*. 2007;114:1526–1529.
3. Rouvas A, Nikita E, Markomichelakis N, Theodossiadis P, Pharmakakis N. Idiopathic retinal vasculitis, arteriolar macroaneurysms and neuroretinitis: clinical course and treatment. *J Ophthalmic Inflamm Infect*. 2013;3:21.
4. Kincaid J, Schatz H. Bilateral retinal arteritis with multiple aneurysmal dilatations. *Retina*. 1983;3:171.
5. Ramin N, Talieh M, Nasim A, Hossein H, Masoud S. Idiopathic retinal vasculitis, aneurysms and neuroretinitis syndrome associated with positive perinuclear antineutrophil cytoplasmic antibody. *J Ophthalmic Vis Res*. 2011;6(4):330–333.
6. Karagiannis D, Soumplis V, Georgalas I, Kandarakis A. Ranibizumab for idiopathic retinal vasculitis, aneurysms, and neuroretinitis: favorable results. *Eur J Ophthalmol*. 2010;20:792–794.
7. Cheema RA, Al-Askar E, Cheema HR. Infliximab therapy for idiopathic retinal vasculitis, aneurysm, and neuroretinitis syndrome. *J Ocul Pharmacol Ther*. 2011;27:407–410.