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Bilateral neovascular glaucoma in idiopathic retinal vasculitis, aneurysms, and neuroretinitis syndrome

The association of bilateral retinal macroaneurysms with retinal vasculitis and neuroretinitis was first described by Kincaid and Schatz.[1] The condition later was renamed as idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN).[2] The retinal vasculitis may result in capillary occlusion and capillary nonperfusion. This causes retinal ischemia, leading to release of vascular endothelial growth factors, and subsequently results in anterior segment neovascularization causing neovascular glaucoma (NVG). We report a case of a young girl with IRVAN who presented with neovascular sequelae of the disease. She was treated with pan-retinal photocoagulation for posterior segment ischemia and was given intracameral bevacizumab injection, which was followed by mitomycin-C-augmented trabeculectomy for NVG in both eyes.

Case Report

A 17-year-old female presented to our tertiary eye care centre with complaints of gradually progressive diminution of vision in her left eye (LE) for >3 months. She had no systemic complaints. Her best corrected visual acuity (BCVA) was 6/9 right eye (RE) and 6/60 LE. On examination, her intraocular pressure (IOP) was 18 and 40 mm Hg in RE and LE, respectively. Slit lamp biomicroscopy revealed florid neovascularization of iris (NVI) in the LE with keratic precipitates, whereas NVI was absent in the RE. On gonioscopy, fine-angle neovascularization along with presence of goniosynechiae was seen in the RE. In the LE she had total synechial angle closure (Fig. 1). Stereoscopic fundus examination with 90D lens showed temporal disc pallor in both eyes along with mild blurring of disc margins (Fig. 2). The vertical cup/disc ratio was 0.4:1 RE and approximately 0.8:1 LE. On indirect ophthalmoscopy, both eyes had multiple aneurysmal dilatations of arterioles, vascular sheathing, sclerosis of peripheral arterioles, and exudation over macula (Fig. 3a). Fluorescein angiography of both eyes confirmed the presence of aneurysms and capillary nonperfusion (CNP) areas (Fig. 3b). The arm–retinal circulation time was 8 seconds, thus ruling out circulation delay in the patient.

Systemic investigations were done. Her hemoglobin level was 11.5 mg/dL and total leucocyte count was 8500, with polymorphonuclear leucocytes being 64%, lymphocytes 34%, monocytes 2%, and eosinophils 2%. Erythrocyte segmentation rate was found to be 14 mm at 1st

Fig. 1—Goniophotograph of the left eye showing total synechial closure with keratic precipitates.

Fig. 2—Left eye disc photograph showing neuroretinitis.

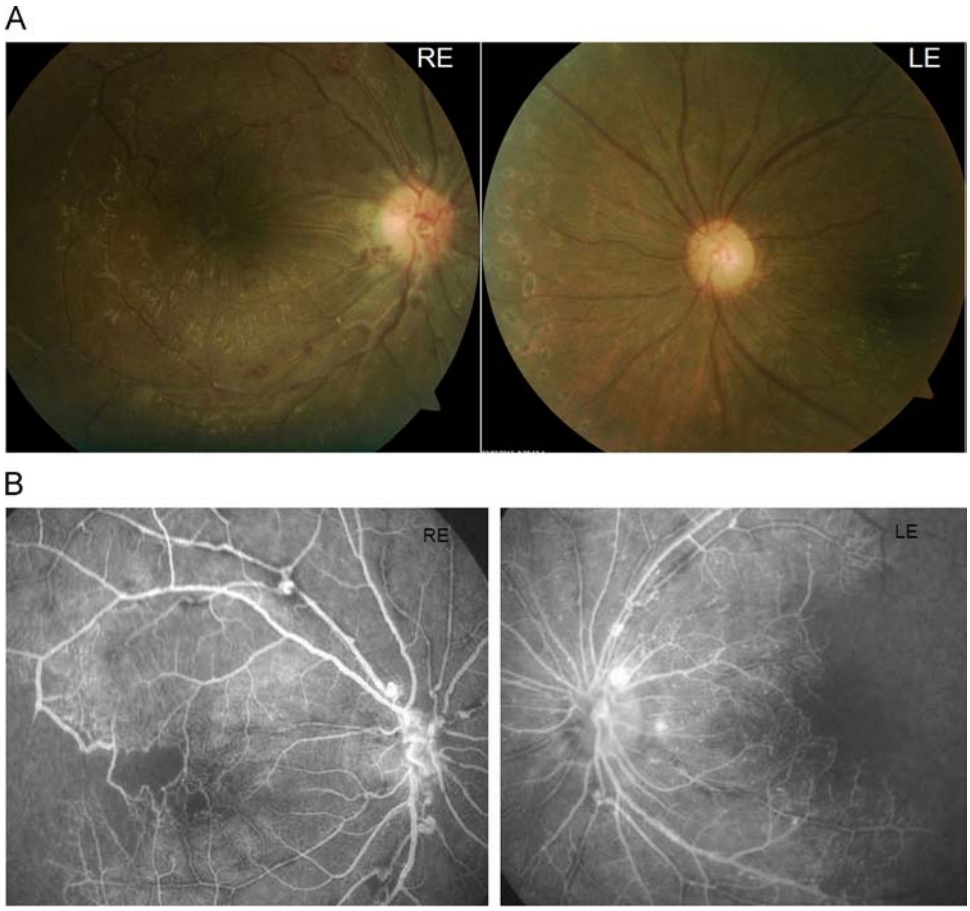

Fig. 3—(A) Fundus photographs of right eye (RE) and left eye (LE) taken after the first phase of pan-retinal photocoagulation showing aneurysmal dilations of arterioles, vascular sheathing, flame-shaped hemorrhages along the vessels, and sclerosis of arterioles in periphery and macular exudation. (B) Fundus fluorescein angiography of both the eyes showing aneurysmal dilations of arterioles with capillary nonperfusion areas.

hour, which was normal, and peripheral blood smear did not reveal any abnormalities.

Findings of serological tests, namely, Venereal Disease Research Laboratory test (VDRL), ELISA for HIV 1 and 2, rheumatoid arthritis factor, and antinuclear antibody (ANA), were negative. C-reactive protein (CRP) and serum angiotensin converting enzyme (SACE) levels were within the normal range. Results of stool and urine routine and microscopic examination were normal. Findings from the chest x-ray study were normal, and the result of Mantoux test was negative. A thorough cardiovascular examination ruled out Takayasu's disease and carotid stenosis. On the basis of

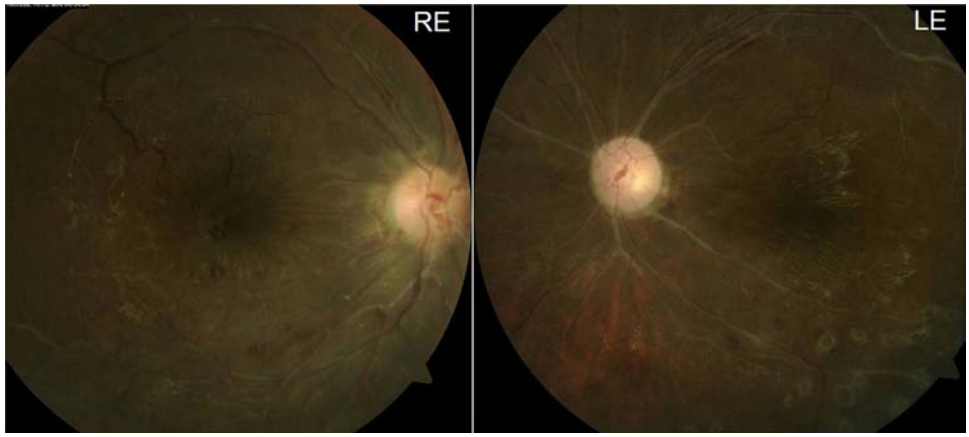

Fig. 4—Fundus photographs of right eye (RE) and left eye (LE) taken after 12 months of trabeculectomy in her LE

the fundus examination, fluorescein angiographic findings, and absence of systemic abnormality, a diagnosis of idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) with bilateral neovascular glaucoma (NVG) was made.

The patient was immediately started on maximal topical and systemic antiglaucoma therapy for IOP control of the LE. This included systemic acetazolamide 500 mg administered 3 times in a day and 2 topical antiglaucoma medications, timolol 0.5% and brimonidine 0.2%. However, her IOP remained high. First-stage photocoagulation (PRP) was performed in both eyes. Intracameral bevacizumab (1.25 mg/0.05 mL) was injected in the LE, and mitomycin (0.04%)-augmented fornix–based trabeculectomy was performed 36 hours later in her LE. After trabeculectomy, her IOP was well controlled without any antiglaucoma medications at 2 weeks. Second stage of PRP was completed at this time for both eyes, after which angle neovascularization regressed in her RE also.

She had stable IOP and visual acuity in both eyes for up to 8 weeks after surgery, after which she presented with pain and decreased vision in her RE. On examination, she had florid NVI with IOP of 54 mm Hg RE and 18 mm Hg LE. Mitomycin (0.04%)-augmented trabeculectomy was performed in her RE also. The patient maintained IOP of 12–14 mm Hg in both the eyes without antiglaucoma therapy for 12 months after this surgery. Her BCVA was 6/18 and 6/60 in RE and LE, respectively. However, in the next 1 year, because of progressive vasculitis, sheathing, and sclerosis (Fig. 4), she experienced 2 episodes of vitreous hemorrhage in each eye. Consequently, she underwent pars plana vitrectomy along with retinal endolaser in both the eyes, after which her IOP again increased in both the eyes, for which topical antiglaucoma medications had to be re-instituted. Although her IOP remained controlled on topical antiglaucoma medications, her visual acuity dropped progressively to 6/36 RE and finger counting close to face in LE over the next 12 months.

Discussion

Because IRVAN is a diagnosis of exclusion,[2–4] one needs to rule out other causes of retinal vasculitis and neuroretinitis. Chang et al.[2] defined 3 major criteria (retinal vasculitis, aneurysmal dilations at arterial bifurcations, and neuroretinitis) and 3 minor criteria (peripheral CNP, retinal neovascularization, and macular exudation) to diagnose IRVAN. Samuel et al.[3] described a case series of 22 patients and proposed a staging system for IRVAN, in which NVG was categorized as stage 5. Hence, our patient suffered from stage 5 IRVAN in both eyes.

An essential part of management for IRVAN involves panretinal PRP that needs to be performed at the earliest evidence of nonperfusion areas in the retina. Although topical steroids may be added as a temporizing measure to control anterior segment inflammation, the role of systemic steroids and immunosuppressive agents is controversial.[3] NVG when present should be managed with mitomycin-C-augmented trabeculectomy[5] after intracameral Bevacizumab as was performed for this patient. Not many reports on the long-term outcomes of managing bilateral NVG among patients with IRVAN syndrome are available in the literature.[2] The use of Bevacizumab as an adjunct in the therapy for IRVAN was described by Jad Akesbi et al.,[6] who reported on a patient with IRVAN having bilateral NVI with open angles and normal IOP who was successfully treated with intravitreal Bevacizumab followed by PRP in one eye and pars plana vitrectomy with endolaser in the other eye for vitreous hemorrhage. Injecting intracameral or intravitreal bevacizumab before trabeculectomy or vitrectomy, respectively, helps control ocular neovascularization till surgery is performed besides controlling excessive bleeding intraoperatively.[7]

Our case emphasizes poor long-term visual prognosis for this rare disease caused by the relentless retinal ischemia even with timely medical and surgical interventions and regular follow-up.

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