Sir,
An atypical case of idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) syndrome

IRVAN syndrome is a retinal vasculitic disorder characterized by idiopathic retinal vasculitis, aneurysms, and neuroretinitis.¹ Young women are most frequently affected, and most cases are not associated with any systemic disorder.^{2,3} The typical features of this condition are prominent multiple aneurysmal dilatations of retinal arterioles, neuroretinitis manifesting as diffuse staining of the optic nerve head in the late fundus fluorescein phase, and extensive nonperfusion of peripheral capillaries. Here, we report an atypical case of IRVAN syndrome, and describe the fundus fluorescein and indocyanine green angiography findings of the disease.

Case report

A 55-year-old woman presented to our vitreo-retinal unit complaining of poor vision in her left eye. She had had this problem for 2 weeks. Her medical history included mitral valve replacement at 21 years of age due to acute rheumatic fever at that time. She was still taking antiaggregant treatment (dipyridamole, drisentin). The systolic and diastolic blood pressures were 110 and 70 mmHg, respectively, and there was no electrocardiographic abnormality.

On initial ophthalmologic examination, the patient's visual acuity was counting fingers at 1 m in the left eye and 20/20 in the right eye. Intraocular pressure was 16 mmHg bilaterally. The anterior chamber of the left eye showed 1+ cells and aqueous flare. The left fundus showed diffuse intra and subretinal haemorrhage, while fundus examination of the right eye was normal (Figure 1a, b). The retinal arteries on the left exhibited multiple aneurysmal dilatations. There was also diffuse

perivascular exudation in the temporal region of the fovea and the superior portion of the optic disc. The peripheral retinal arteries were attenuated, and peripheral retinal neovascularization was noted in the superotemporal region. No cells were detected in the vitreous.

Fundus fluorescein angiography (FFA) of the left eye revealed dye leakage and hyperfluorescence of the optic disc, and dye leakage at the macula and superotemporal region. The leakage lesions were observed in late-stage FFA only. FFA also demonstrated the above-mentioned aneurysmal dilatations of the retinal arteries. 'Tied-knot-like' dilatations formed from the aneurysms were particularly prominent in the area of the superior temporal artery (Figure 2a, b). Hypofluorescent areas indicating haemorrhage and peripheral temporal ischaemia were observed, and regions of capillary non-perfusion and neovascularization were also noted (Figure 2c, d). FFA of the right eye revealed only minor unremarkable abnormalities.

Tied-knot-like sacculations were better visualized with ICG angiography because of the lack of dye leakage (Figure 3). The aneurysms remained filled and stained until the late stages.

Systemic enquiry was unremarkable and extensive laboratory investigations for underlying thrombophilia, connective tissue disease were normal. Doppler imaging of the carotid arteries and chest radiograph were also normal. The patient was negative for human leucocyte antigens B51 and B27.

Based on these findings, topical steroid treatment (dexamethasone, Dexamethasone, 4X1) was initiated and scattered retinal photocoagulation therapy was applied to the avascular peripheral retina and to the macroaneurysms, which were surrounded by exudations. At 6 months after photocoagulation, the visual acuity in the left eye was 20/200 and the patient's condition was quiescent, with reduced haemorrhage and

Figure 1 Diffuse intra and subretinal haemorrhages and perivascular exudations in the left eye (a) and (b) revealed normal right fundus.

Figure 2 FFA of the left eye revealed 'tied-knot'-like dilatations formed from the aneurysms in the region of the superior temporal artery (a), leakage and hyperfluorescence of the optic disc in the late stages (b), regions of capillary nonperfusion and neovascularization (c, d).

exudation. Repeat FFA at this stage revealed regression of the neovascularization and decreased exudation.

Comment

Kincaid and Schatz³ were the first to describe the clinical association of macroaneurysms with retinal vasculitis and neuroretinitis. Chang *et al*¹ described the biggest series of this condition and were the first to coin the acronym IRVAN. This condition usually affects young, healthy females, and its major features are multiple tied-knot-like aneurysmal dilatations of the retinal arteries, exudative retinopathy, diffuse staining of the optic disc, and nonperfusion of peripheral capillaries.^{4,5} IRVAN syndrome affects the retinal and optic disc vasculature, and also the choroidal circulation in some cases. Vascular dilatation may be a result of the inflammatory process in the retinal artery walls.

Since both the retinal and choroidal vessels can be damaged in this disorder, the differential diagnosis includes a range of inflammatory and infectious vascular diseases. Behçet's disease, sarcoidosis,⁶ multiple sclerosis,⁷ and collagen-related vascular disorders such as polyarteritis nodosa and systemic lupus

erythematosus may also cause neuroretinitis and macroaneurysms. In previously reported cases of this syndrome, no specific systemic pathology has been identified. As in other cases, the comprehensive work-up of our patient revealed no underlying systemic disease. The only notable abnormality in our case was the replaced mitral valve.

Retinal arterial macroaneurysms are acquired vascular abnormalities that are rarely seen in individuals less than 60 years of age.^{8,9} Leakage of vascular contents through the arterial walls causes exudation and macular oedema, which severely reduces visual acuity. These large saccular dilatations are strongly associated with long-standing hypertension. In our case, there was no history of hypertension.

Based on the findings of exudative retinopathy, nonperfusion of peripheral capillaries, diffuse staining of the optic nerve head in late FFA (neuroretinitis), and mild uveitis, we diagnosed this healthy middle-aged woman with IRVAN syndrome. The patient's age, the unilateral nature of her condition and the haemorrhage from the aneurysms that we observed are not typical features of this disorder. The haemorrhage may be due to the fragile arterial wall caused by chronic inflammation

Figure 3 The tied-knot-like sacculations were better visualized with indocyanine green angiography because of the lack of dye leakage with this method.

or secondary to the antiaggregant treatment that the patient has been given.

Since oral steroids are reportedly ineffective in IRVAN syndrome,¹ we prescribed only topical steroid for her mild anterior uveitis. Pan-retinal photocoagulation may be effective for select cases with extensive peripheral ischaemia and retinal neovascularization. At 6 months of follow-up, the visual acuity in her left eye was 20/200. Careful long-term follow-up is recommended for all patients with this syndrome.

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