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Sir,

Central retinal vein and ophthalmic artery occlusion in primary antiphospholipid syndrome

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Primary antiphospholipid syndrome is characterized by the production of moderate to high levels of antiphospholipid antibodies, associated with thrombotic phenomena (arterial or venous) and recurrent spontaneous abortion due to placental vascular insufficiency, in the absence of any other recognizable autoimmune disease.[1]

Primary antiphospholipid syndrome has been found to be associated with ocular ischaemia and retinal vascular occlusion, due to arteriolar or venular thrombosis.[2–6] We report a case of a 60-year-old woman with primary antiphospholipid syndrome, who developed consecutive occlusion of the central retinal vein and ophthalmic artery. This is the first reported case in patients with this syndrome.

Case report

A 60-year-old woman presented with a 1-day history of slurred speech and left-sided weakness. She also described an episode of transient dimming of vision in the right eye that lasted 4 h. She had a history of mild hypertension and noninsulin-dependent diabetes mellitus that was diagnosed 6 months before, both of which were adequately controlled by lifestyle modification and dietary measures alone. Neurological examination revealed a left-sided haemiparesis and a left upper motor neuron facial nerve palsy. Visual acuity was 6/7.5 OU. Funduscopic examination was normal. Magnetic resonance imaging of the brain demonstrated the presence of a right corona radiata infarct. Duplex carotid ultrasonography showed moderate dampening of the velocity and waveform of the right internal carotid artery blood flow compared to the left. The patient was started on ticlopidine.

After 5 months, she presented with a sudden reduction in vision acuity in the right eye to 6/18. Intraocular pressures were normal. Fundus examination revealed a right central retinal vein occlusion. She was screened for autoimmune and procoagulant disorders. Pertinent laboratory investigations were as follows: the erythrocyte sedimentation rate was 57 mm/h; IgG anticardiolipin was raised to 33 GPL units/ml; antinuclear antibody and lupus anticoagulants were negative. Serum total complement, C3, C4, and protein S, protein C and antithrombin III were normal. The diagnosis of primary antiphospholipid syndrome was established and the patient was started on anticoagulation with warfarin.

After 1 month, she presented with sudden deterioration of her right vision to perception of light only. A right relative afferent pupillary defect was present. There was neovascularization of the iris, with a raised intraocular pressure of 26 mmHg. Fundus examination revealed the presence of a right ophthalmic artery occlusion, with severe whitening of the retina and markedly attenuated retinal arterioles and venules (Figure 1). Fundus fluorescein angiography demonstrated blocked choroidal and retinal artery filling. Transcranial doppler ultrasound of the ophthalmic arteries revealed an absent signal over the right ophthalmic artery and a normal signal from the left. The international normalized ratio (INR) was 1.6. Immediate panretinal photocoagulation was instituted. She was started on oral acetazolamide 250 mg bid and topical timolol 0.5% bid and atropine 1% tid to the affected eye. Warfarin therapy was optimized to INR 2.5–3.0. There was gradual resolution of the iris neovascularization and eventual normalization of the intraocular pressure. However, the final visual acuity in the right eye was hand movements only. She remained asymptomatic in the left eye.

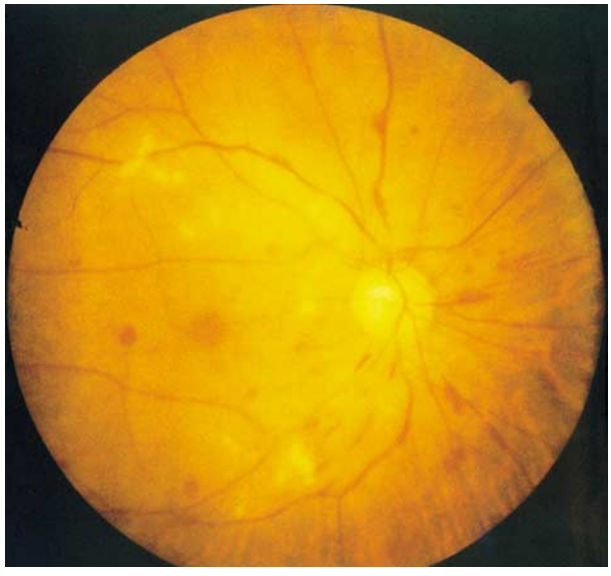

Figure 1 Fundus photograph of the right eye following central retinal vein and ophthalmic artery occlusion.

Comment

Patients with primary antiphospholipid syndrome are at risk of developing systemic and cerebral thromboembolism. The frequency of ocular vaso-occlusive disorders in patients with this syndrome ranges from 0.5 to 8%, with the majority affecting the retinal vasculature.[2–5] Both the arterial and venous systems may be involved.

Clinical features include microaneurysms, vitreous and preretinal haemorrhage, anterior ischaemic optic neuropathy, as well as retinal and choroidal vascular occlusions.[2–5] Iris neovascularization tends to occur early,[6] as was demonstrated in our patient. Castanon *et al*[3] reported a high prevalence of ocular disease in patients with primary antiphospholipid syndrome, with 88% of patients (15 out of 17 patients) demonstrating funduscopic abnormalities, and 29% of patients having vaso-occlusive retinopathy.

Consecutive central retinal vein and ophthalmic artery occlusion is extremely rare in patients with primary antiphospholipid syndrome. This is the first reported case of this occurrence.

Antiphospholipid antibodies are believed to promote thrombotic events due to its action on the phospholipid component of platelet membranes and vascular endothelium, as well as on thrombotic factors such as local prostacyclin, antithrombin III, and protein C activation.[6,7] Anticoagulation with warfarin has been advocated in patients with vaso-occlusive retinopathy as it may assist in the reperfusion of the ischaemic retina and reduce the risk of neovascularization.[8]

Ophthalmologists should consider screening for antiphospholipid antibodies in patients without obvious cardiovascular risk factors, who present with ocular and systemic vaso-occlusive disease.

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Sir,

Ocular syphilis: the return of an old acquaintance
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Syphilis is an overlooked cause of uveitis with past reports showing delays in diagnosis leading to irreversible visual loss.[1] The epidemiology of syphilis is changing, with infection now beginning to re-emerge after being thought to have been eradicated.[2]

Case report

A 30-year-old homosexual male, with no past medical or ophthalmic history, presented with a 3-week history of a red right eye associated with mild photophobia. Examination revealed a mild anterior uveitis with no posterior synechiae formation, vitreous activity, or retinal pathology. Usual treatment for anterior uveitis was commenced—topical steroid and mydriasis—and the patient was sent for blood tests, including RPR, FTA-ABS, ACE estimation, and HLA-B27.