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Sir,

Recurrent retinal artery occlusion after the disappearance of lupus anticoagulant

Antiphospholipid antibodies are a cause of recurrent arterial and venous thrombosis, and may lead to retinal vascular occlusion, especially in young people.[1] Therapeutic options to prevent further vascular occlusions are varied, but it is recommended that treatment be continued until at least 6 months after the disappearance of the antibody.[2,3] We describe a patient with lupus anticoagulant who had recurrent branch retinal artery occlusion (BRAO) long after the disappearance of the antibody and despite treatment with oral steroids and aspirin.

Case report

A 29-year-old women presented in November 1995, 1 week after the onset of a relative scotoma in the left eye. Visual acuity was 6/5 in both eyes and a superior BRAO was found in the left eye (Fig. 1). Ocular examination was otherwise unremarkable and no disc drusen were observed on clinical examination or subsequent MRI examination of the optic nerves. Her blood pressure was 130/90 mmHg and systemic examination was normal. There was no significant past ophthalmic or medical history, though she smoked 5 cigarettes daily. She had stopped taking the combined oral contraceptive pill a year previously, but there was no history of spontaneous abortion, migraine or intravenous drug abuse. The presence of lupus anticoagulant was confirmed that month on serological testing according to established criteria,[5] but had become absent when testing was repeated in January 1996. No other hypercoagulability state was detected on either occasion. The following investigations were also negative or normal: full blood count, erythrocyte sedimentation rate, glucose, cholesterol and an autoantibody screen including anti-nuclear factor and anti-neutrophil cytoplasmic antibody.

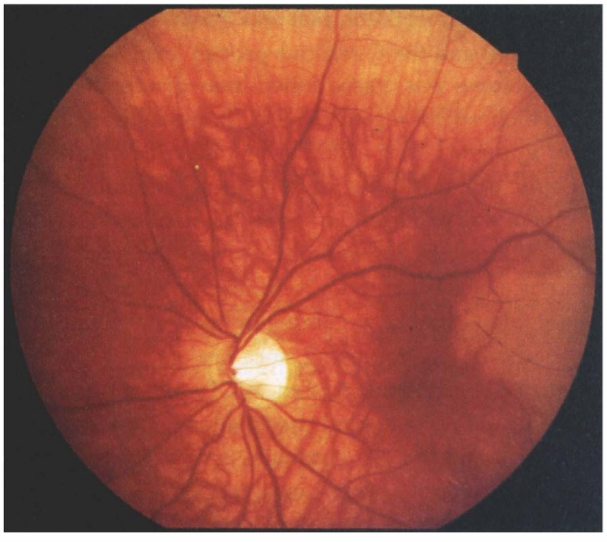

Fig. 1. *Colour fundus photograph of the left eye showing an area of retinal oedema supero-temporal to the fovea that occurred secondary to a branch retinal artery occlusion.*

The patient presented again in April 1996 with a relative scotoma, above fixation, in the right eye. Examination revealed a supero-temporal BRAO. She was commenced on treatment with aspirin 300 mg and prednisolone 30 mg daily. In February 1997 a left supero-temporal BRAO caused a further, relative, paracentral scotoma, although the acuity remained 6/5 in both eyes. No other neurological symptoms were reported. Repeat investigation for a hypercoagulability state was unremarkable and the lupus anticoagulant was absent on each of these occasions. Echocardiography and carotid duplex imaging were unremarkable. Pure tone audiometry suggested sensorineural thresholds within normal limits. MRI examination of the brain was unremarkable and no evidence was found to suggest a more diffuse microangiopathy.

Comment

Although antiphospholipid antibodies are frequently found in patients with autoimmune and malignant disease, they are most commonly detected in patients who are otherwise healthy.[2] Both lupus anticoagulant and anticardiolipin antibody may cause recurrent arterial and venous occlusion, recurrent abortion and thrombocytopenia. Retinal vascular occlusion, ischaemic optic neuropathy and amaurosis fugax have also been described.[3,4] Over 50% of patients with the antiphospholipid syndrome suffer further vascular occlusion after their initial thrombotic event.[6]

In this patient, lupus anticoagulant was detected at the initial presentation but has been consistently undetectable since. (Serological testing was based on established criteria,[5] with screening and confirmatory tests based on prolonged APTT, prolonged dRVVT and phospholipid dependency.) However, levels of lupus anticoagulant fluctuate during the course of the disease and the antibodies may disappear spontaneously, so repeated testing is necessary when the clinical suspicion is high.[7] Since the initial presentation, two further arterial occlusions have occurred, 15 and 25 months after the disappearance of the antibody. Recurrent BRAO, often in young women, has previously been described by others.[8,9] Although there are features in common with these previous reports, the presence of the lupus anticoagulant and the absence of symptoms or signs of vestibular or other neurological disease has led us to believe that the disease process in this patient is different.

Therapeutic options to prevent further vascular occlusion are varied and may involve antithrombotic agents alone or in combination with steroid immunosuppression to modulate the prothrombotic effect of the antibodies, and aggressive therapy with both modalities has been recommended for younger patients.[2–4] It is recommended that anticoagulation be continued until 6 months after the disappearance of the antibody.[2]

Given the lack of a consistent response to a specific therapy in the antiphospholipid syndrome and the possibility of future pregnancy, this patient was initially treated with oral prednisolone and with aspirin rather than intensive warfarin treatment (INR>3).[3,4,10] During 14 months of treatment she has had one further BRAO. In the light of experience with this case we would now anticoagulate with warfarin all patients with antiphospholipid syndrome and recurrent retinal vascular occlusion, and we would continue treatment indefinitely.

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Sir,

Bilateral anterior ischaemic optic neuropathy following amiodarone

Amiodarone, initially used in Europe as a coronary vasodilator and anti-anginal agent, is now widely used as an anti-arrhythmic for supraventricular and ventricular tachyarrhythmias. However, the drug has numerous pulmonary, hepatic, thyroid and neurotoxic adverse effects. Corneal microdeposits develop in nearly all patients. These do not significantly affect vision and are reversible on discontinuing the medication. Rarely, more profound effects on vision can develop with the drug.[1]

Case report

A 58-year-old man presented with a 5 day history of acute blurring of the lower half of the visual field in his left eye. He had no headache, jaw claudication, scalp tenderness or polymyalgia rheumatica. He had been taking amiodarone 200 mg b.d., bumetanide 1 mg o.d. and captopril 25 mg for the previous 2 months, having been diagnosed as having hypertrophic cardiomyopathy.

On examination, the visual acuity was 6/5 in the right eye and 6/5 in the left eye. A left relative afferent pupillary defect was present. The anterior segments showed vortex keratopathy and the intraocular pressures were normal. The superior half of the left optic disc was oedematous and pale, with a few splinter haemorrhages. The right disc was normal. Visual fields confirmed an inferior altitudinal defect in the left eye. He was generally fit and well. The blood pressure was 140/90 mmHg and he was in sinus rhythm. Haematological investigations, liver function tests, tests for syphilis, autoantibody screening and the coagulation profile including thrombophilia screening were all normal. The erythrocyte sedimentation rate was 5 mm/h (Westergren) and the C-reactive protein level < 10 g/l.

Amiodarone was discontinued immediately in consultation with the cardiologist. Six weeks later, the patient presented with a similar picture affecting the right optic disc (Fig. 1), with an inferior altitudinal field loss. There was pallor of the superior half of the left disc. Fundus fluorescein angiography showed the typical appearance of fresh anterior ischaemic optic neuropathy in the right eye and disc staining in the left. He had had no episodes of feeling unwell since his last visit and continued to be in sinus rhythm. A CT scan of the orbits and brain was normal. Oral prednisolone 60 mg was commenced and tapered gradually over 4 weeks. Unfortunately, there was no visual recovery in either eye.

The patient has been followed up for 18 months. He has a visual acuity of 6/6 in both eyes, atrophy of the upper halves of both optic discs and bilateral non-progressive inferior altitudinal field defects.