Combined central retinal artery and vein occlusion in a patient with systemic lupus erythematosus

Sir, The ocular manifestations of lupus include mucocutaneous lid involvement, secondary Sjögren’s syndrome, retinal vascular disease and neuro-ophthalmic disease [1]. Retinopathy is the most common ocular manifestation in lupus and usually consists of cotton-wool spots with intraretinal haemorrhages. The more severe form of lupus retinitis consists of retinal arteriolaritis and vascular occlusion, resulting in capillary non-perfusion, retinal haemorrhage and venous stasis. When larger vessels are involved, branch or central retinal artery or vein occlusion may result, with secondary retinal neovascularization and vitreous haemorrhage [1, 2]. It has been shown that patients with lupus and raised concentrations of antiphospholipid antibodies have a higher risk of developing retinal vaso-occlusive disease [3, 4]. Here we describe a case of combined central retinal artery and vein occlusion in a lupus patient without a raised concentration of antiphospholipid antibodies. To our knowledge, this is the first description of such a case.

A 23-yr-old man was admitted due to sudden visual loss in his right eye. He had been diagnosed as suffering from systemic lupus erythematosus 6 yr ago on the basis of oral ulcers, lupus nephritis with severe hypertension and renal failure, thrombocytopenia and abnormal titres of antinuclear antibodies (ANA). His history of venous or arterial thrombotic events was unremarkable. He was treated with anti-hypertensive agents (candesartan cilexetil and enalapril maleate) and systemic steroids (prednisone 60 mg/day). Two years ago, central nervous system (CNS) lupus was diagnosed after the appearance of convulsions, and the patient was started on phenytoin sodium. An MRI scan of the brain was non-specific. The patient had been treated with all these drugs, including the systemic steroids, until the current episode. There were no ocular problems until 1 day before his admission, when he suddenly noted blurred vision in his right eye. He was evaluated by an ophthalmologist and was diagnosed as having a central retinal vein occlusion in that eye. A few hours later he noted a second dramatic decrease in vision, and prompt evaluation revealed no light perception in his right eye. Examination of the anterior segment was normal and the lens was clear. Funduscopic evaluation (Fig. 1) revealed a swollen optic disc, arterial sheathing, scattered intraretinal haemorrhages in four quadrants and a cherry-red macular spot, establishing the diagnosis of combined central retinal artery and vein occlusion. Despite paracentesis and supplementation with intravenous steroids and anticoagulants, there was no improvement in the visual acuity of his right eye and the eye became amaurotic. The visual acuity of his left eye was 20/20, with a normal anterior segment and fundus during the episode.

Laboratory evaluation revealed abnormal titres of ANA (homogeneous pattern) and anti-single-stranded DNA antibodies, with low C3 and C4 complement levels. Antiphospholipid antibodies (detected by enzyme-linked immunosorbent assay) and lupus anticoagulants were normal, as were anti-double-stranded DNA antibodies, anti-thrombin III and protein S. Other laboratory results, including complete blood count, prothrombin time and partial thromboplastin time, were normal. The concentration of antiphospholipid antibodies remained normal after 8 months of follow-up.

The patient had a unilateral combined central retinal artery and vein occlusion. While reviewing the literature since 1966, using the Medline database, we found data on patients with lupus who had either isolated central retinal artery or vein occlusion, but the present case provides, to our knowledge, the first description of combined central retinal artery and vein occlusion. The only other descriptions of such a case concerned a lupus patient with elevated antiphospholipid antibodies [5] and a patient with severe bilateral retinal vasculitis with thrombosis of most of the retinal vasculature [6].

The facts that antiphospholipid antibody titres were normal and CNS lupus had been diagnosed previously in our patient suggests that the pathogenesis of the ocular disease was probably vasculitis. Jabs et al. [1] postulated that CNS lupus is a vasculitis that is identical to the vasculitis occurring in vaso-occlusive retinal lesions. A thrombotic event could not be ruled out, although there were no serological or haematological predisposing factors.

The distinction between thrombotic and inflammatory aetiology was very important during the acute phase of the episode because of the treatment options, which differ depending on whether immunological vasculitis or a thrombotic episode is the cause of the occlusion. If vasculitis is the cause, the patient should be offered the option of additional immune-suppressing treatment.
therapy. If a thrombotic episode is the cause of the occlusion, anti-thrombotic agents must be used. Because of the chronicity of the disease and the need to protect the remaining eye, treatment, once initiated, must continue for a long period. It seems possible that, in our patient, the arterial occlusion could have been secondary to the venous occlusion and raised intraocular pressure. After recovering from the acute episode, our patient was offered immune-suppressing therapy but refused it.

Regardless of the treatment chosen, patients with this condition should be followed up carefully in order to detect, as early as possible, any fundus changes in the second eye.

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