Sir,
Bilateral combined central retinal artery and vein occlusion in systemic lupus erythematosus resulting in complete blindness

Systemic lupus erythematosus (SLE) is a chronic autoimmune disorder that affects multiple organ systems, including the eye. Visual morbidity is usually due to retinal vascular manifestations of the disease. Severe vaso-occlusive retinopathy is a rare form of retinopathy often associated with poor visual prognosis. We report an unusual case of combined bilateral central retinal and vein-occlusion in a patient with SLE that resulted in complete blindness.

Case report

A 42-year-old Sudanese woman was referred to our department for profound bilateral visual loss. Six months before, she was diagnosed to have SLE on the basis of photosensitive erythematous maculopapular skin rashes with hair loss, polyarthrititis, autoimmune haemolytic anaemia, neutropenia, thrombocytopenia, and abnormal titres of antinuclear antibodies (ANAs) and antidouble-strand DNA (anti-dsDNA) antibodies. At the time of initial examination, visual acuity (VA) was no light perception in both eyes. The pupils were in mydriasis and not reactive to light. Fundus examination revealed pale optic discs and extensive and severe arteriolar

narrowing in both eyes. Superficial and deep intraretinal haemorrhages were scattered throughout the retina (Figure 1a and b). Fluorescein angiography showed severe ischaemia of both the macula and the peripheral retina (Figure 1c and d).

Laboratory examination confirmed the diagnosis of SLE and revealed autoimmune haemolytic anaemia with reticulocytosis, neutropenia, lymphopenia, and raised titres of ANAs, anti-dsDNA, antinucleoprotein (anti-Sm, anti-RNP, and anti-Ro/SSA) and perinuclear antineutrophilic cytoplasmic (p-ANCA) antibodies. Antiphospholipides antibodies (APAs), both anticardiolipin and lupus anticoagulants were negative. Serologies for syphilis, human immunodeficiency virus 1 and 2, hepatitis B and C, and haemocultures were all negative. Cerebrospinal fluid and urine analysis were within the normal limits. A magnetic resonance image of the central nervous system (CNS) was compatible with the diagnosis of CNS vasculitis.

Treatment was started with intravenous followed by oral prednisone and cyclophosphamide. Two months later, VA was still no light perception in the right eye and with light perception in the left eye (LE). Fundus examination showed persistent, extensive vaso-obliteration. Both arteries and veins appeared as yellow-white fibrotic cords, while the optic discs had become paler. A small tuft of neovascularisation over the disc surface was noted in the LE (Figure 2a and b).

Figure 1 Fundus colour photograph of the right (a) and left eye (b) of a patient with systemic lupus erythematosus showing bilateral and combined central retinal artery and vein occlusion. Note the extensive and severe arteriolar narrowing. The veins are thin with superficial and deep intraretinal haemorrhages scattered throughout the retina and around their walls giving them a beaded appearance. The arterioles supplying the macula are markedly narrowed and sheathed with no blood column in their lumina. A bull's-eye pattern is present in the macula of both eyes. (c and d) Fluorescein angiography shows severe ischaemia of the posterior pole and the peripheral retina. Note the widespread retinal capillary non-perfusion with dye arrest at about one to two disc diameters away from the disc. There is no dye filling of the retinal capillary bed beyond this area even at the very late phase of the angiography.

Figure 2 Fundus colour photograph of the right (a) and left eye (b) of the same patient 2 months later. Both arteries and veins appeared as yellow–white fibrotic cords free of blood with empty lumina, also called as ghost vessels. Optic discs are pale and atrophic. There is a small tuft of neovascularisation over the disc surface of the LE (black arrow).

Comment

Retinal vascular changes are frequently seen in patients with SLE.¹ The classic retinal findings are haemorrhages and cotton-wool spots and VA is generally unaffected. A rare form of retinopathy seen in patients with SLE is the so-called severe vaso-occlusive retinopathy. It is often associated with neovascularisation and poor visual prognosis.² An association usually exists between this severe retinopathy and APAs but in our case both lupus anticoagulant and anticardiolipin were negative.

Occlusions of larger vessels including isolated central retinal artery (CRAO) or vein (CRVO), and combined forms secondary to SLE have been reported in the literature and are usually unilateral.^{2–5} Although an uncommon event, combined CRAO and CRVO, has been reported in a variety of clinical settings, including in association with syphilis,⁶ Behçet's disease,⁷ cat-scratch disease,⁸ septic cavernous sinus thrombosis,⁷ infective endocarditis,⁷ temporal arteritis,⁹ Churg–Strauss syndrome,¹⁰ thrombotic thrombocytopenic purpura,¹¹ leukaemia and lymphoma,⁷ orbital inflammatory pseudotumor,¹² and posterior scleritis.¹³

Our case is atypical, because it combines central retinal artery and central retinal vein occlusion in both eyes of a patient with SLE without raised concentration of APAs leading to devastating bilateral visual loss, despite treatment. To the best of our knowledge, such a clinical picture has not been described previously and this case demonstrates that lupus retinopathy is a potential cause of visual morbidity.

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