Case report

SEVERE OCCLUSIVE RETINAL VASCULITIS IN SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

Paradee Kunavisarut, M.D., Kessara Pathanapitoon, M.D., Yodpong Chantarasorn, M.D., Janejit Choovuthayakorn, M.D., Aniki Rothova, Ph.D.

1Department of Ophthalmology, Faculty of Medicine, Chiang Mai University, 2Uveitis Centre, Department of Ophthalmology, University Medical Centre Utrecht, The Netherlands

Abstract

Severe vaso-occlusive retinopathy, as the primary manifestation of systemic lupus erythematosus (SLE), is relatively rare. We report a 30-year-old female, who gradually suffered bilateral visual loss. Fundus examination revealed bilateral severe occlusive retinal vasculitis and development of neovascularizations. Her clinical manifestations and laboratory findings were compatible with the diagnosis of systemic lupus erythematosus.


Keywords: occlusive retinal vasculitis, systemic lupus erythematosus, bevacizumab

Systemic lupus erythematosus (SLE) is a chronic systemic autoimmune disease, which may affect multiple organ systems. It is characterized by the production of pathological autoantibodies that lead to end-organ damage via inflammatory mechanisms. Keratoconjunctivitis sicca is the most common manifestation in ophthalmic lupus, while the posterior segment involvements have important implications for visual function. The retinal arterial occlusion is less common than office ocular complication. The presence of retinal manifestations parallels disease activity elsewhere. We, herein, describe a case of systemic lupus erythematosus, which who initially presented with bilateral severe vaso-occlusive retinopathy.

Case report

A previously healthy 30-year-old Thai female presented with gradual bilateral visual loss for 2 months. Visual acuity was finger counting in both eyes; retinal examination revealed severe occlusive vasculitis with diffuse sheathing of the arteries and development of neovascularizations located on both
optic discs. Multiple areas of retinal edema with hemorrhages and lipid exudates were present and all vessels exhibited gross caliber changes (Fig. 1). She had malar rash, discoid rash, oral ulcer and a history of photosensitivity, arthralgia, and alopecia for one year. She used no medications. Systemic lupus erythematosus was initially suspected, so she consulted a rheumatologist for further investigations. Complete blood count included hemoglobin at 8.3 g/dL without hemolytic blood features, and normal white blood cell and platelet count. Erythrocyte sedimentation rate (ESR) was 139 mm/hour. Urine analysis found trace proteinuria, without granular cast, and urine protein = 0.35 g per day. An antinuclear antibody test was positive at a titer of 1:1280. Renal and liver function tests were normal. The diagnosis of systemic lupus erythematosus was made following the American College of Rheumatology criteria. The results of serologic evaluation for HIV and syphilis were negative. Anticardiolipin antibodies, both Ig G and Ig M, were negative. Her blood pressure was normal. The patient was treated with systemic corticosteroid (1 mg/kg/day) and posterior subtenon injection with triamcinolone (20 mg/0.5 cc), panretinal laser photocoagulation and intravitreal injection of 1.25 mg of bevacizumab in both eyes. Within 6 weeks, retinal edema decreased and the majority of exudates and hemorrhages had been absorbed. Retinal neovascularizations disappeared in the right eye and were grossly reduced in the left one (Fig. 2). Optic disc pallor developed and arterial sheathing as well as extremely variable caliber of the retinal vessels persisted. In the left eye, adjacent to the optic disc, a development of fibrous tissue in previous neovascular membranes could be seen, which caused traction on the retina. Unfortunately, within one year, the neovascularizations of optic discs in both eyes reappeared. The patient was subsequently treated with extensive laser photocoagulation as well as intravitreal bevacizumab injections in both eyes. However, vitreous hemorrhage developed in both eyes, and pars plana vitrectomy was required to remove vitreous hemorrhage and release macular traction. During the surgery, additional endophoto-

Figure 1. Fundus photograph at the onset of the disease demonstrating severe occlusive vasculitis in systemic lupus erythematosus.
coagulation was also performed. Several months later, the neovascularizations of optic discs regressed. Her final visual acuity was 0.1 in both eyes.

**Discussion**

Retinal vascular lesions are one of the most common forms of intraocular involvement in patients with SLE, and present in 2-30% of all SLE patients, depending on the activity and severity of the disease. The retinopathy in SLE usually consists of cotton-wool spots, with or without intraretinal hemorrhage, and does not compromise visual acuity unless it is located in the macular area. A more severe variant of retinal vaso-occlusive disease is less common, and may present with localized features as central and branch retinal vascular occlusions or diffuse occlusive retinal disease, as in our patient. Clinicians should be aware of clinical pictures that resemble hypertensive retinopathy, which could be present secondary to lupus renal hypertension. Still, this patient had lupus retinopathy, which occurred as an independent manifestation of the underlying disease process in the absence of hypertension. Severe occlusive SLE retinopathy is associated with CNS involvement, which did not present in our patient however. The presence of antiphospholipid antibodies is mostly found when a search is made in patients with severe occlusive SLE retinopathy. However, the test was negative in our patient. The visual prognosis of diffuse occlusive retinal vascular disease is poor, and retinal neovascularizations with subsequent vitreous hemorrhage and tractional retinal detachment commonly develop. In our patient, visual acuity was slightly improved in both eyes after treatment. Systemic SLE treatment combined with scatter laser photocoagulation, and/or vitreoretinal surgery, contributed to the treatment of retinal neovascularizations. The role of anti-VEGF agents in the treatment of SLE retinal vasculopathy or further autoimmune diseases have not been systematically evaluated yet. We illustrated the promising role of anti-VEGF agents as an adjuvant in the treatment of retinal neovascularizations from severe occlusive retinal vasculitis.

In conclusion, this study demonstrated...
that SLE might initially manifest with ocular symptoms, which are potentially severe, and it also illustrated novel treatment possibilities with anti-VEGF agents.

References