Choroiditis in Systemic Lupus Erythematosus: Systemic Steroid Therapy and Focal Laser Treatment

Masahiko Shimura*, Yuko Tatehana*, Kanako Yasuda*, Shin-ichiro Saito†, Takeshi Sasaki† and Makoto Tamai*

*Department of Ophthalmology, Tohoku University, School of Medicine, Sendai, Japan;
†Department of Rheumatology and Hematology, Tohoku University, School of Medicine, Sendai, Japan

Purpose: To report a rare case of choroiditis in association with systemic lupus erythematosus (SLE).

Case: A 49-year-old woman with a 17-year history of SLE experienced acute vision impairment of her left eye during the remission stage of systemic SLE. Fundus examination revealed a gray-white subretinal exudate with serous retinal detachment. Angiographic examination disclosed choroidal inflammation at the macula and a breakdown of the blood retinal barrier. Retinal burns were applied to the subretinal exudate with an argon laser as in the treatment of central serous retinopathy. Afterward, her visual acuity showed prompt recovery due to the regression of the serous retinal detachment. However, the choroidal inflammation remained until the systemic condition was controlled with steroid therapy.

Results: Laser treatment of a subretinal exudate was helpful for the resolution of serous detachment and the prompt improvement of visual acuity, whereas systemic steroid therapy was effective for choroidal inflammation.

Conclusions: Systemic steroid therapy is thought to be effective for SLE choroiditis; however, this therapy is also known to cause serous retinal detachment. Thus, in SLE choroiditis, laser photocoagulation at a leakage point, in addition to systemic steroid therapy, may be helpful for the prompt restoration of vision in patients with serous retinal detachment.


Key Words: Choroidopathy, indocyanine green angiography, laser photocoagulation, steroid, systemic lupus erythematosus.

Introduction

Systemic lupus erythematosus (SLE) is a chronic, multisystem disease characterized by autoantibody formation and immune complex disease. Ocular manifestations of SLE are common; however, choroidopathy or choroiditis with serous detachment of the retina or pigment epithelium, or both, is less common and is usually seen in severely ill or hypertensive patients. There have been 28 cases of lupus choroidopathy reported in the English literature since 1968. All of the patients had active systemic vascular disease at the onset of their choroidopathy, and in most of the cases (23 cases; 83%) there was resolution of the choroidopathy when their systemic disease was brought under control. All of the cases had been treated with systemic steroid therapy. There has been little reported regarding steroid-induced chorioretinopathy or regarding the effectiveness of local treatment such as photocoagulation.

In this case, choroiditis with serous retinal detachment was seen in an SLE patient and laser treatment resulted in successful resolution of the serous detachment and complete recovery of visual acuity within a couple of days. SLE choroidopathy persisted longer, clearly shown by indocyanine green angiography (ICGA), and gradually improved with control of the underlying systemic SLE condition.

Case Report

A 49-year-old woman with a 17-year history of SLE was admitted to the rheumatic disease unit of our hospital...
because of deterioration of her condition. At the time of admission, she had a fever (37.5°C) and numbness of the lower limbs. A physical examination showed 90/48 mm Hg blood pressure, slight edema in the dorsum of both hands, but no arthritis, skin eruption, or neurological abnormalities. A serological examination showed a positive antinuclear antibody and anti-double-strand DNA antibody (dsDNA). Results of laboratory tests included the following: total protein 6.3 g/dL, albumin 3.0 g/dL, total bilirubin 0.5 mg/dL, γ-glutamyl transpeptidase 22 IU/L, glutamic oxaloacetic transaminase 39 IU/L, glutamic pyruvic transaminase 25 IU/L, lactic acid dehydrogenase 842 IU/L, cholinesterase 98 IU/L, blood urine nitrogen 8 mg/dL, creatinine 0.6 mg/dL, white blood cells 3500/µL, red blood cells 3.99 × 10¹²/µL, platelets 72,000/µL, hemoglobin 10.6 g/dL. High serum amyloid A (SAA) (301 µg/mL), positive anti-dsDNA antibody (400 IU<), and low 50% hemolytic unit of complement serum (CH50) (10.5 U/mL) indicated active SLE.

The patient was treated with prednisone (40 mg/day), and subsequent steroid therapy was gradually reduced. Three months after hospitalization, she noticed a sudden haziness in her left eye. At that time, 20 mg/day of systemic steroid was still being administered. Her fever, numbness of the lower limbs, and dorsal hand edema all had disappeared. On general examination, SAA (8>) and dsDNA (10>), had returned to a normal range except for the relatively low value of CH50 (19.1). On ophthalmological examination, her best corrected visual acuity was 20/20 OD and 20/200 OS; and intraocular pressure was 16 mm Hg in both eyes. Ophthalmoscopy disclosed focal shallow serous elevations of the sensory retina without cotton-wool spots or hemorrhages (Figure 1A), and a small area of gray-white subretinal exudate at the upper edge of the serous detachment (Figure 1A, arrow). Fluorescein angiography (FA) in her left eye revealed early leakage at the subretinal exudates and serous elevation of the sensory retina was clearly seen in the late phase.

Figure 1. (A) Fundus photograph of the left eye of a 49-year-old woman with a 17-year history of systemic lupus erythematosus (SLE) shows a gray-white subretinal exudate (arrow). (B) Fluorescein angiography shows leakage from the gray-white subretinal exudate and a large retinal serous detachment including the fovea. (C) Indocyanine green angiography (ICGA) in early phase reveals small hypofluorescent lesions in the fovea (arrows) and at the gray-white lesion. (D) ICGA in late phase shows small hyperfluorescent lesions in the fovea (arrows) and at the gray-white lesion.
(Figure 1B). At the subretinal exudates, ICGA detected hypofluorescent lesions in the early phase, and hyperfluorescence in the late phase. ICGA also disclosed early hyp- and late hyperfluorescent lesions around the fovea (Figure 1C and D, arrows). These indicated early circulation disturbance and late increased permeability at the choriocapillaries and retinal pigment epithelium (RPE). For better control of active SLE, 50 mg/day of cyclophosphamide was added and the dosage of systemic steroid was increased to 40 mg/day. During the subsequent 2 weeks of observation, CH50 had ameliorated to over 20.0; however, the area of serous detachment increased while the visual acuity of her left eye decreased to 20/400. Because of the decreasing visual acuity and apparent lack of beneficial effect of systemic therapy on the retina, laser photocoagulation (argon laser 130 mW, 0.15 seconds, 150 µm spot size, 28 spots) was performed on the gray-white subretinal exudate. Two days later, serous detachment had regressed and the corrected visual acuity of her left eye was restored to 20/20 (Figure 2A), although the scattered hyperfluorescent lesions in the late phase of ICGA remained (Figure 2B, arrows). Subsequently, cyclophosphamide was discontinued and systemic steroid treatment was reduced. One year after that, the systemic condition was well under control without prednisone administration. In addition, the choroidal foci seen in ICGA had healed (Figure 3).

**Discussion**

Ocular manifestations of SLE are common and include the following: (1) lid involvement by mucocutaneous disease; (2) anterior segment findings including keratoconjunctivitis sicca, conjunctivitis, episcleritis, scleritis, keratitis, and iridocyclitis; (3) posterior segment abnormalities including asymptomatic microvascular changes, cotton-wool spots, intraretinal hemorrhages, retinal artery and vein occlusions, retinal neovascularization, and vitreous hemorrhages; and (4) neuro-ophthalmic lesions with optic neuritis, papillitis and papilledema. The retinopathy with SLE generally consists of cotton-wool spots with or without retinal hemorrhages. Even less common are reports of choroidopathy with serous detachment of the retina or pigment epithelium or both.

In this case, FA showed serous retinal detachment and ICGA clearly revealed focal choroidal inflammation in association with SLE. Several mechanisms of serous detachment (serous detachment of pigment epithelium) can be involved in such cases. First, there is the possibility of increased permeability of the choroidal or retinal pigment epithelium, resulting from systemic vasculitis. Second, choroidal inflammation may cause leakage and serous detachment. In the present case, serous detachment of the pigment epithelium had healed, indicating that choroidal inflammation had resolved, and the patient's visual acuity had improved. However, the scattered hyperfluorescent lesions in the late phase of ICGA remained (Figure 2B, arrows).
detachment in SLE can be considered: (1) corticosteroid-induced RPE dysfunction, (2) choroidal vasculopathy causing ischemia-induced RPE dysfunction, or (3) choroidal inflammation-induced RPE dysfunction. The decrease in visual acuity was mainly due to serous retinal detachment, because laser treatment brought a quick regression of the serous detachment and a subsequent dramatic improvement of visual acuity without apparent relation to systemic SLE activity. Therefore, it was strongly suspected that the focal inflammatory lesion itself was the cause of the retinal detachment, and not widespread vasculopathy. SLE choroiditis around the fovea found in ICGA resolved slowly at the remission of systemic SLE activity. Even after the resolution of serous retinal detachment and visual acuity, ICGA still revealed some early hypo- and late hyperfluorescent lesions around the fovea. These indicated that choroidal foci still existed with no relation to visual acuity. During the 1 year of follow-up examinations, the foci gradually regressed with time and finally disappeared. Thus, further observation of choroidal circulation with the use of ICGA, even after complete recovery of visual acuity in SLE choroiditis, is recommended.

The exact pathogenetic mechanism of SLE choroiditis remains unclear. Histopathologic studies have shown the existence of inflammatory cells, deposits of immunoglobulin, and complement in choroidal vessels. Therefore, SLE choroiditis is thought to be caused by focal inflammation at the choriocapillaris, and subsequent focal vascular compromise results in ischemia in overlying RPE cells, which causes a breakdown of the normally impermeable outer blood—ocular barrier provided by this layer. Therefore, systemic corticosteroid therapy is believed to be an effective treatment. Some evidence suggests, however, that the systemic application of corticosteroid can result in choroidal hyperpermeability, which may lead to a breakdown of the blood—retinal barrier or alter RPE ion transport capabilities, and cause serous retinal detachment. During the clinical course of active SLE, systemic steroid therapy is thought to have two opposite effects. On the one hand, systemic steroid therapy helps the resolution of choroidopathy by reducing the inflammation of systemic vasculitis. On the other hand, it accelerates a breakdown of the blood—retinal barrier and leads to serous retinal detachment. In this case, it is likely that both effects were observed; resolution of choroidal inflammation and aggravation of inflammatory blood—retinal barrier damage inducing serous retinal detachment. According to reported cases, adequate control of the systemic condition by systemic corticosteroid therapy with or without immunosuppressive agents sometimes fails to induce a complete resolution of serous retinal detachment. Therefore, the present case of SLE choroiditis, which was refractory to systemic corticosteroid therapy, indicates a preference for performing focal laser treatment for accurate resolution of serous retinal detachment and restoration of visual acuity.

The authors thank Dr. Yasuhiko Hirabayashi for helpful comments and clinical assistance, and Mr. Kurt Schelbner and Ms. Betty Parker for critically reading the manuscript and correcting the English.

References