A case of severe choroidal detachment in both eyes due to systemic lupus erythematosus

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ABSTRACT

Purpose: We report a case of severe choroidal detachments (CDs) in both eyes caused by systemic lupus erythematosus (SLE).

Observations: The patient was a 50-year-old woman who presented with conjunctival edema in both eyes, visual dysfunction, and generalized fatigue. At the first visit, the best corrected visual acuity (BCVA) was 20/70 OD and 20/70 OS, and the intraocular pressure (IOP) was 22 mmHg OD and 27 mmHg OS. She had serous retinal detachments (SRDs), CDs, ciliary dissections, and a shallow anterior chamber with partial angle closure in both eyes. Systemic findings included hypoalbuminemia, pleural fluid, generalized fatigue, and brown papules on the back and both legs. First, we suspected Vogt-Koyanagi-Harada disease and administered two courses of methylprednisolone pulse therapy, but the CDs in both eyes gradually deteriorated and worsened to the extent that the optic nerve in both eyes could not be observed, and the BCVA deteriorated to 20/200 OD and 6/200 OS. Further multidisciplinary evaluations for diagnosing collagen diseases revealed vasculitis in the skin histopathology examination, positive results for anti-double stranded DNA antibody and anti-SS-A antibody, and hypo-complementemia in the blood examination, and she was diagnosed with severe SLE in the dermatology department. After administration of high dose intravenous γ-globulin therapy, albumin infusion, and intravenous cyclophosphamide pulse therapy, the SRDs and severe CDs improved along with improvement in hypoalbuminemia, pleural fluid, and generalized fatigue. Moreover, the shallow anterior chamber and high IOP improved to normal in both eyes. The CDs and SRDs completely disappeared, and the BCVA improved to 20/13 OU 6 months after the SLE therapy.

Conclusion and importance: In patients with observed SRDs and CDs accompanying hypoalbuminemia, it is necessary to consider collagen diseases such as SLE.

1. Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease characterized by systemic inflammatory lesions that cause damage to various organs by tissue deposition of immune complexes on the vascular walls. SLE commonly begins between 20 and 40 years of age and shows a male-to-female ratio of 1:10, with a greater predilection among younger women. Initial symptoms in most patients are arthrits, skin findings such as hot flashes and erythema, fever, and malaise. In SLE patients, ocular involvement is common (prevalence up to 30%) and extremely varied. The ocular lesions in SLE patients are mainly reported to occur in the retina, optic nerve, uvea, and conjunctiva, and approximately 25% of ocular lesions are retinal lesions. Typical retinal findings in SLE include retinal hemorrhages, white spots, retinal vasculitis, serous retinal detachment, and choroidal detachment (CD). There have been a report about SLE patients with mild CD, but no reports about SLE patients with severe CD. Here, we report a case of severe CD in both eyes due to SLE that caused remarkable visual dysfunction and was difficult to diagnose and treat.

2. Case report

The patient was a 50-year-old female beautician. She had eczema on the dorsum due to perming fluid and was diagnosed with self-sensitizing
Mild CDs were observed on the inferotemporal fundus (Fig. 1C and D), the anterior chamber, lens opacity, or vitreous opacity was not observed. In addition, ciliary dissections were observed all around. Inflammation in the anterior chamber, lens opacity, or vitreous opacity was not observed. In addition, ciliary dissections were observed all around. Inflammation in the anterior chamber, lens opacity, or vitreous opacity was not observed.

At the first visit, her best-corrected visual acuity (BCVA) was 20/70 OD and 20/70 OS, and the intraocular pressure (IOP) was 22 mmHg OD and 27 mmHg OS. Conjunctival edema and shallow anterior chambers with partial angle closure were observed in both eyes (Fig. 1A and B). In addition, ciliary dissections were observed all around. Inflammation in the anterior chamber, lens opacity, or vitreous opacity was not observed. Mild CDs were observed on the inferotemporal fundus (Fig. 1C and D), and optical coherence tomography (OCT) revealed SRDs in the macular area in both eyes (Fig. 1E and F). There was no optic nerve abnormality, retinal hemorrhage, or exudate. Scleral thickening and obvious short axis were not observed on B-mode ultrasound (Fig. 2). Furthermore, she presented with general malaise and depression, and pigmentation and brown papules were noted on the back and both legs (Fig. 3A and B). Cerebrospinal fluid examination by lumbar puncture was not performed because she did not wish to undergo the procedure. On the basis of these findings, we first diagnosed Vogt-Koyanagi-Harada disease and started one course of methylprednisolone pulse therapy (1000 mg/day for 3 days). Following the pulse therapy, 1 mg/kg/day prednisolone was administered orally. After the treatment, the SRDs and the CDs did not improve and the patient’s general fatigue worsened. Hematological examination showed hypoalbuminemia and negative findings for human leukocyte antigen-DR4 (Table 1). Computed tomography (CT) of the chest and abdomen showed massive thoracoabdominal fluid and intestinal edema (Fig. 4). On the 12th day of hospitalization, the CDs in both eyes worsened, and the optic nerve of the left eye became difficult to observe (Fig. 5). Fluorescein angiography (FA) and indocyanine green angiography showed granular hyperfluorescent lesions, mild leakages, and partial hypofluorescent lesions, suggesting choroidal circulation insufficiency, but not obvious retinal vasculitis (Fig. 6). The patient received the second course of methylprednisolone pulse therapy (1000 mg/day for 3 days). However, there was no improvement, the CDs worsened, the optic nerve of both eyes could not be observed, and the BCVA deteriorated to 20/200 OD and 6/200 OS (Fig. 7). In order to confirm the presence of other diseases such as connective tissue disease, malignant tumor, and digestive system disease, we consulted other departments. Subsequently, inflammatory cell infiltration of lymphocytes around the capillary walls in the superficial dermis was observed as vasculitis on skin histopathology examination.

(Fig. 8), and positive results for anti-double stranded DNA antibody and anti–SS-A antibody, and hypocomplementemia were observed in the blood test in the dermatology department (Table 2). In addition, gastrointestinal scintigraphy revealed accumulation in the stomach, which led to the diagnosis of protein-losing gastroenteropathy in the gastroenterology department (Fig. 9). There were no findings suggesting malignant tumors in the systemic screening test. Based on the above results, the patient was diagnosed with SLE because four of the clinical findings and two of the immunological items in the Systemic Lupus International Collaborating Clinics classification criteria for SLE were recognized (Table 3). Furthermore, the SLE Disease Activity Index 2000 score for assessing the severity was 16 points, corresponding to severe (Table 4). On the 16th day of hospitalization, gamma globulin (20 g/day for 5 days) was infused intravenously for the treatment of severe SLE. On the 21st day of hospitalization, an albumin preparation (12.5 g/day for 3 days) was also infused intravenously, after which the pleural fluid, hypoalbuminemia, and general fatigue gradually improved. In addition, the CDs and SRDs gradually improved. On the 33rd and 60th days of hospitalization, intravenous cyclophosphamide pulse therapy (750 mg/day for 1 day) was performed, and the CDs and SRDs further improved. On the 41st day of hospitalization, the CD in the left eye disappeared. Although CD in the right eye and mild SRDs in both eyes were observed, she was discharged on the 48th day of hospitalization because her systemic condition improved and CT showed the disappearance of the thoracoabdominal fluid. Thereafter, the SRDs and CDs in both eyes completely disappeared on the 26th day after discharge (Fig. 10). Moreover, the shallow anterior chamber with partial angle closure improved and the IOP was normalized to 10 mmHg in both eyes. At present (24 months after discharge), she is being followed up in an outpatient setting, taking 5 mg of prednisolone, and is showing good progress with an improvement in her BCVAs to 20/13 in both eyes without recurrence of systemic and ocular symptoms.

**Fig. 1.** Anterior segment and fundus findings at the first visit (A, C, and E: Right eye, B, D, and F: Left eye). Conjunctival edema and a shallow anterior chamber with partial angle closure in anterior segment optical coherence tomography (OCT) were observed in both eyes (A and B). The color fundus photograph shows partial choroidal detachments (CDs) on the inferotemporal fundus in both eyes (red arrows, C and D). The macular OCT revealed subretinal detachments (SRDs) in the macula in both eyes (E and F). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)
3. Discussion

According to the report by Tamiya et al., 7 10 of 40 SLE patients showed accompanying chorioretinopathy, with only retinal hemorrhages and cotton wool spots in 4 cases, SRDs in 3 cases, and retinal vasculitis, central retinal vein occlusion, bullous retinal detachment, and acute retinal necrosis in 1 case each. Furthermore, the higher the activity of SLE, the greater was the frequency of comorbidities of chorioretinopathy, and 5 of the 7 cases were considered to be highly active and showed comorbidities of chorioretinopathy. Matsumoto et al. reported one SLE patient with mild CD; this patient had already been diagnosed with SLE, and the CDs occurred while taking 50 mg of steroid. 8 In systemic examinations, pleural fluid was recognized by CT and

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**Table 1**

| Variable                              | Result | Reference range, adults |
|---------------------------------------|--------|-------------------------|
| White blood cell (WBC)                | 5600   | 3300-10700 (/μL)        |
| Lymphocyte                            | 8      | 26.8–43.8 (%)           |
| Red blood cell (RBC)                  | 511    | 353-466 (/μL)           |
| Hemoglobin                            | 13.6   | 12.1–15.0 (g/dL)        |
| Platelets                             | 232    | 150-400 (bil/L)         |
| Urea nitrogen                         | 11     | 8.0–22.0 (mg/dL)        |
| Creatinine                            | 0.56   | 0.60–1.40 (mg/dL)       |
| Total protein                         | 6.5    | 6.3–8.0 (g/dL)          |
| Albumin                               | 2.4    | 3.9–4.9 (g/dL)          |
| Aspartate aminotransferase (AST)      | 68     | 10-40 (U/L)             |
| Alanine aminotransferase (ALT)        | 37     | 5-40 (U/L)              |
| Lactate dehydrogenase (LDH)           | 327    | 100-238 (U/L)           |
| Total bilirubin                       | 0.4    | 0.3–1.2 (mg/dL)         |
| C-reactive protein (CRP)              | 0.62   | 0.0–0.8 (mg/dL)         |
| Human leukocyte antigen-DR4 (HLA-DR4) | –      | –                       |
| Urinalysis of protein                 | 0.4    | 0 (mg/dL)               |
| Urinalysis of blood                   | –      | 0                       |

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**Fig. 2.** B-mode ultrasound images. B-mode ultrasound images did not show obvious short axis length or scleral thickening (A: right eye, B: left eye).

**Fig. 3.** Cutaneous symptoms at the first visit. Pigmentation and brown papules were noted on the back (A) and both legs (B). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

**Fig. 4.** Computerized tomography (CT) of the chest (A) and abdomen (B). CT shows massive thoracoabdominal fluid (asterisks, A and B), pleural thickening (arrowheads, A), and intestinal edema (arrowheads, B).
Fig. 5. Color fundus photography (CHP) and macular optical coherence tomography (OCT) images after the first pulse steroid therapy (A: right eye, B: left eye). The CHP images show severe choroidal detachments (CDs) around the fundus in both eyes (uppers, A and B). The macular OCT images reveal subretinal detachments (SRDs) in the macula area in both eyes (lowers, A and B). The SRDs in both eyes slightly decreased compared to those at the first visit, but the CDs worsened in both eyes at the first visit.

Fig. 6. Fluorescein angiography (FA) and indocyanine green angiography (ICGA) images (A: right eye, B: left eye, early-phase images: uppers, late-phase images: lowers). Both FA and ICGA images showed granular hyperfluorescent lesions and mild leakages, and partial hypofluorescent lesions suggesting choroidal circulation insufficiency (asterisks, A and B), but not obvious retinal vasculitis in both eyes. In addition, ICGA images had poor visualization of choroidal vessels, and no obvious leakage and pooling from the choroidal vessels. In both eyes, the areas of choroidal detachment were hypofluorescent due to the block.

Fig. 7. The color fundus photograph (CHP) and macular optical coherence tomography (OCT) images after second pulse steroid therapy (A: right eye, B: left eye). The CHP images showed severe choroidal detachments all around the fundus such that the optic nerve of both eyes could not be observed, and the best-corrected visual acuity deteriorated to 20/200 OD and 6/200 OS (uppers, A and B). The macular OCTs reveal subretinal detachments in the macula area in both eyes (lowers, A and B).
a decrease in total serum protein level was also observed in the blood test. The CDs disappeared after one course of steroid pulse therapy (0.5 g/day for 3 days) and the intravenous vasodilator lipoprostaglandin E1, and visual acuity had also improved. On the other hand, the SLE patient described in this report had severe CDs in both eyes and the CDs worsened so much that the optic nerves of both eyes could not be observed. In addition, this patient was resistant to steroid pulse therapy, and it was difficult to make a diagnosis and treat.

In the dermatology examination, although the result for antinuclear antibody was negative, this patient could be diagnosed as having SLE because 4 or more criteria (at least 1 clinical and 1 immunologic criteria) of the SLE diagnostic criteria were met.

The SLE patient was diagnosed as showing SLE because four of the clinical findings and two of the immunological items in the classification criteria 2012 of SLE were recognized.

Table 2
The results of hematological examination.

| Variable                        | Result          | Reference range, adults |
|---------------------------------|-----------------|-------------------------|
| Antinuclear Antibodies titer    | <1:40           | <1:160                  |
| Anti-double stranded DNA        | 19              | <10.0 (IU/mL)           |
| Anti Sm                         | <5              | <7 (U/mL)               |
| Anti SS-A (ELISA)               | 87.4            | <10 (U/mL)              |
| Complement C3                   | 50              | 80-200 (mg/dL)          |
| Complement C4                   | 3               | 12-45 (mg/dL)           |

Table 3
The systemic lupus erythematosus (SLE) classification criteria 2012.

SLICC criteria
Clinical criteria Immunologic criteria

| Acute cutaneous lupus | Antinuclear Antibody |
|-----------------------|-----------------------|
| Chronic cutaneous lupus| Anti-double stranded DNA |
| Nonscarring alopecia  | Anti Sm               |
| Oral or nasal ulcers   | Antiphospholipid       |
| Joint disease          | Low complement         |
| Serositis              | Direct Coombs test (in the absence of hemolytic anemia) |

Renal

| Neurologic |
|------------|
| Hemolytic anemia |

Leukopenia or

| Lymphopenia |
|-------------|

| Thrombopenia |
|--------------|

The patient was diagnosed as showing SLE because four of the clinical findings and two of the immunological items in the classification criteria 2012 of SLE were recognized.

Table 4
The systemic lupus erythematosus disease activity index 2000 (SLEDAI-2K).

| SLEDAI-2K |
|-----------|
| Criteria  | Weight | Criteria  | Weight |
|-----------|--------|-----------|--------|
| Recent onset seizure | 8 | Proteinuria | 4 |
| Psychosis | 8 | Pyuria | 4 |
| Organic brain syndrome | 8 |  | |
| Visual disturbance     | 8 | Inflammatory-type rash | 2 |
| New onset sensory or motor neuropathy involving cranial nerves | 8 | Alopecia | 2 |
| Lupus headache          | 8 | Oral or nasal mucosal ulcers | 2 |

Fever >38 °C |

| Heme-granular or RBC urinary casts | 4 | Platelets <100 × 10^9/L | 1 |
| Hematuria                            | 4 | WBC <3 × 10^9/L | 1 |

The SLEDAI-2K stratifies the severity of systemic lupus erythematosus (SLE). In this patient, the SLEDAI score was 16 points, corresponding to severe.

because 4 or more criteria (at least 1 clinical and 1 immunologic criteria) of the SLE diagnostic criteria were met. In this patient with SLE, antigen-antibody complexes such as immunoglobulin and complement
may have been deposited on various capillary walls, causing systemic vascular inflammation and vascular hyperpermeability and hypoalbuminemia. In addition, one of the systemic symptoms of SLE is gastrointestinal symptoms; especially, protein leakage-type gastroenteropathy causes protein leakage in the intestine. In this SLE patient, the protein leakage-type gastroenteropathy was also possibly one of the factors that exacerbated hypoalbuminemia. Severe CD was presumably caused by the leakage of plasma components from the choroidal vessels after the decrease in plasma oncotic pressure due to prominent hypoalbuminemia. The same mechanism can explain the appearance of prominent pleural fluid. and Aronson et al.

In this SLE patient, a similar change in the whole-organ vasculitis caused by deposition of the antigen-antibody complex may have occurred chiefly on the choroidal vessel walls. Thus, the choroidal vessel cavity was possibly occluded, which caused SRDs and severe CDs. In other words, the SRDs and severe CDs in this patient were directly attributable to the deposition of antigen-antibody complexes on the choroidal vessels by SLE and indirectly attributable to factors such as hypoalbuminemia due to systemic vasculitis and protein leakage gastroenteropathy.

In this patient, high IOP due to the shallow anterior chamber with partial angle closure was observed in both eyes from the first visit. Kobashigawa et al. reported an SLE case with a shallow anterior chamber, and noted that inflammation of the ciliary body was the pathogenic mechanism. In autopsy reports on the eyes of SLE patients, immune complex deposits were observed in the ciliary epithelium, conjunctival epithelium, and choroidal microvessels, suggesting cellular damage in these parts. In addition to the mechanism described above, prominent hypoalbuminemia, and a decrease in plasma oncotic pressure due to protein leakage-type gastroenteropathy associated with SLE might cause movement of plasma components from the blood vessels to the ciliary body tissue. Ciliary body detachment and the shallow anterior chamber with angle closure might have occurred. We think that the onset of the shallow anterior chamber with angle closure in this patient was caused by a similar mechanism as the occurrence of SRDs and severe CDs.

Wang et al. previously reported an SLE patient with uveal effusion diagnosed as no ocular findings of short axis length or scleral thickening, but showing SRDs with retinal folds and hyperfluorescent leakage from the peripheral retinal vessels on FA. In addition, intravenous administration of steroids and azathioprine for the treatment of SLE improved systemic symptoms and ocular findings. We think the uveal effusion in the patient reported by Wang et al. was also caused by the same mechanism as that in the SLE patient we have described in this report. However, the SLE patient we reported had more severe CDs and did not improve by steroid pulse therapy; thus, it is presumed that the condition of this patient was more severe. Similar to the SLE patient Wang et al. reported, the patient we describe in this report did not show obvious short axis length or scleral thickening, and administration of high dose of intravenous γ-globulin therapy, albumin infusion, and intravenous cyclophosphamide pulse therapy for the treatment of SLE improved CDs along with the improvement of the general condition. Therefore, we think the severe CDs in the SLE patient we have described in this report are not idiopathic uveal effusion, but occurred secondary to SLE. There have been a few previous reports of patients with chorioretinopathy due to SLE, such as uveal effusion and mild CD, but there have been no reports of patients with severe CD due to SLE; therefore, the diagnostic treatment was difficult.

4. Conclusions

In this report, we present a patient with severe CDs in both eyes due to SLE that caused remarkable visual dysfunction. In patients showing SRDs and CDs accompanying hypoalbuminemia, it is necessary to consider collagen diseases such as SLE and cooperate with other departments immediately.

Patient consent

We hereby acknowledge that the patient provided written informed consent for reporting the examination and imaging findings as deemed necessary for diagnosis, education, research, and quality improvement.

Authorship

All authors attest that they meet the current ICMJE criteria for authorship.
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Declaration of competing interest

We confirm that there is no known interest associated with this publication, and there has been significant support for this work.
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