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CASE REPORT

Toshiyuki Bohgaki · Masaya Mukai · Atsushi Notoya
Makoto Kondo · Michifumi Kohno · Utako Okushiba

Transient myopia with severe chemosis as an initial manifestation of systemic lupus erythematosus

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Abstract A 24-year-old woman suffered from blurred vision and periorbital edema with remittent fever. She was diagnosed as having systemic lupus erythematosus (SLE), complicated with myopia and retinopathy and severe chemosis. Antiphospholipid syndrome (APS), hemophagocytic syndrome, and liver involvement were also proven. We considered that APS might cause chemosis as a result of thrombosis-induced perfusion failure in the conjunctiva. In such cases, APS should be considered and anticoagulation therapy associated with steroid therapy should be initiated. In systemic lupus erythematosus (SLE), chemosis, severe hepatitis, and hemophagocytic syndrome (HPS) are rare complications. It is well known that many cases of SLE are complicated with antiphospholipid syndrome (APS), which causes arteriovenous thrombosis. We report a case of SLE with transient myopia and severe chemosis complicated with severe hepatitis and HPS. As this patient had antiphospholipid antibodies, these ocular complications were considered to be related to APS.

Key words Antiphospholipid syndrome · Chemosis · Hemophagocytic syndrome · Myopia · Systemic lupus erythematosus

Case report

A 24-year-old pregnant woman had suffered from blurred vision and periorbital edema since February 1999. She had been well until these symptoms appeared. On 15 March, she had a planned abortion because of her poor general condition. Two days after the abortion, she was admitted to a local hospital because of a remittent fever and edema of her legs. Liver involvement and proteinuria were noted. She was referred to our hospital on March 19. Her temperature was then 39.0°C. She had bilateral periorbital edema with severe chemosis, malar rash, and mild edema of her legs. There was no physical abnormality in her chest, abdomen, or joints.

The following results were obtained on ophthalmological examination. The bulbar conjunctivas were markedly chemotic, and the anterior chambers were shallow bilaterally. Ocular motility was normal. The corrected visual acuity was 20/18 right eye (−3.0 D), and 20/24 left eye (−3.5 D). There was no inflammation, and her bilateral applanation tensions were normal. Ophthalmoscopy revealed no evidence of edema or detachment of choroid or retina in either eye. Funduscopy revealed bilateral cotton-wool spots and intraretinal hemorrhages (Fig. 1). Fluorescent angiography revealed occluded capillary vessels, leakage, and blockage of background fluorescence due to intraretinal hemorrhage.

Laboratory tests indicated microcytic hypochromatic anemia (hemoglobin 9.5 g/dl), leukopenia (1900/mm^3), and thrombocytopenia (10.4×10^4/mm^3). Hypoalbuminemia (albumin 2.8 g/dl), liver involvement (aspartate aminotransferase 139 IU/l; alanine transaminase 115 IU/l; lactate dehydrogenase 953 IU/l), and urinary abnormalities (proteinuria 2.2 g/day and microhematuria) were detected. Serum creatinine (1.1 mg/dl) and erythrocyte sedimentation rate (120 mm/h) were high. The work-up demonstrated negative serologies for hepatitis B and C, cytomegalovirus, and Epstein–Barr virus. Antinuclear antibodies (1:640 with a homogeneous and speckled pattern), antidouble-strand DNA (anti-DNA) antibodies (63.1 IU/ml), lupus anticoagulant (aPTT method 8.7″; normal range less than 4.5″), and both IgG (4.9; normal range less than 1.0) and IgM (1.0; normal range less than 1.0) isotypes of anticardiolipin antibodies were positive. Other autoantibodies such as antismooth muscle antibody or antimitochonrial antibody were negative. Platelet-associated IgG (1013.5 ng/10^7 cells),

T. Bohgaki · M. Mukai (✉) · A. Notoya · M. Kondo · M. Kohno
Division of Clinical Immunology and Hematology, Department of Medicine, Sapporo City General Hospital, Kita 11-jo, Nishi 13-chome, Chuo-ku, Sapporo 060-8604, Japan
Tel. +81-11-726-2211; Fax +81-11-726-9541
e-mail: masaya-mukai@hokkaido.med.or.jp

U. Okushiba
Department of Ophthalmology, Sapporo City General Hospital, Sapporo, Japan

ferritin (11 815 ng/ml), soluble Fas (sFas, 1.829 ng/ml), soluble Fas ligand (sFasL, 0.579 ng/ml), and tumor necrosis factor α (TNFα, 102 pg/ml) were high. Low complement levels (C3 33.5 mg/dl; C4 3.0 mg/dl; CH50 <11.0 U/ml) were detected. The patient's bone marrow showed hemophagocytosis. The titer of anti-DNA antibodies decreased from 63.1 IU/ml to 49 IU/ml in just 1 day without any medication during the deterioration of liver function (Table 1). The disease activity of her SLE was obviously elevated, and the SLE disease activity index (SLEDAI)[1] was thirty-seven.

The patient was diagnosed as having SLE complicated with myopia due to chemosis. Prednisolone (60 mg/day) was administered following methylprednisolone pulse therapy, and subcutaneous heparin and low-dose aspirin were given. Following these therapies, her periorbital edema, chemosis, myopia, and serological abnormalities rapidly subsided, and her visual acuity returned to 40/20 in the right eye and 40/20 in the left without correction (Table 1).

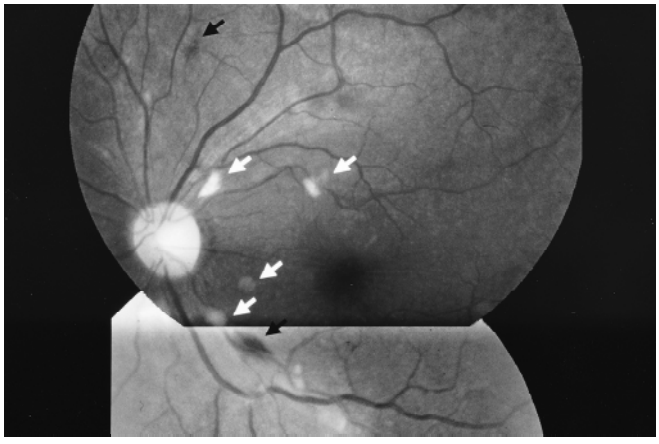

Fig. 1. "Classic" type of lupus retinopathy characterized by cotton-wool spots (*white arrows*) with intraretinal hemorrhages (*black arrows*) in the left eye

Discussion

The incidence of retinopathy in SLE is reported to range from 10% to 20%.[2–4] It has been noted that at least two major clinical forms of retinopathy occur in SLE: the "classic" type characterized by cotton-wool spots, with or without intraretinal hemorrhages, and "Hughes' retinopathy," characterized by the occlusion of retinal vessels by thrombosis related to APS.[5,6] Montehermoso et al.[4] reported that retinal vascular disease was detected in 13 of 82 SLE patients, and that antiphospholipid antibodies were positive in 10 of these 13 patients. Retinopathy, which is relatively rare as an initial manifestation, is related to the activity and duration of the SLE.[5,6] In our patient, transient myopia with the "classic" type of retinopathy, characterized by cotton-wool spots with intraretinal hemorrhages and bilateral severe chemosis, was confirmed. Fluorescent angiography revealed occluded capillary vessels, indicating that her retinopathy might be combined with "Hughes" retinopathy.

Transient myopia is associated with local conditions such as contusion of the eye, inflammation, medication, diabetes mellitus, and the nephrotic syndrome. At least two mechanisms of transient myopia are hypothesized. The axial eyeball length may be increased owing to (i) increasing vitreous volume as a result of decreased colloidal osmotic pressure

Table 1. Clinical course

	Normal range	1999						
		19 March	23 March	24 March	1 April	8 April	22 April	20 May
Prednisolone (mg/day)[a]		–	–	–	60	60	60	40
VA (RE) uncorrected		20/200	ND	ND	40/20	40/20	24/20	30/20
VA (RE) corrected		(18/20 × −3.0 D)	ND	ND	–	–	–	–
VA (LE) uncorrected		14/200	ND	ND	40/20	40/20	24/20	24/20
VA (LE) corrected		(24/20 × −3.5 D)	ND	ND	–	–	–	–
AST (IU/l)	5–40	139	195	240	47	26	24	23
ALT (IU/l)	0–46	115	123	135	95	43	33	38
LDH (IU/l)	180–410	953	2003	2545	728	494	434	584
Al-P (IU/l)	78–362	135	139	209	235	227	244	238
γ-GTP (IU/l)	7–77	42	60	86	257	114	70	47
anti-DNA (IU/ml)	<6	ND	63.1	49	ND	33	38	26
CH50 (U/ml)	30–45	ND	<11.0	<11.0	<11.0	16.4	32.6	41.0
C3 (mg/dl)	72–135	ND	33.5	35.6	39.6	48.0	71.5	91.7
C4 (mg/dl)	9.9–31.5	ND	3.0	2.7	1.8	3.0	7.9	14.0
CRP (mg/dl)	<0.3	<0.3	2.79	2.33	1.09	1.17	1.36	<0.3
WBC (/mm^3)	4500–9000	1900	1900	2000	7700	9000	5500	8300
Lymphocytes (%)	20–30	ND	27	21	10	8	25	9
Ferritin (ng/ml)	5–70	ND	ND	11 815	ND	516	ND	39

The patient had myopia due to severe chemosis on admission. She also had liver involvement and hemophagocytic syndrome. The titer of anti-DNA antibodies decreased with the deterioration in liver function, as in a previous case report.[11] After treatment with steroid pulse therapy on 28–30 March, myopia, liver involvement, and hemophagocytic syndrome improved rapidly

[a] Methylprednisolone pulse (25–27 March)

VA, visual acuity; RE, right eye; LE, left eye; AST, aspartate aminotransferase; ALT, alanine transaminase; LDH, lactate dehydrogenase; Al-P, alkaline phosphatase; γ-GTP, gamma glutamyl transpeptidase; anti-DNA, anti-DNA antibodies; CRP, C-reactive protein; WBC, white blood cell; ND, not done

Table 2. Clinical and laboratory characteristics of SLE with chemosis

Source	Patient sex/age (years)	Systemic disease	Laboratory data	SLE duration	Myopia/ visual acuity	Medication (daily)	APS
Zaini[8]	Female / 21	Periorbital edema	ANA 1:80, low complement levels, LE cells (+), ESR 109 mm/h, Alb 2.9 g/dl, BUN 20 mg/dl, Cr 0.8 mg/dl	0 month	No	PSL (initial dose not reported)	Not reported
Ayazi[6]	Female / 18	Anasarca, polyarthralgia, photosensitive rash, hair loss, proteinuria	ANA 1:512, LE cells (+)	3 months	Yes RE (20/25 × −9.0 D) LE (20/30 × −10.0 D)	PSL (120 mg)	Not reported
Shu[7]	Female / 46	Periorbital oedema, proteinuria	ANA (+) (peri + homo), anti-DNA (+), low complement levels, ESR 116 mm/h, Alb 2.5 g/dl, BUN 30 mg/dl, Cr 1.1 mg/dl	14 years	Yes RE (20/15 × −3.75 D) LE (20/25 × −3.50 D)	PSL (40 mg)	Not reported
Leahey[9]	Female / 36	Periorbital edema, dusky erythematous macular eruption, malar rash, hair loss, polyarthritis, Raynaud's phenomenon	ANA 1:640 (sp + homo), low complement levels, hypoalbuminemia, BUN and Cr elevated	0 month	No	PSL (30 mg)	Not reported
Bohgaki et al. (this case)	Female / 24	Periorbital edema, malar rash, proteinuria	ANA 1:640 (sp + homo, anti-DNA (+), low complement levels, ESR 120 mm/h, Alb 2.7 g/dl, BUN 28 mg/dl, Cr 1.1 mg/dl, pancytopenia	0 month	Yes RE (20/18 × −3.0 D) LE (20/24 × −3.5 D)	PSL pulse, heparin, low-dose aspirin	LAC (+), IgGaCL (+), IgMaCL (+)

Reported cases of SLE with chemosis. Chemosis was detected in four out of five cases, including this case, at the beginning of the SLE, and was the first sign of exacerbation of lupus in the remaining case.[7] All cases responded well to systemic steroid therapy. No case except this one reported the existence of antiphospholipid antibodies

ANA, antinuclear antibodies; peri, perinuclear pattern; homo, homogeneous pattern; sp, speckled pattern; ESR, erythrocyte sedimentation rate; Alb, serum albumin; BUN, blood urea nitrogen; Cr, serum creatinine; anti-DNA, anti-DNA antibodies; RE, right eye; LE, left eye; D, dioptre; PSL, prednisolone; LAC, lupus anticoagulant; IgG isotype of anticardiolipin antibody; IgMaCL, IgM isotype of anticardiolipin antibody

related to hypoalbuminemia, and/or (ii) a change in lenticular curvature with anterior displacement of the lens due to ciliary body edema.[7] Regarding the mechanism of the latter theory, Shu et al.[8] have suggested that immune complexes might play a role in transient myopia. In addition to this mechanism, a perfusion defect caused by venous thrombi should also be considered. Reports of SLE patients with chemosis are summarized in Table 2.[7–10] All the patients were female. Chemosis was detected in three out of four cases at the beginning of SLE,[7,9,10] and was the first sign of exacerbation of lupus in the remaining case.[8] All cases responded well to systemic steroid therapy. Although APS was not described, chemosis may result from regional perfusion failure in the conjunctiva caused by thrombosis related to APS. In our patient, ocular inflammation and endocrine disorders were not proven. Retinopathy was mild, and myopia with chemosis did not improve with furosemide. Although a pathological examination was not carried out, APS might also have caused these ocular symptoms because of capillary occlusion.

The clinical courses of retinopathy, liver involvement,[11] and HPS were all in parallel with the activity of SLE. As myopia and chemosis may also reflect the activity of SLE,[9] these rare complications should be considered as a lupus manifestation.

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