Mod Rheumatol (2013) 23:172–174
DOI 10.1007/s10165-012-0629-6

CASE REPORT

Atypical severe central serous chorioretinopathy in a patient with systemic lupus erythematosus improved with a rapid reduction in glucocorticoid

Hiroe Sato · Satoshi Ito · Shingo Nagai · Akira Murasawa · Yoko Wada · Shuichi Murakami · Takeshi Kuroda · Masaaki Nakano · Ichiei Narita

Received: 17 January 2012 / Accepted: 5 March 2012 / Published online: 25 March 2012

Abstract A 36-year-old woman was diagnosed with systemic lupus erythematosus (SLE). Seven days after beginning glucocorticoid treatment, she developed reduced visual acuity, and atypical severe central serous chorioretinopathy (CSC) was confirmed. Since glucocorticoid use is an important risk factor for CSC, the PSL was reduced, tacrolimus was added, and the visual acuity improved rapidly. Reduction in glucocorticoid combined with the use of immunosuppressive agents is one option for preventing a deterioration in atypical severe CSC while still controlling SLE.

Keywords Systemic lupus erythematosus · Central serous chorioretinopathy · Glucocorticoid · Tacrolimus

Central serous chorioretinopathy (CSC) is an unusual complication of systemic lupus erythematosus (SLE), and there are only a few reports of it [1], especially the severe variant of CSC. CSC is characterised by an accumulation of subretinal fluid, which is accompanied by neurosensory retinal detachment and impairment of the barrier function of the retinal pigment epithelium (RPE) [2, 3]. Vascular hyperpermeability or circulatory failure of the choroidal vessels are predisposing factors [2, 3]. Typical CSC usually occurs in 20–50-year-old males, affects one eye, and improves spontaneously. Stress is an important risk factor. The severe form of CSC is associated with multifocal leakage on fluorescein angiography, usually affects both eyes, and the prognosis for visual acuity is sometimes poor [3, 4]. Glucocorticoid use is an important risk factor for multifocal, atypical severe or chronic CSC, which is clinically indistinguishable from multiple posterior pigment epitheliopathy (MPPE) [2, 3, 5, 6]; discontinuing or reducing the glucocorticoids is an effective way to prevent exacerbations, and aids retinal reattachment [3, 6].

Here, we report the case of a Japanese female with atypical severe CSC that developed after starting glucocorticoid treatment for SLE.

A 36-year-old woman visited our hospital because of eruptions on her face and back. She also had photosensitivity, stomatitis, polyarthralgia, diarrhoea, and anasarca. The patient's blood pressure was 102/76 mmHg and her blood count was normal. C-reactive protein was negative (0.20 mg/dL), and the total protein and serum albumin levels were low (4.5 and 1.8 g/dL, respectively). Antinuclear and anti-double-stranded DNA antibody tests were positive [59.6 index value (normal range <20 index value) and 32 IU/mL (normal range <12 IU/mL), respectively]. Complement activity was low [<14 U/mL (normal range 32–53 U/mL)]. Proteinuria was 0.28 g/day and renal function was normal. Computed tomography (CT) showed a right pleural effusion and ascites. A diagnosis of SLE was made. Methylprednisolone 500 mg was administered for

H. Sato (✉) · Y. Wada · S. Murakami · T. Kuroda · I. Narita
Division of Clinical Nephrology and Rheumatology, Niigata University Graduate School of Medical and Dental Sciences, 1-757 Asahimachi-Dori, Chuoku, Niigata, Niigata 951-8510, Japan
e-mail: hiroe-212@umin.ac.jp

H. Sato · S. Ito · A. Murasawa
Department of Rheumatology, Niigata Rheumatic Center, 1-2-8 Honcho, Shibata, Niigata 957-0054, Japan

S. Nagai
Department of ophthalmology, Shibata Hospital, 1-2-8 Honcho, Shibata, Niigata 957-0054, Japan

M. Nakano
Medical Laboratory Science, School of Health Sciences, Faculty of Medicine Niigata University, 2-746 Asahimachi-Dori, Chuoku, Niigata, Niigata 951-8510, Japan

3 days and oral prednisolone (PSL) (40 mg/day) was started. Her symptoms improved gradually, and the oedema decreased with albumin and diuretics. Seven days after beginning treatment, she developed reduced visual acuity and metamorphopsia in both eyes. The fundus examination revealed subretinal fluid and optical coherence tomography showed neurosensory retinal detachment in both eyes (Fig. 1). Fluorescein angiography showed multiple granular leakages in the macular and extramacular areas (Fig. 1), and atypical severe CSC was confirmed. As glucocorticoid treatment can cause CSC, the PSL was reduced to 20 mg/day for 8 days, and tacrolimus was added at 3 mg/day; visual acuity improved rapidly (Fig. 2). The pleural effusion and ascites also decreased, and the SLE disease activity was well controlled.

In this case, glucocorticoid therapy was thought to have triggered the severe atypical CSC because the visual disturbance followed the glucocorticoid treatment. As in previous reports, either oral or topical glucocorticoid treatment can trigger CSC [5]. Furthermore, endocrine disorders that increase endogenous catecholamines and glucocorticoids, such as Cushing's syndrome and steroid-producing tumours, have also been reported to be risk factors [7, 8]. A case–control study of the risk factors for CSC, including classic and chronic CSC, indicated that

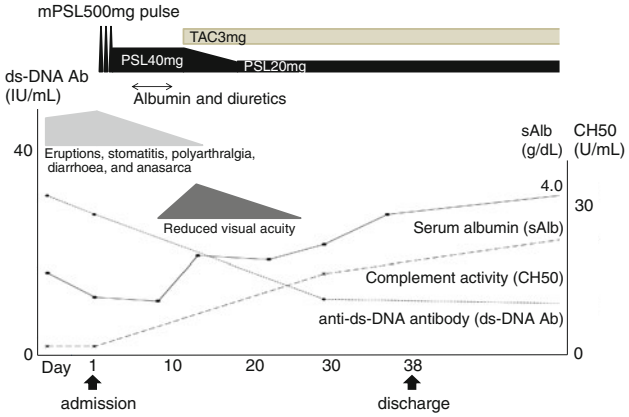

Fig. 2 The clinical course of this case. *mPSL* methylprednisolone, *PSL* prednisolone, *TAC* tacrolimus

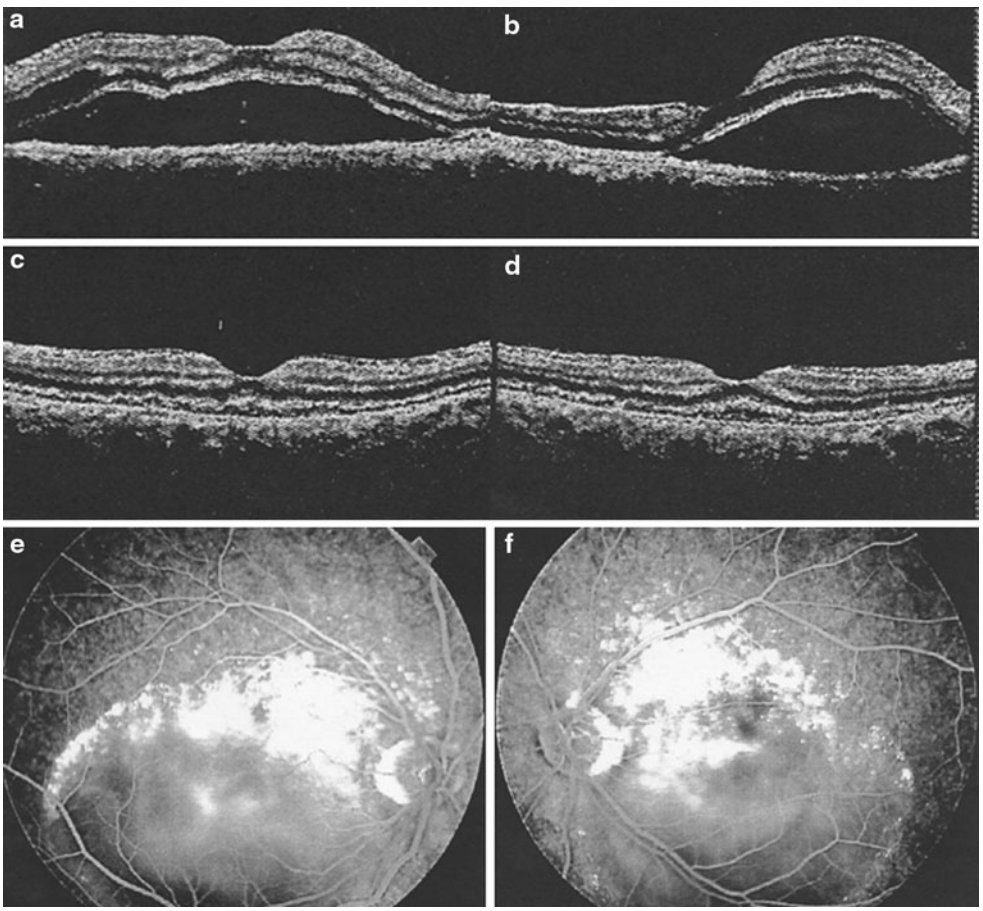

Fig. 1 Optical coherence tomography showed neurosensory retinal detachment in the right (**a**) and left (**b**) eyes. After reducing the dose of prednisolone, the pigment epithelial detachment improved in both eyes (**c**, **d**). Fluorescein angiography showed multiple granular leakages in the macular and extramacular areas in the right (**e**) and left (**f**) eyes

systemic steroid use had the highest odds ratio (odds ratio 37.1, 95 % confidence interval 6.2–221.8) [2]. Increased plasma cortisol aggravates the vascular permeability and capillary fragility in the choriocapillaris [3].

Although laser photocoagulation therapy contributes to retinal reattachment, retinal reattachment was possible only after discontinuing the glucocorticoid in 87.5 % of atypical severe CSC eyes without laser photocoagulation therapy [3]. Therefore, discontinuing the glucocorticoid is the first step in treating severe atypical CSC. However, discontinuing or reducing glucocorticoids can be fatal if the SLE is not controlled. In this case, the SLE treatment had just been started and her SLE activity had not been controlled completely. We decided to reduce the PSL carefully and add an immunosuppressive agent which could act relatively quickly. Tacrolimus was reported to reduce proteinuria and the SLE disease activity index (SLEDAI) faster than oral cyclophosphamide or azathioprine in patients with lupus nephritis (class V) [9]. Suzuki et al. also showed that SLEDAI improved within 4 weeks after starting tacrolimus in SLE patients with mild active manifestations [10]. Thus, we added tacrolimus 3 mg/day and reduced the glucocorticoid dose rapidly to 20 mg. This treatment improved the atypical severe CSC while still controlling the SLE.

Clinically, lupus choroidopathy is similar to CSC, and is often associated with severe SLE activity [11, 12]; additionally, it can cause typical CSC [1]. Immune complex deposition in the choroidal vessels [13] and autoantibodies against the RPE are pathological conditions [14], and intensive glucocorticoid therapy is often effective [11, 12]. We believe that our patient's atypical severe CSC was not directly related to the lupus activity, but potential lupus choroidopathy might have been present. Furthermore, severe hypoalbuminaemia and anasarca were accompanying factors, and these affect the vascular hyperpermeability and circulatory failure of the choroidal vessels. Lupus choroidopathy and choroidal circulation impairment are considered potential causes, which were aggravated by glucocorticoid therapy and led to the development of atypical severe CSC.

Ophthalmological examinations should be performed before starting glucocorticoid treatment to detect lupus choroidopathy and other conditions. After starting glucocorticoid treatment, atypical severe CSC is a rare complication of SLE; however, once it occurs, permanent visual loss can develop. Glucocorticoid reduction combined with immunosuppressive agents is an option to prevent a deterioration of atypical severe CSC while still controlling SLE.

Conflict of interest None.

References

1. Khng CG, Yap EY, Au-Eong KG, et al. Central serous retinopathy complicating systemic lupus erythematosus: a case series. Clin Experiment Ophthalmol. 2000;28:309–13.
2. Haimovici R, Koh S, Gagnon DR, et al. Risk factors for central serous chorioretinopathy: a case–control study. Ophthalmology. 2004;111:244–9.
3. Sharma T, Shah N, Rao M, et al. Visual outcome after discontinuation of corticosteroids in atypical severe central serous chorioretinopathy. Ophthalmology. 2004;111:1708–14.
4. Sharma T, Badrinath SS, Gopal L, et al. Subretinal fibrosis and nonrhegmatogenous retinal detachment associated with multifocal central serous chorioretinopathy. Retina. 1998;18:23–9.
5. Karadimas P, Kapetanios A, Bouzas EA. Central serous chorioretinopathy after local application of glucocorticoids for skin disorders. Arch Ophthalmol. 2004;122:784–6.
6. Wakakura M, Song E, Ishikawa S. Corticosteroid-induced central serous chorioretinopathy. Jpn J Ophthalmol. 1997;41:180–5.
7. Thoelen AM, Bernasconi PP, Schmid C, et al. Central serous chorioretinopathy associated with a carcinoma of the adrenal cortex. Retina. 2000;20:98–9.
8. Bouzas EA, Scott MH, Mastorakos G, et al. Central serous chorioretinopathy in endogenous hypercortisolism. Arch Ophthalmol. 1993;111:1229–33.
9. Szeto CC, Kwan BC, Lai FM, et al. Tacrolimus for the treatment of systemic lupus erythematosus with pure class V nephritis. Rheumatology (Oxford). 2008;47:1678–81.
10. Suzuki K, Kameda H, Amano K, et al. Single center prospective study of tacrolimus efficacy and safety in the treatment of various manifestations in systemic lupus erythematosus. Rheumatol Int. 2011;31:757–63.
11. Kamdar NV, Erko A, Ehrlich JS, et al. Choroidopathy and kidney disease: a case report and review of the literature. Cases J. 2009;2:7425.
12. Nguyen QD, Uy HS, Akpek EK, et al. Choroidopathy of systemic lupus erythematosus. Lupus. 2000;9:288–98.
13. Aronson AJ, Ordonez NG, Diddie KR, et al. Immune-complex deposition in the eye in systemic lupus erythematosus. Arch Intern Med. 1979;139:1312–3.
14. Matsuo T, Nakayama T, Koyama T, et al. Multifocal pigment epithelial damages with serous retinal detachment in systemic lupus erythematosus. Ophthalmologica. 1987;195:97–102.