Case report

Therapeutic dilemma in serpiginous choroiditis

Chieh-Yin Chenga, San-Ni Chena,b, Jiunn-Feng Hwanga,b, Chun-Ju Linac,d,*

a Department of Ophthalmology, Changhua Christian Hospital, Changhua, Taiwan
b School of Medicine, Chung Shan Medical University, Taichung, Taiwan
c Department of Ophthalmology, China Medical University Hospital, Taichung, 40402, Taiwan
d China Medical University, Taichung, 40402, Taiwan

ABSTRACT

A 43-year-old woman had blurred vision in the left eye for 4 years. Her best-corrected visual acuity was 20/20 in the right eye and 20/200 in the left eye. The fundus showed grayish-yellow, jigsaw-puzzle-shaped lesions at the level of the retinal pigment epithelium and choriocapillaries emanating from the optic nerve head in both eyes. Fluorescein angiography showed late leakage in active lesions and hypofluorescence in hyperpigmented areas. Oral prednisolone and cyclosporine were given first. However, after posterior subtenon triamcinolone injections in both eyes and one intravitreal triamcinolone injection in the left eye, macular edema worsened. Steroid-induced central serous chorioretinopathy was suspected, so we tapered prednisolone rapidly and changed to azathioprine. Subsequent optical coherence tomography demonstrated retinal pigment epithelial detachment in the right eye subsided gradually. Fundus autofluorescence imaging showed progressively quiescent lesions. Unfortunately, acute myocardial infarction, atrial fibrillation, and ischemic stroke developed after 6 weeks azathioprine. In the acute phase of serpiginous choroiditis, corticosteroids are most commonly used. However, steroid therapy may be complicated with steroid-induced central serous chorioretinopathy. When we shift to other systemic immunosuppressive regimens, such as azathioprine, the possibility of acute myocardial infarction should be kept in mind.

Copyright © 2013, The Ophthalmologic Society of Taiwan. Published by Elsevier Taiwan LLC. All rights reserved.

1. Introduction

We report a case with serpiginous choroiditis complicated with steroid-induced central serous chorioretinopathy. After discontinuation of steroids and shift to azathioprine, her ocular condition improved. However, acute myocardial infarction and ischemic stroke developed.

2. Case report

A 43-year-old woman noticed progressively blurred vision in the left eye for 4 years, but she paid little attention to it. Blurred vision in the right eye occurred recently, so she came to our outpatient clinic for help. She denied any systemic diseases except seizure in childhood. Her best-corrected visual acuity was 20/20 in the right eye and 20/200 in the left eye. Fundus examination demonstrated bilateral grayish-yellow, jigsaw-puzzle-shaped lesions at the level of the retinal pigment epithelium (RPE) and choriocapillaries, which emanated from the optic nerve head with distinct borders in both eyes (Fig. 1A and B). Her complete blood counts, differential counts, blood chemistry tests, C-reactive protein, erythrocyte sedimentation rate, rapid plasma regain, human leukocyte antigen typing, and chest X-ray all revealed insignificant findings. Optical coherence tomography (OCT; Cirrus HD-OCT Model 4000, Carl Zeiss Meditec, Dublin, CA, USA) showed RPE detachment (RPED) in the right eye (Fig. 1C). Absence of inner segment and outer segment area as well as subretinal fluid was noted in the left eye (Fig. 1D). Fluorescein angiography showed late leakage in active lesions and hypofluorescence in hyperpigmented areas. Serpiginous choroiditis was diagnosed, so 30 mg prednisolone daily and 100 mg cyclosporine twice daily were started.

The patient complained of facial swelling and leg pitting edema after 3 weeks of steroid treatment, so we tapered prednisolone gradually. We tried subtenon injections of 40 mg triamcinolone in both eyes to reduce systemic side effects, but OCT showed that RPED increased in the right eye and intraretinal fluid appeared in the left eye despite prednisolone and cyclosporine treatment.
Intravitreal 0.5 mg ranibizumab was also given in the right eye with no response (Fig. 2C). Therefore, we added mycophenolate mofetil 250 mg twice daily. After 1 month, RPED and retinal edema still progressed. We tried intravitreal injection of 4 mg triamcinolone in the left eye, but macular edema worsened (Fig. 2D). Steroid-induced central serous chorioretinopathy was suspected, so we tapered prednisolone rapidly and changed to 50 mg azathioprine twice daily. The subsequent OCT demonstrated that RPED resolved in the right eye after azathioprine treatment (Fig. 2E). The macular edema decreased a little after azathioprine treatment in the left eye which had received one intravitreal triamcinolone injection (Fig. 2F).

Unfortunately, acute myocardial infarction, atrial fibrillation, and ischemic stroke over the right middle cerebral artery territory developed after 6 weeks azathioprine. Therefore, azathioprine was stopped. After aggressive medical care and rehabilitation, left hemiparesis and aphasia with dysarthria improved gradually. At 8 months follow-up, the autofluorescence images (Fig. 3A and B) demonstrated quiescent lesions. OCT showed no RPED in the right eye and macular contours were back to normal (Fig. 3C). Macular

![Fig. 1](image1.png)

**Fig. 1.** (A) Fundus photography of the right eye showed grayish-yellow lesions. (B) Fundus photography of the left eye showed grayish-yellow and jigsaw-puzzle-shaped lesions, emanating from the optic nerve head with distinct borders, with variable amounts of pigmentation. (C) Optical coherence tomography (OCT) of the right eye showed mild retinal pigment epithelium detachment. (D) OCT of the left eye showed absence of photoreceptor inner segment/outer segment band and collection of subretinal fluid.

![Fig. 2](image2.png)

**Fig. 2.** (A) Optical coherence tomography (OCT) showed increased retinal pigment epithelium detachment (RPED) in the right eye after one subtenon injection of 40 mg triamcinolone, despite prednisolone and cyclosporine treatment. (B) OCT showed intraretinal fluid in the left eye after one subtenon injection of 40 mg triamcinolone, despite prednisolone and cyclosporine treatment. (C) Intravitreal 0.5 mg ranibizumab was also administered to the right eye with no response. (D) After one intravitreal injection of triamcinolone in the left eye, retina edema worsened. (E) OCT demonstrated that RPED resolved in the right eye after azathioprine treatment. (F) OCT demonstrated that macular edema decreased a little after azathioprine treatment in the left eye, which had received one intravitreal injection of triamcinolone.
edema subsided but absence of photoreceptor inner segment/outer segment band was still noted in the left eye (Fig. 3D).

3. Discussion

Serpiginous choroiditis is a rare, chronic progressive and inflammatory condition of unknown origin. It primarily involves the choroid and RPE. Most cases occur in middle age or older (about 4th–8th decades).1-2 There is also a slight male predominance. The clinical course has multiple recurrences, leading to potentially significant visual loss.3 Visual prognosis is related to the macular involvement.

In a retrospective study, Hooper and Kaplan first reported rapid remission in five patients with serpiginous choroiditis who were treated with a combination of prednisone, cyclosporine, and azathioprine.4 Combination of azathioprine and corticosteroids was also advocated.5 Local treatment such as periocular triamcinolone and intravitreal ranibizumab injection demonstrated favorable results.6,7 In our case, oral prednisolone and cyclosporine were tried, but macular edema worsened. Steroid-induced central serous chorioretinopathy was suspected.8 We finally used azathioprine, and macular edema and RPED gradually resolved in both eyes.

Unfortunately, after 6 weeks azathioprine, acute myocardial infarction, atrial fibrillation, and ischemic stroke over the right middle cerebral territory developed. In the literature, the relationship between myocardial infarction and ischemic stroke has been discussed. Approximately 15–30% of ischemic strokes are cardioembolic in origin, and atrial fibrillation, valvular heart disease, coronary artery disease, congestive heart failure, and myocardial infarction are significant risk factors for stroke.9 Atrial fibrillation is the most important predictor of myocardial infarction-related ischemic stroke.10

A postmarketing study based on 30 reports from the US Food and Drug Administration and user community in September 2012 showed 12,727 people reported side effects when taking azathioprine. Among them, 30 people (0.24%) reported acute myocardial infarction. Perhaps the present patient’s medical problems were related to the cascade of azathioprine-related acute myocardial infarction.

In the acute phase of serpiginous choroiditis, corticosteroids are most commonly used to control the inflammation. Although the possible side effect of steroid-induced central serous chorioretinopathy is rarely reported, we should be alert to it and change to other systemic immunosuppressive regimens if necessary. In addition, azathioprine-related myocardial infarction needs to be kept in mind, although it does have a very low incidence rate. We can perform pretreatment evaluation, such as thorough systemic physical examination and electrocardiography. We have to be cautious that medication not only can treat the diseases, but also can cause side effects that might lead to unwanted results.

References