A case of Churg–Strauss syndrome and central retinal artery occlusion with good visual recovery

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Here we report a case of Churg–Strauss syndrome (CSS) and central retinal artery occlusion (CRAO), with good visual recovery. A 58-year-old Japanese man with CSS experienced acute painless loss of vision in his right eye. CRAO was diagnosed by fundoscopic findings (retinal whitening with a cherry-red spot). Steroid pulse therapy (methylprednisolone at 1 g daily for 3 days) followed by combined treatment with prednisolone (30 mg/day) and cyclophosphamide (150 mg/day) was administered; his visual acuity recovered to 20/30 in 1 month, and no recurrence has occurred for 1 year. Steroid pulse therapy may be effective for CRAO in CSS patients.

Key words: Central retinal artery occlusion, Churg–Strauss syndrome, steroid pulse therapy

Churg–Strauss syndrome (CSS) is an uncommon systemic disease characterized by bronchial asthma, hypereosinophilia, and granulomatous small vessel vasculitis. Ocular involvement is unusual in CSS. There have only been a few case reports of CSS associated with central retinal artery occlusion (CRAO) and most of the patients had a poor visual outcome.[1-4] Here, we report a patient with CSS and unilateral CRAO that responded to steroid pulse therapy.

Case Report

A 58-year-old man had been diagnosed with CSS from bronchial asthma, eosinophilia, polyneuropathy, and infiltration in lungs 2 years before. Bilateral foot drop and hypereosinophilia developed 2 weeks before the present condition and he was treated with oral prednisolone (15 mg/day). On referral to the Department of Neurology of our hospital, he had a diagnosis of peroneal nerve paralysis by CSS and steroid pulse therapy (methylprednisolone at 1 g/day) was started. The following day, he experienced acute painless loss of vision in his right eye and was immediately admitted to the Department of Ophthalmology.

On initial examination, the best corrected visual acuity was hand motion on the right side and 20/20 on the left. Slit-lamp examination revealed bilateral early cataracts. There was marked pallor of the retina with a cherry-red spot in the fovea on fundoscopy of the right eye [Fig. 1a]. Fluorescein angiograms showed delayed arterial filling in the right eye [Fig. 1b]. Eyeball massage and anterior chamber paracentesis were performed as initial treatment, but retinal perfusion did not recover. Laboratory tests showed a white blood cell count of 15,250/mm^3, with hypereosinophilia (24%). C-reactive protein (reference range is 0.3 mg/dl or less) increased to 6.4 mg/dl. Perinuclear antineutrophil cytoplasmic antibodies (p-ANCA) and cytoplasmic antineutrophil cytoplasmic antibodies (c-ANCA) were negative. Because it was thought that the patient's CRAO had been caused by an inflammatory process, steroid pulse therapy (methylprednisolone at 1 g daily for 3 days) was continued and was followed by combined treatment with oral prednisolone (30 mg/day) and low-dose cyclophosphamide (150 mg/day) together for management of intractable CSS. There was rapid resolution of foot drop and normalization of hypereosinophilia. His visual acuity improved more slowly and recovered to 20/30 one month after CRAO. Result of fluorescein angiography performed at that time showed resolution of the retinal ischemia of the right eye [Fig. 2].

Oral prednisolone was tapered slowly to 10 mg/day over 6 months. No recurrence has occurred over 18 months, and the most recent visual acuity was 20/30.

Discussion

This was an uncommon case of CRAO in a patient with CSS. Ocular involvement is infrequent in CSS and the ocular manifestations can be classified into two types, which are those related to ischemic vasculitis and those due to an orbital inflammatory pseudotumor.[5] In the present case, ANCA was normal but the clinical manifestations were

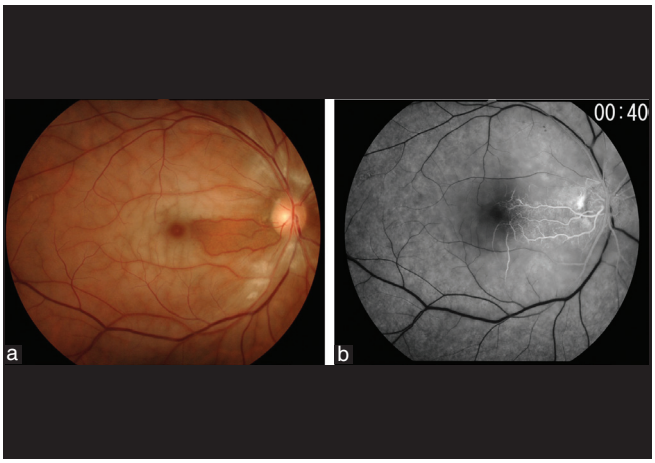

Figure 1: (a) Color fundus photograph of the right eye, showing a cherry-red spot, retinal pallor, and attenuated retinal arteries without emboli. (b) Fluorescein angiograms of the right eye at 40 s after injection, showing delayed arterial filling

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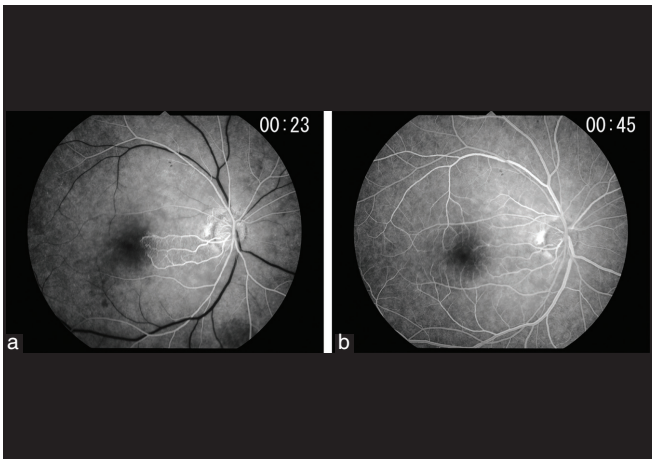

Figure 2: Fluorescein angiograms of the right eye at (a) 23 s and (b) 45 s after injection, showing resolution of the retinal ischemia of the right eye 1 month after CRAO

consistent with ischemic vasculitis. There have been a few previous reports about CSS patients without ANCA who developed CRAO.[1,3,4] These cases may indicate that the ANCA status is not a useful criterion for the classification of ocular inflammation in CSS.

CSS is treated with 30-40 mg/day of prednisolone in a light case and a moderate case. In a severe case, it is treated with 60 mg/day prednisolone or by steroid pulse therapy combined with immunosuppressant. When it is steroid resistant, a high-dose intravenous gamma globulin may be used. Because peroneal nerve paralysis appeared during treatment with oral prednisolone, we thought that he was a severe CSS case. For that reason, steroid pulse therapy and low-dose cyclophosphamide were used together in this case.

According to previous reports, CRAO associated with CSS had a poor visual outcome.[1-4] It might be due to low-dose systemic corticosteroids (prednisolone ≤ 60 mg) or delay in performing steroid pulse therapy. In the present patient, steroid pulse therapy had already been started on the day before the occurrence of CRAO and the final visual acuity was 20/30. Therefore, early steroid pulse therapy may be effective for CRAO in CSS patients. But it remains possible that in this case, steroid pulse therapy increased platelets' aggregation because coagulation parameters were not examined during a steroid pulse therapy. In addition, more cases are needed to confirm our hypothesis.

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