

Central Retinal Vein Occlusion During Remission of Ulcerative Colitis

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Background: Retinal vascular disease is a rare complication of ulcerative colitis.

Case: We report a patient who developed unilateral nonischemic central retinal vein occlusion (CRVO) (papillophlebitis) without any other retinal vascular disease during remission of ulcerative colitis.

Observations: The best-corrected visual acuities were 1.5 OD and 0.7 OS. Dilated and tortuous retinal veins and retinal bleeding were seen in the left eye. Macular edema and leakage from the papilla and the retinal veins of the left eye were evident on fluorescein angiography. After increased dosage of systemic prednisolone was prescribed, the retinal vascular changes resulting from CRVO (papillophlebitis) in the left eye gradually abated.

Conclusions: Retinal vascular diseases should be monitored during both remission and activation of intestinal symptoms of ulcerative colitis. **Jpn J Ophthalmol 1999;43:213–216** © 1999 Japanese Ophthalmological Society

Key Words: Central retinal vein occlusion, inflammatory bowel disease, papillophlebitis, retinal vascular disease, ulcerative colitis.

Introduction

Ulcerative colitis sometimes induces ocular involvement such as iridocyclitis, keratitis, corneal ulceration, conjunctivitis, episcleritis, and central serous chorioretinopathy. However, retinal vascular diseases associated with ulcerative colitis are rare. To our knowledge, only four cases of retinal vascular diseases, ie, central retinal artery occlusion,^{1,2} retinal periphlebitis,³ and cilioretinal arterial occlusion with central retinal vein occlusion (CRVO),⁴ have been reported to occur during the activation of intestinal symptoms. In this report, we describe a patient who developed CRVO (papillophlebitis) without any other retinal vascular disease during remission of ulcerative colitis.

Case Report

A 34-year-old Japanese man with a 14-year history of chronic, intermittent type ulcerative colitis diagnosed by colonoscopy was admitted to the Department of Internal Medicine, Mie University Hospital, on July 27, 1994, because of activation of ulcerative colitis. He had severe bloody diarrhea; systemic prednisolone (60 mg/day), prednisolone enema (40 mg/day), and sulfasalazine enema (1 g/day) were prescribed. When his abdominal symptoms abated, the medications were tapered gradually. On September 8, the patient was discharged with systemic prednisolone (30 mg/day) and systemic sulfasalazine (4 g/ day). As his abdominal condition was in remission, the systemic prednisolone and systemic sulfasalazine were gradually tapered. Systemic prednisolone was discontinued on March 15, 1995. The erythrocyte sedimentation rate (ESR) (13 mm/hour; normal, <15 mm/hour), platelet count (204,000 cells/µL; normal, 180,000-365,000 cells/ μ L), thrombin time (10.6 seconds; normal, 10-13 seconds), and the activated partial thromboplastin time (37.7 seconds; normal, 32-48 seconds) were normal. Because his ulcerative colitis was in remission, systemic sulfasalazine (2 g/ day), the usual maintenance therapy, had been prescribed.

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On May 25, 1995, with the complaint of blurred vision in the left eye, the patient consulted an ophthalmologist. His corrected visual acuities were 1.2 OD and 1.2 OS. Although the anterior segments of both eyes were normal, dilated and tortuous retinal veins and retinal bleeding were seen in the left eye (Figure 1). Nonischemic CRVO (papillophlebitis,⁵ venous stasis retinopathy) was diagnosed. Systemic prednisolone (10 mg/day) was administered. Because retinal bleeding gradually worsened in the left eye (Figure 2), and his left visual acuity decreased, he was referred to the Department of Ophthalmology, Mie University Hospital, on August 29, 1995.

When we first examined the patient, his corrected visual acuities were 1.5 OD and 0.7 OS. Macular edema and leakage from the papilla and the retinal veins of the left eye were evident on fluorescein angiography (Figure 3). However, retinal arterial filling was normal. Arm-to-retina circulation time was 11.1 seconds (normal, 10-15 seconds), and the retinal circulation time was 13.3 seconds (normal, 5-11 seconds). He did not have symptoms of active ulcerative colitis, such as diarrhea, fever, tachycardia, or anemia. Diabetes mellitus, hypertension, heart disease, pulmonary disease, and renal disease were not found. Although the ESR (4 mm/hour), the platelet count (232,000 cells/µL), and the thrombin time (10.5 seconds) were still normal, the activated partial thromboplastin time was 30.7 seconds. Because 10 mg/day of prednisolone had been ineffective in the prior treatment of this patient, we prescribed 30 mg/ day of systemic prednisolone. As the retinal vascular



Figure 2. Despite systemic corticosteroid administration (10 mg/day), retinal bleeding gradually worsened in left eye.

changes resulting from CRVO in the left eye gradually abated, systemic prednisolone was tapered over 10 months. His ulcerative colitis was still in remission.

On May 1, 1996, fluorescein angiography showed that the dilatation and tortuosity of the retinal vein and leakage from the papilla had resolved. Macular edema and leakage from the peripheral retinal veins had decreased (Figure 4). Arm-to-retina circulation time and retinal circulation time were 10.3 seconds, and 8.9 seconds, respectively. His corrected visual acuities were 1.5 OD and 1.2 OS. The retinal bleeding and macular edema resolved on June 21, 1996.



Figure 1. Central retinal vein occlusion (papillophlebitis) in left eye. Dilated and tortuous retinal veins and retinal bleeding are seen.



Figure 3. Fluorescein angiography demonstrates dilatation and tortuosity of posterior pole retinal veins, macular edema, and leakage from papilla and retinal veins.



Figure 4. After systemic corticosteroid administration (30 mg/day), fluorescein angiography shows that dilatation and tortuosity of retinal veins and leakage from papilla have resolved. Macular edema and leakage from peripheral retinal veins have subsided.

The ESR (8 mm/hour), the platelet count (223,000 cells/ μ L), and the thrombin time (11.3 seconds) were normal, and the activated partial thromboplastin time also returned to normal (37.0 seconds). The patient's corrected visual acuity was 1.5 bilaterally. CRVO has not recurred.

Discussion

CRVO in young adults who present with the complaint of blurred vision or photopsia has sometimes been called papillophlebitis, which is characterized by unilateral optic disc edema, and dilatation and tortuosity of the major retinal veins with a variable amount of retinal hemorrhage.⁵ Papillophlebitis⁵ usually manifests as nonischemic CRVO or venous stasis retinopathy. CRVO in older patients is occasionally associated with systemic abnormalities, such as hypertension, diabetes, and systemic vascular diseases. However, vasculitis may cause occlusion in young adults.⁶ Ulcerative colitis and Crohn's disease sometimes induce arterial and venous thrombosis in any vascular system.^{4,7} Although ulcerative colitis is distinct from Crohn's disease, both diseases are recognized as inflammatory bowel disease. In Crohn's disease, retinal vascular diseases, such as branch retinal artery occlusion,8 obliterative retinal arteritis and phlebitis,9 CRVO,10 and retinal vasculitis with branch retinal artery occlusion¹⁰ also have been reported. Although Keyser and Hass⁴ reported CRVO with cilioretinal arterial occlusion in ulcerative colitis, the present patient with CRVO had no symptoms of any other retinal vascular disease. Causes other than ulcerative colitis were not found during examinations conducted by physicians, ophthalmologists, and in laboratory findings in this case. We believe that the ulcerative colitis was the cause of the CRVO.

In some previously reported cases of inflammatory bowel disease, coagulation abnormalities, such as increased levels of platelet counts, factor V, factor VIII, and fibrinogen, and decreased levels of antithrombin III have occurred.⁷ However, these abnormalities did not occur in all cases.⁴ In the present case, only the activated partial thromboplastin time was slightly shorter than normal. When CRVO was seen, the activated partial thromboplastin time (30.7 seconds) was shorter than before CRVO was diagnosed (37.7 second) and after CRVO resolved (37.0 seconds).

The effective therapy for retinal vascular disease in ulcerative colitis has not been established;⁴ systemic corticosteroids were used to treat the retinal thromboembolic events in Crohn's disease.8,9 CRVO in young adults sometimes resolves spontaneously. However, because the prognosis in about 40% of CRVO cases is poor, corticosteroids are sometimes administered.⁵ In the present case, we prescribed systemic prednisolone because at the time CRVO was diagnosed the patient had not taken prednisolone for about 2 months because his abdominal condition was quiescent. Furthermore, venous stasis retinopathy with macular edema, which was also seen in this patient, has usually been treated effectively by systemic corticosteroids.⁶ Retinal periphlebitis,³ another venous retinal disease associated with ulcerative colitis, was also reported to respond to systemic prednisolone. Although 10 mg/day of systemic prednisolone had been ineffective in this patient when he was being treated at another hospital, 30 mg/day effectively treated the CRVO after the patient was referred to our hospital. Retinal arterial disease did not accompany CRVO in this case, but ulcerative colitis sometimes induces retinal arterial disease. The prognosis of retinal artery occlusion in ulcerative colitis is generally pessimistic.^{1,2,4}

The chronic intermittent ulcerative colitis seen in this patient was marked by long periods of quiescence interspersed with acute attacks lasting weeks to months. In the present case, CRVO was found during remission of ulcerative colitis; the patient did not have symptoms of the active disease, such as diarrhea, fever, tachycardia, anemia, and an elevated ESR. The development of retinal vascular changes in other cases of ulcerative colitis has been reported during active intestinal symptoms.¹⁻⁴ The administration of systemic prednisolone to treat CRVO may have prevented activation of the abdominal symptoms of ulcerative colitis in this case. The retinal vascular changes in ulcerative colitis should be monitored during periods of activation as well as during remission of intestinal symptoms.

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