Central Retinal Vein Occlusion Combined with Cilioretinal Artery Occlusion

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A healthy 65-year-old man with sudden profound visual loss in his right eye presented with clinical signs of central retinal venous occlusion and retinal whitening, indicative of a cilioretinal arterial obstruction. He had been diagnosed with cilioretinal artery occlusion at a private ophthalmology clinic three days before being referred to our department. On fluorescein angiogram of the affected eye, the proximal portion of the retinal arteries filled with dye 27.3 seconds after injection, indicating a delay in retinal arterial filling. Moreover, the cilioretinal artery did not fill at that phase, but went on to fill 45.1 seconds after injection. Over 63.4 seconds after the filling of the retinal arteries, the laminar flow of the retinal venous vessels appeared. This was not until 90.7 seconds after injection. This patient was elderly, had no systemic diseases, and showed non-ischemic CRVO, prolonged retinal arterial filling on fluorescein angiography, and poor prognosis in visual acuity. His clinical course seemed to favor the pathogenetic hypothesis of a primary arterial affection.

Key words: cilioretinal artery occlusion, central retinal vein occlusion, delayed retinal arterial filling

INTRODUCTION

The association of cilioretinal arterial occlusion with central retinal vein occlusion (CRVO) has been described by many authors.1-13 Brown et al.4 reported that obstruction of the cilioretinal artery may occur either in isolation or in conjunction with central venous obstruction or ischemic optic neuropathy. Combined CRVO and cilioretinal artery occlusion is uncommon in the elderly. Most reported patients are young adults. The pathogenesis of this condition has not yet been established. The association of retinal infarction along the course of a cilioretinal artery with CRVO has been documented.2,4,9 It has been suggested that the infarction is a result of a decrease in perfusion caused by either an increase in venous pressure or a reduction in arterial pressure.1 However, Noble11 suggested that the major cause of the decrease in perfusion of the cilioretinal artery is an increase in the resistance of the central retinal vein. It has been suggested that the association of cilioretinal artery occlusion with CRVO may occur more frequently than it appears, since retinal hemorrhages can obscure cilioretinal obstruction.9

Here we present a healthy 65-year-old patient showing retinal whitening related to a cilioretinal arterial obstruction and retinal signs of CRVO.
Fig. 1. A, Color photograph: White edematous retinal infarction of the area supplied by the cilioretinal artery is seen. The retinal veins are dilated and tortuous with scattered hemorrhages. B, FAG (27.3 seconds after dye injection): Fluorescein angiogram of the right eye demonstrated delayed filling of dye in the proximal portion of the retinal arteries. A cilioretinal artery is not still perfused, and the surrounding retina shows hypofluorescence caused by ischemic edema. C, FAG (45.1 seconds after dye injection): The cilioretinal artery is filling with dye. The cilioretinal arteries continued to fill as dye appeared in the retinal arteries 7.8 seconds later. D, FAG (90.7 seconds after dye injection): Over 63.4 seconds after the filling of retinal arteries, laminar flow of the retinal venous vessels appeared. This did not occur until 90.7 seconds after injection, which indicates a prolongation in the retinal arteriovenous transit time.

CASE REPORT

A 65-year-old man noticed a sudden profound visual loss in his right eye. At first examination of this patient in a private ophthalmology clinic, a whitish retinal edema corresponding to a cilioretinal artery in the right eye was the most prominent finding. The retinal veins appeared slightly dilated and tortuous at that time. The best-corrected visual acuity was 20/200 in the right eye and 20/20 in the left eye. Three days later, he consulted our Department of Ophthalmology. Fundus examination showed retinal whitening along a cilioretinal artery, changes in the venous vessels, and retinal hemorrhages in the right eye (Fig. 1A). The visual acuity decreased to hand motions in the right eye. The intraocular pressure was 17 mmHg bilaterally. The anterior segment was normal. No vitreous cells were present. The left eye was unremarkable. Fluorescein angiogram of
Fig. 2. A, Color photograph: Four months after the initial visit, the retinal whitening along the affected cilioretinal artery was less prominent and the veins were less dilated and tortuous. B: Circulation of retinal vessels had improved on FAG, showing the laminar flow of the retinal veins at 26.3 seconds after injection.

the right eye demonstrated a delayed filling of dye in the proximal portion of the retinal arteries 27.3 seconds after injection (Fig. 1B). Moreover, the cilioretinal artery did not fill at that phase, but went on to fill 45.1 seconds after injection (Fig. 1C). The cilioretinal arteries continued to fill as dye appeared in the retinal arteries 17.8 seconds later. Over 63.4 seconds after the filling of the retinal arteries, the laminar flow of the retinal venous vessels appeared. This was not until 90.7 seconds after injection, which indicates a prolongation in the retinal arteriovenous transit time (Fig. 1D). The circulation in the cilioretinal arteries did not demonstrate a pulsatile quality with repeated intermittent retrograde flow. Four months after the patient’s initial visit to the Department of Ophthalmology, the retinal whitening along the affected cilioretinal artery was less prominent and the veins were less dilated and tortuous (Fig. 2A). The circulation of the retinal vessels had improved on fluorescein angiography, showing a laminar flow of the retinal veins at 26.3 seconds after injection (Fig. 2B), and no leakage at the optic nerve or staining of the vein walls in the late phase. The visual acuity was 20/400 in the right eye. The patient was treated with pentoxifylline (Trental®) 300 mg three times daily for three months.

Systemic evaluation failed to reveal any clinical evidence of thromboembolic conditions, cardiac diseases, neurologic abnormalities, diabetes mellitus, or connective tissue disorders. Normal or negative laboratory determinations included the following: complete blood cell count with differential, platelet count, sedimentation rate, urinalysis, blood lipid profiles, fasting blood sugar (FBS), 2 postprandial blood sugar (2 PPBS), rheumatoid factor, protein C3 and C4, C reactive protein, antithrombin III, Coombs’ test, prothrombin time (PT), activated partial thromboplastin time (PTT), cryofibrinogen, anticardiolipin IgG and IgM, lupus anticoagulant, and VDRL. The electrocardiogram, echocardiogram, and chest x-ray showed no significant abnormality.

**DISCUSSION**

At the first examination of this patient, a whitish retinal edema corresponding to a cilioretinal artery was the most prominent finding. Three days later, the retinal whitening persisted and the characteristic funduscopic changes of CRVO predominated. This patient was elderly, had no systemic diseases, and showed non-ischemic CRVO, prolonged retinal arterial filling on fluorescein angiography, and poor prognosis in visual acuity.

Brazitikos et al. studied two groups of patients with combined cilioretinal artery occlusion and CRVO. In the first group, the patients showed the ophthalmoscopic changes characteristic of CRVO and delayed filling of the cilioretinal artery during fluorescein angiography. All patients in this group (average age: 53 years) had systemic etiologic fac-
tors that could account for retinal vascular occlusion. The visual acuity did not recover in any of these patients. The patients in the second group were younger (average age: 31 years) and three of the four patients showed no signs of systemic disease. The cilioretinal artery filled normally on fluorescein angiography. Visual acuity recovered rapidly, but a paracentral scotoma remained. The clinical course of our patient, an otherwise healthy, elderly man showing a non-ischemic CRVO with reduction of visual acuity, seems to be related to neither the first nor the second group.

Prolonged branch retinal arterial filling has been described in cases of cilioretinal arterial occlusion combined with CRVO. In this patient, the affected cilioretinal artery and the central retinal artery revealed prolonged retinal arterial filling during fluorescein angiography. Rhythmic changes of cilioretinal arterial filling in cases of CRVO have been reported by Zylbermann et al. and Schatz et al. The repeated reversals of blood flow in the cilioretinal artery may reflect the different arterial pressures during systole and diastole. This phenomenon of intermittent retrograde flow was noted in four patients by McLeon and Ring and in five patients by Schatz et al. However, we did not observe such a finding in our patient during fluorescein angiography.

The pathogenetic mechanisms of cilioretinal artery occlusion combined with CRVO have not yet been established. However, a few hypotheses have been proposed. The first hypothesis is the development of cilioretinal artery occlusion secondary to the raised capillary pressure caused by CRVO. The second hypothesis is that a primary reduction in the perfusion pressure of the cilioretinal and retinal arteries may lead to decreased retinal circulation and subsequent venous stasis and thrombosis. A fall in systemic blood pressure and inflammatory, atherosclerotic or atherosclerotic retinal arterial disease have been suggested as possible causes of reduced arterial perfusion pressure. In addition, the combination of decreased plasminogen activity and elevated lipoprotein(a) should also be considered as a possible cause of retinal vein and artery occlusion. Orth deduced why cilioretinal artery occlusion in older patients with a central retinal vein occlusion is uncommon. These older patients frequently have arteriosclerotic disease involving the central retinal artery. This aging process within the walls of the central retinal artery may cause the perfusion pressure to approximate the lower pressure within the cilioretinal artery. Thus, the capillary bed pressure is reduced and does not impede to approximate cilioretinal artery perfusion.

In our patient, initial retinal whitening along a cilioretinal artery, followed by signs of venous stasis, seemed to favor the pathogenetic hypothesis concerning a primary arterial affection. In addition, this patient demonstrated prolonged, irregular filling of the branch retinal arteries on fluorescein angiography. This may have been caused by a disorder that primarily affects the retinal arteries or may have occurred as a secondary effect of central retinal vein occlusion.

REFERENCES


