

Paracentral Acute Middle Maculopathy in a Patient with Cilioretinal Artery Occlusion

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Abstract

Case Report

Introduction: A 49-year-old woman presented at Inoda Eye Clinic with inferior visual field defect in the left eye. Ophthalmoscopic examination revealed well-demarcated retinal edema of the superior region at the posterior pole in the left eye. The area of retinal edema was consistent with the area of the cilioretinal artery, and therefore, cilioretinal artery occlusion was suspected. Optical coherence tomography showed hyperreflective bandlike lesions involving the middle layers of the retina at the level of the inner nuclear layer. Optical coherence tomography angiography showed decreased flow sign in outer retina layer and choriocapillaris layer rather than in superficial layer. Based on these findings, the patient was diagnosed with paracentral acute middle maculopathy due to cilioretinal artery occlusion. Optical coherence tomography angiography is a useful diagnostic examination instead of fluorescein angiography for evaluation of cilioretinal artery occlusion.

Keywords: Paracentral acute middle maculopathy, cilioretinal artery occlusion, optical coherence tomography, optical coherence tomography angiography.

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INTRODUCTION

The retinal artery is a terminal artery responsible for blood flow in the inner retinal layer. Cilioretinal arteries, which belong to the posterior ciliary artery system, usually arise from the peripapillary choroid or directly from one of the short posterior ciliary arteries [1]. Clinically evident cilioretinal artery occlusions (CilioRAO), thought to be quite rare, appear to comprise about 5.3%-7.1% of all retinal artery occlusions (RAO) [2, 3]. Fluorescein angiography (FA) has been a standard method for the diagnosis of RAO. However, recently, noninvasive microcirculation evaluation by optical coherence tomography angiography (OCTA) has become possible. There have been few reports in the literature presenting a CilioRAO evaluated by OCTA [4-6].

Paracentral acute middle maculopathy (PAMM) was originally described by Sarraf *et al.*, [7] and has been noted to be associated with both central retinal vein and RAO. PAMM is a clinical sign of ischemia of the deep retinal vascular layers.

Herein, we report a case of PAMM due to CilioRAO occlusion evaluated by OCTA.

CASE REPORT

A 49-year-old woman presented at the Inoda Eye Clinic 2 days after development of a sudden, painless, inferior visual field defect in the left eye. The patient had a history of mitral and aortic valve regurgitation. On ophthalmic examination, the best-corrected visual acuity in the right eye was 20/20 and in the left eye acuity was 20/20. Ocular pressures were normal. Slit lamp examination showed no abnormalities in both eyes. Funduscopy of the left eye showed well-demarcated retinal edema of the superior region at the posterior pole, sparing the fovea (Figure 1A). The cilioretinal artery extended from the optic disc. The area of retinal edema was consistent with the area of the cilioretinal artery, and therefore, CilioRAO was suspected. Optical coherence tomography (OCT) showed hyperreflective bandlike lesions involving the middle layers of the retina at the level of the inner nuclear layer (Figure 1B). OCTA showed decreased flow sign in outer retina layer (Figure 1D) and choriocapillaris layer (Figure 1E) rather than in superficial layer (Figure 1C).

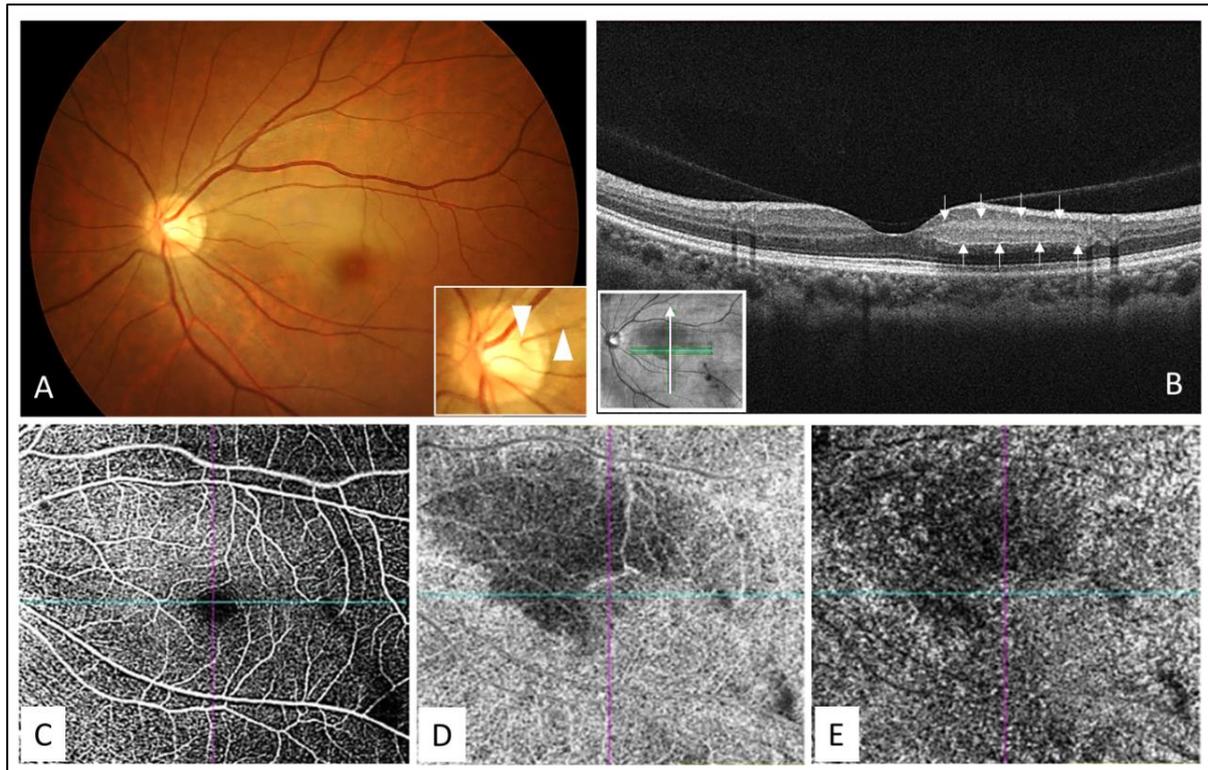


Figure 1: (A) Left fundus photograph shows a cilioretinal artery originating from the optic disc (inset arrows). White changes are seen in the retina around the cilioretinal artery, sparing the fovea. (B) Optical coherence tomography showed hyperreflective bandlike lesions involving the middle layers of the retina at the level of the inner nuclear layer (arrows). (C-E) Optical coherence tomography angiography showed decreased flow sign in outer retina layer (D) and choriocapillaris layer (E) rather than in superficial layer (C)

Based on these findings, the patient was diagnosed with PAMM due to CilioRAO. She was referred to a higher medical institution for further examinations.

DISCUSSION

Generally, cilioretinal arteries have been shown to originate from short posterior ciliary arteries, and they appear ophthalmoscopically to emerge from the optic disc or disc margin separately from the central retinal artery or its branches. Previously, cilioretinal arteries had been identified by their characteristic hook appearance on ophthalmoscopy. The presence of the cilioretinal artery was confirmed in 379 of the 956 eyes (39.6%) [1]. Clinically evident CilioRAO, thought to be quite rare, appear to comprise about 5.3%-7.1% of all RAO [2, 3]. CilioRAO can be classified clinically into three categories: (1) nonarteritic CilioRAO alone, (2) arteritic CilioRAO associated with giant cell arteritis, and (3) CilioRAO associated with central retinal vein occlusion [8]. The present case corresponds to nonarteritic CilioRAO alone.

Generally, central RAO showed decreased flow sign in both the superficial and deep layers, but the superficial layer was more injured. However, there have been few reports in the literature presenting a CilioRAO evaluated by OCTA [4-6]. Nakayama *et al.*, [4]

described the case of a 78-year-old man with CilioRAO. According to their report, decreased flow sign was observed in all layers in OCTA. Ikebukuro *et al.*, [5] described the case of a 70-year-old man with CilioRAO. According to their report, there was more damage in the deep layer than in the superficial layer. In the present case, there was more damage in the deep layer, which may suggest that CilioRAO may be more damaging to the capillary network in the deep layer than in the superficial layer. Furthermore, there have been few reports in the literature presenting PAMM with CilioRAO [4-6]. In the largest series, Pichi *et al.*, [6] evaluated 53 eyes with CilioRAO. According to their report, PAMM was present in 100% of isolated CilioRAO. They stated that this finding may be due to hypoperfusion of the involved cilioretinal artery, rather than complete occlusion. Ischemic injury may occur in only the most susceptible retinal tissue that may be normally associated with lower oxygen saturation levels due to the deeper location or due to greater oxygen demand and consumption or other mechanisms.

CONCLUSION

Although this is only a case report and more cases are needed, OCTA is a useful diagnostic examination instead of FA for evaluation of CilioRAO.

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