A Case of Paracentral Acute Middle Maculopathy Presenting Scotoma and Normal Visual Acuity

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Dear Editor,

Paracentral acute middle maculopathy (PAMM) is an optical coherence tomography (OCT) finding of hyper-reflective band in inner nuclear layer (INL) [1,2]. PAMM is caused by hypoperfusion of the deep vascular complex (i.e. intermediate capillary plexus [ICP] and deep capillary plexus [DCP]) or specifically the DCP [1]. PAMM is associated with retinal vascular diseases and systemic disorders [1,3]. We report a case of PAMM in an elderly patient followed for one year who presented scotoma and had normal visual acuity.

A 67-year old male presented with blurred vision and scotoma in the left eye that started five days ago. His medical history included hypertension, hypothyroidism, hypercholesterolemia. Uncorrected visual acuity (UCVA) was 20/20, and intraocular pressure was normal in both eyes. Anterior segment was normal. Fundus showed perifoveal drusen in both eyes, deep grayish lesions at parafovea, and cotton wool patches (CWPs) around the optic disc in the left eye (Fig. 1A). Humphrey visual field (HVF) showed inferior defects in the right eye and scattered defects in the left eye (Fig. 1B). OCT revealed multiple hyper-reflective bands in INL in the left eye and no significant findings in the right eye. The position of the grayish lesions on fundus corresponded to the hyper-reflective INL lesions (Fig. 1C). OCT angiography (OCTA) demonstrated decreased vascular density of DCP in the left eye (Fig. 1D). Cardiologic workup was unremarkable. Neurologic workup demonstrated luminal irregularity at bilateral distal internal carotid arteries (ICAs) in brain magnetic resonance imaging. After one week, UCVA was 20/22 in both eyes. Fundus showed diminished grayish lesions and increased CWPs in the left eye (Fig. 1E). HVF revealed decreased defects. OCT showed slightly decreased hyper-reflective bands in INL. Two weeks later, his symptom improved slightly. UCVA was stationary. CWPs decreased (Fig. 1F). HVF improved. OCT showed decreased hyper-reflective INL lesions. OCTA showed hypoperfusion of ICP and DCP (Fig. 1J). After two months, UCVA was similar. Fundus findings, HVF and OCT improved further (Fig. 1G). After six months, he rarely had visual impairment. UCVA was 20/20. CWPs almost disappeared (Fig. 1H). HVF was stationary. The hyper-reflective INL lesions improved and focal INL thinning was observed. The hypoperfusion of ICP and DCP decreased on OCTA (Fig. 1J). After one year, UCVA was stationary and the objective findings were nearly stationary (Fig. 1I). INL thinning developed slightly further. OCTA improved further (Fig. 1J).
To the best of our knowledge, this is the third case report describing PAMM in Korea. One study presented a 38-year-old healthy male with acute vision loss (BCVA: 20/60) whose systemic workup revealed a patent foramen ovale [4]. Another study reported a 43-year-old woman with visual field defects in both eyes (BCVA: 20/25 in the right eye, 20/40 in the left eye) after severe headache [5]. The two case reports had short follow-up period of 3 months and 2 weeks, respectively.

Our case report is different from the two studies in several respects. Firstly, the patient in our case presented blurred vision and scotoma, but had visual acuity of 20/20. Secondly, our case had longer follow-up period than the previous two reports. We observed gradual improvement of the hyper-reflective INL bands and the consequent thinning of INL on OCT. We also found the restoration of hypoperfusion in ICP and DCP in OCTA during one year. Thirdly, we followed up the patient weekly during the first 2 weeks and could closely monitor the acute change of the fundus, HVF, OCT and OCTA. The patient had vascular risk factors including hypertension. He also had luminal irregularity at bilateral distal ICAs. These factors might induce transient hypoperfusion in deep vascular complex. Transient increase of CWPs with inner retina’s hypoperfusion also supports the vascular etiology. The severity of retinal ischemia is determined by the severity of blood flow impairment; delayed or slow flow leads to PAMM; more severe obstruction of flow leads to combined middle/inner retinal infarction (i.e. partial arterial occlusion) [1].

In conclusion, we report a case of PAMM followed for one year for a patient having normal visual acuity. Even in the case with normal vision, PAMM could be considered as a differential diagnosis for a patient with paracentral scotoma. Caution and evaluation are required with combined systemic vascular risk factors.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.
References


Fig 1. Initial and follow-up findings of a 67-year old male with paracentral acute middle maculopathy in the left eye.

(A) At first visit, fundus photography (FP) showed perifoveal drusen in both eyes, some deep grayish faint lesions at parafoveal area (white arrows), and multiple cotton wool patches (CWPs) around the optic disc (black arrows) in the left eye. (B) At first visit, Humphrey visual field (HVF) showed focal inferior defects in the right eye and scattered defects in the left eye. (C) At first visit, spectral-domain (SD) optical coherence tomography (OCT) revealed no significant findings in the right eye and multiple hyper-reflective bands in inner nuclear layer (INL) in the left eye (white arrows). The position of the grayish lesions on fundus corresponded to the hyper-reflective INL lesions in the left eye. (D) At first visit, OCT angiography (OCTA) demonstrated relatively decreased vascular density of deep capillary plexus (DCP) in the left eye, compared to the right eye. (E-J) Follow up examinations of the left eye for one year with FP, HVF, SD-OCT, and OCTA; The OCTA device was changed from Deep Range Imaging (DRI) OCT triton (Topcon, Japan) to Spectralis OCTA (Heidelberg, Germany) after initial visit. (E) At one week follow-up, FP showed diminished parafoveal grayish lesions and increased CWPs around the optic disc in the left eye. HVF revealed decreased scattered defects compared to the previous test. OCT showed slightly decreased multiple hyper-reflective bands in INL (white arrows). (F) At two week follow-up, CWPs decreased. HVF improved. OCT showed decreased hyper-reflective bands in INL. (G) At two month follow-up, FP findings, HVF and OCT improved further. (H) At six month follow-up, CWPs almost disappeared (parafoveal yellowish changes were not the CWPs but the artifact due to the reflection). The scattered minimal defect was stationary in HVF. The multiple hyper-reflective INL lesions improved and focal INL thinning was observed (white dashed arrow). (I) At one year follow-up, FP was nearly stationary (parafoveal yellowish changes were not the CWPs but the artifact due to the reflection). HVF was unremarkable. INL thinning developed slightly further on OCT (white dashed arrow). (J) During the follow up, initial partial hypoperfusion of the intermediate capillary plexus (ICP) and deep capillary plexus (DCP) decreased gradually. The vascular density of the ICP and DCP was almost restored at one year follow-up. Informed consent was obtained from the patient for publication of this case report and relevant images.