CASE REPORT

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A Case of Self-Documented Paracentral Acute Middle Maculopathy After Cataract Surgery

Abstract

Background: *This report illustrates a case of paracentral acute middle maculopathy (PAMM) following routine cataract surgery. This case provides unique insight into the development and progression of the condition through extensive serial ophthalmic imaging.*

Case presentation: *Several hours after cataract surgery of his left eye, a 57-year-old ophthalmic photographer presented to the clinic with a left paracentral scotoma. Serial infrared reflectance (IR) and spectral domain optical coherence tomography (SD-OCT) images were obtained prior to, immediately following, and 10 days after the initial event.*

Conclusions: *Although the etiology of PAMM remains unknown, several precipitating events have been described in the literature. This is one of the first reported cases of PAMM temporally associated with cataract surgery. Given that two previous cases of PAMM were associated with systemic administration of epinephrine, intracameral epinephrine may have played a role in this case. However, the underlying cause remains to be determined.*

Introduction

Ophthalmic photography has long played a vital role in the documentation and diagnosis of many retinal abnormalities. Armed with sophisticated imaging technologies, ophthalmic photographers are often the first to encounter or recognize unique clinical findings and suggest a working diagnosis. This report describes a case of paracentral acute middle maculopathy (PAMM) occurring within hours of uncomplicated cataract surgery. This case is unique in that it affected an experienced ophthalmic photographer who self-documented his condition with infrared reflectance (IR) and spectral-domain optical coherence tomography (SD-OCT) images prior to surgery as well as numerous serial images following surgery.

It is not uncommon for ophthalmic photographers to image their own eyes as a test subject when trying new equipment, demonstrating techniques, or troubleshooting technical issues. In this case, baseline images had been self-captured while demonstrating the capabilities of the SD-OCT. In addition to this baseline, the photographer had been tracking serial progression of a subtle ERM prior to cataract surgery, and had proactively taken SD-OCT images one day prior to surgery as a reference against potential development of post-operative cystoid macular edema (Figure 1).

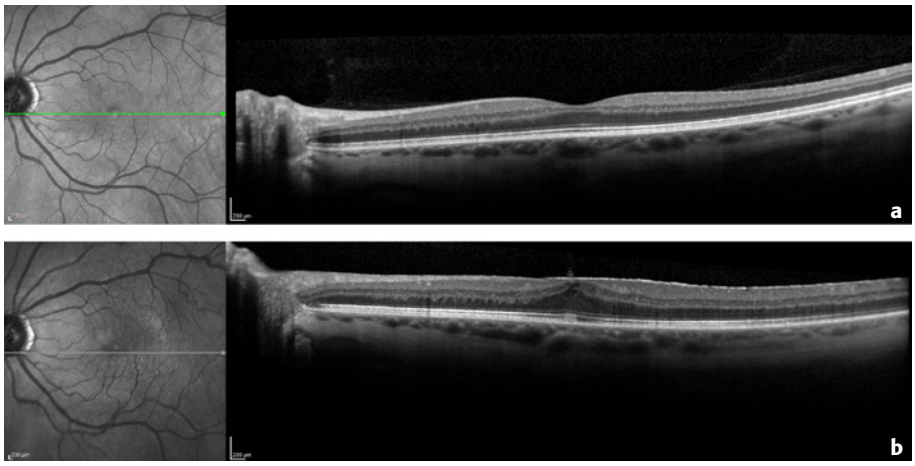

Figure 1: (a) Baseline SD-OCT image self-captured two years prior to surgery while demonstrating capabilities of the instrument. (b) Image captured by the photographer/patient of the same eye captured three days prior to cataract surgery as a pre-operative baseline.

Case Report

A 57-year-old Caucasian man presented with a complaint of sudden onset of a gray, crescent-shaped paracentral scotoma one day after undergoing uncomplicated cataract surgery on the left eye. Previous ocular history was significant for pigment dispersion syndrome and mild ocular hypertension. Eighteen months prior, a retinal tear with an associated vitreous hemorrhage in the operative eye was treated with pars plana vitrectomy and laser photocoagulation. The best corrected visual acuity (BCVA) at this time was 20/25. The BCVA immediately prior to cataract surgery was 20/40 with both cataract and mild epiretinal membrane (ERM) likely contributing to the decreased vision. Of note, ten days prior to cataract surgery, intraretinal fluid was noted in the left eye and treated with topical steroid drops. Past medical history was otherwise unremarkable.

One day following cataract surgery, BCVA was 20/20 in the right eye and 20/30 in the left eye. Intraocular pressure (IOP) was noted to 29 mm Hg in both eyes. Dilated funduscopic examination of the left eye revealed a mild and stable epiretinal membrane with new areas of retinal whitening in the temporal macula as shown in the color and red-free fundus photographs in Figure 2.

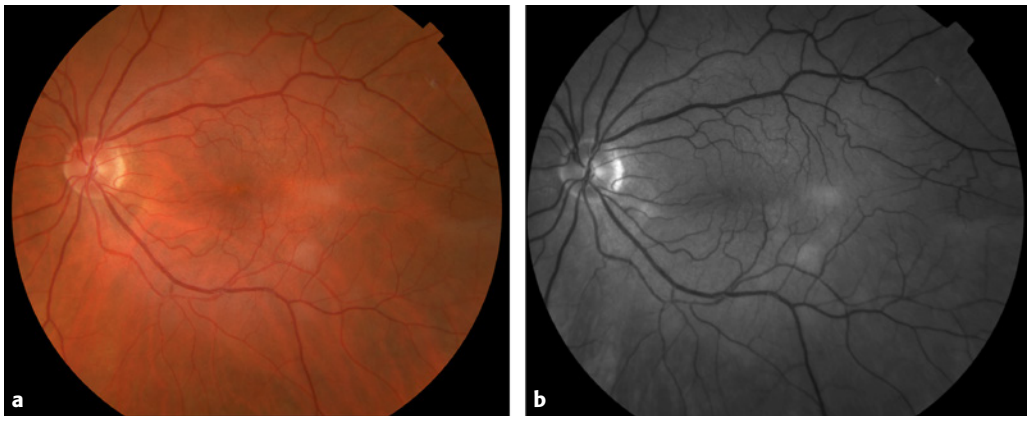

Figure 2: (a) Fundus photograph of the left eye taken one day after cataract surgery reveals a small arc of retinal whitening temporal to the fovea. (b) Red free fundus photograph highlights the same lesion. *Fundus photographs taken by James D. Strong, CRA, OCT-C, FOPS.*

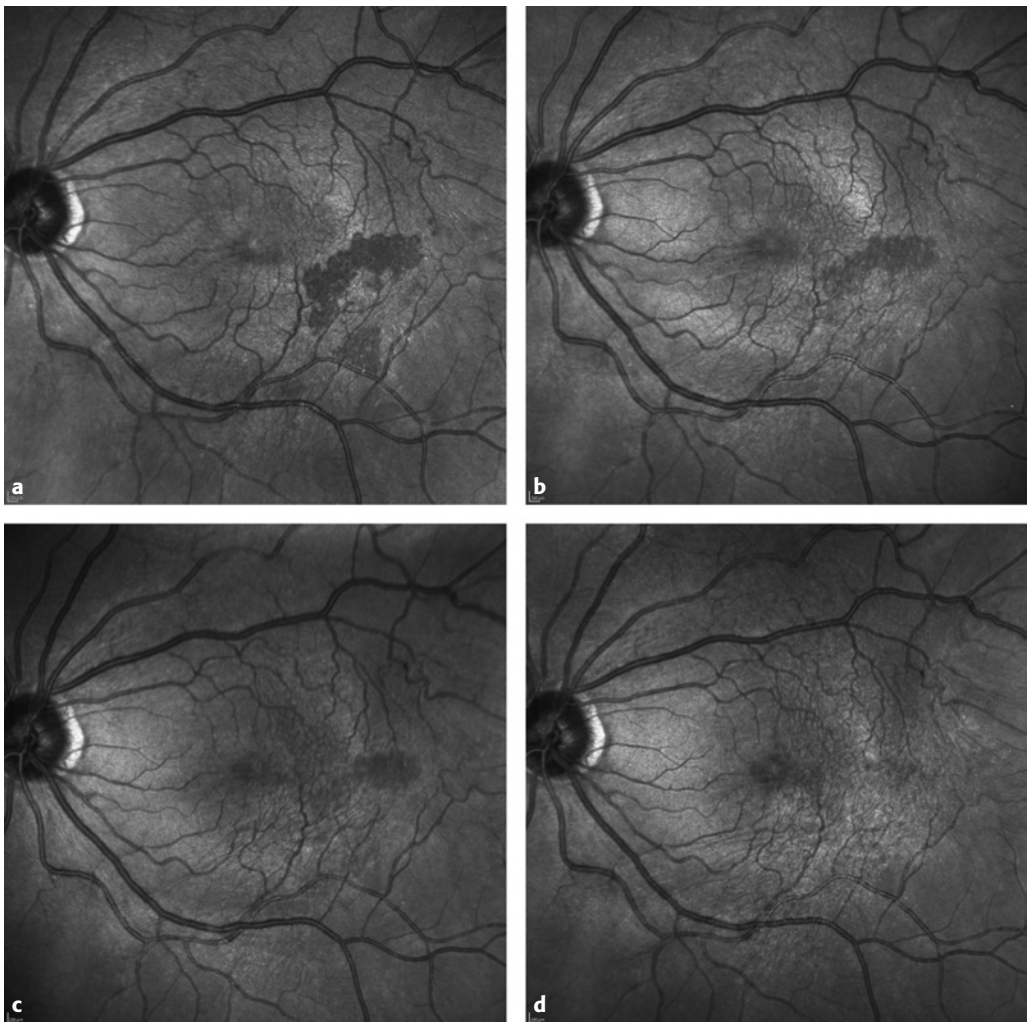

Figure 3: (a) Infrared reflectance cSLO photograph of fundus obtained on post-op day one showing a well-defined wedge-shaped hyporeflective lesion just temporal to the fovea in the left eye. Follow up IR images show a gradual decrease in the intensity of hyporeflectivity over the next several days until hyporeflectivity becomes nearly undetectable. (b-d) Post op days 3, 7, and 17 respectively.

Infrared reflectance confocal scanning laser ophthalmoscopy and spectral domain SD-OCT performed prior to surgery provided normal baseline images of the patient's retina. Infrared reflectance images obtained one day following surgery revealed a well-demarcated hyporeflective lesion (Figure 3a). Corresponding SD-OCT images demonstrated intraretinal hyperreflectivity that extended

from the inner plexiform layer (IPL) to the outer plexiform layer (OPL) of the retina in an area just temporal to the fovea (Figure 4a). The intensities of hyporeflectivity on cSLO and hyperreflectivity on SD-OCT decreased gradually over the next several days, becoming nearly undetectable by day 10 (Figures 3b-d, 4b-d).

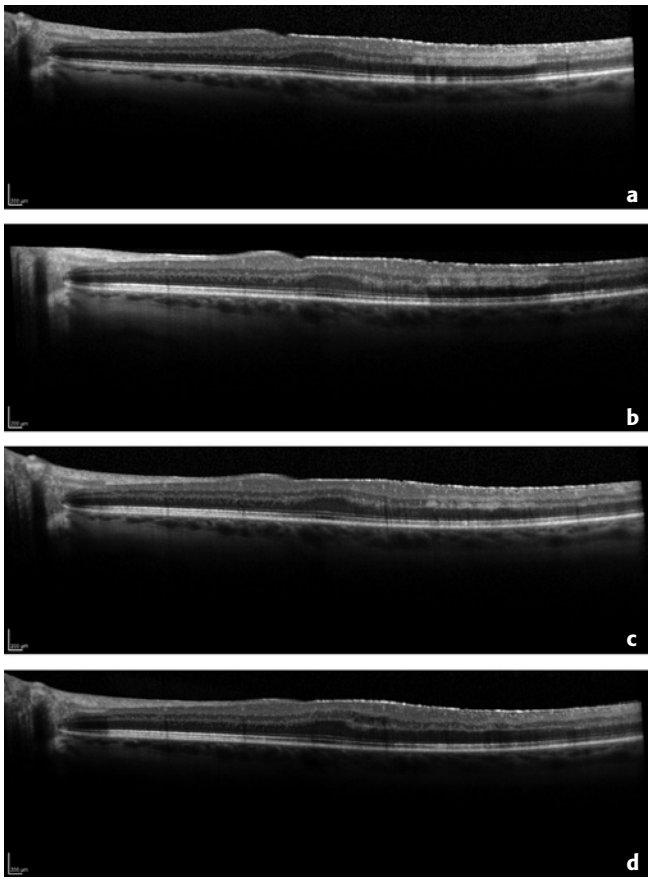

Figure 4: (a) SD-OCT shows an area of hyperreflectivity extending from the IPL to the OPL corresponding to the area of the lesion found by cSLO. (b-d) SD-OCT shows corresponding decrease in intensity of hyperreflectivity in area of lesion following the same time course as IR cSLO images.

Paracentral Acute Middle Maculopathy

Paracentral acute middle maculopathy (PAMM) is a condition that was first reported by Sarraf et al in 2013 as a rare variant of acute macular neuroretinopathy (AMN) detected with multimodal imaging.[1] PAMM typically presents with acute onset paracentral scotoma in the absence of other visual changes. Funduscopic findings tend to be subtle, with wedge-shaped grayish-white lesions appearing in the parafoveal region. These lesions are more apparent with multimodal imaging techniques including monochromatic red-free fundus photographs, confocal infrared reflectance imaging, and spectral domain optical coherence tomography (SD-OCT).

Subsequent reports now recognize PAMM as a distinct and independent entity characterized by SD-OCT hyperreflectivity of the inner nuclear layer (INL) of the retina that progresses to INL thinning.[2] It has been hypothesized that PAMM lesions result from ischemic injury to the deep capillary plexus of the retina, which supplies the inner plexiform, inner nuclear, and outer plexiform layers. This explanation has recently been substantiated by OCT angiography studies demonstrating deep capillary ischemia.[3,4] Several vascular risk factors for PAMM are well-recognized, including vasopressor exposure and retinal vascular diseases including diabetic retinopathy, Purtscher retinopathy, and others. In recent years, diverse additional associations with PAMM lesions have been reported, including pregnancy,[5] optic disk swelling and meningitis,[6] ingestion of synephrine-containing preworkout supplement,[7] and retinal artery occlusion following cataract surgery.[8]

Multimodal Imaging

PAMM is a condition that relies almost exclusively on modern imaging for detection. PAMM lesions aren't always apparent on examination, even in the acute phase. Visibility can be enhanced with short-wavelength monochromatic imaging using the fluorescein exciter filter (488 nm). This wavelength is more effective than the standard red-free (530-560 nm) filter to highlight areas of retinal whitening in artery occlusions or the gray-white PAMM lesions because of increased scatter of shorter wavelengths. Blue-green light is also more completely absorbed by retinal pigmentation and blood vessels, providing a darker background against which reflection and scattering in the anterior layers of the fundus is enhanced (Figure 5).

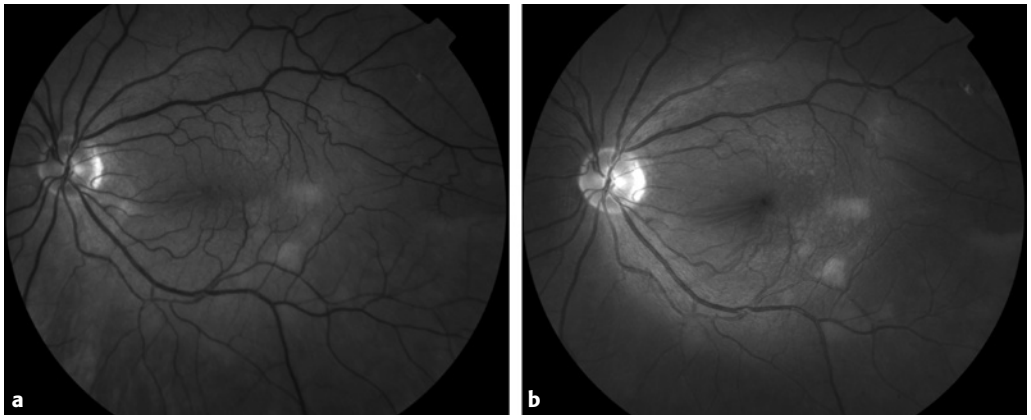

Figure 5: Monochromatic fundus photography comparison. (a) Standard monochromatic green "red free" photograph with green filter at 540 nm. (b)Monochromatic blue-green filter at 488 nm. Increased scatter at shorter wavelengths enhances the visibility of areas of ischemic retinal whitening. *Fundus photographs taken by James D. Strong, CRA, OCT-C, FOPS.*

The combination of confocal NIR imaging and SD-OCT provided by the

Heidelberg Spectralis (Heidelberg Engineering, Heidelberg Germany) is particularly useful in identifying and tracking progression of PAMM lesions. PAMM is now a far more common finding than originally suggested by the early reports, in part because of increased use of multimodal imaging and recognition of the clinical findings.

Infrared reflectance imaging is the best modality to delineate the borders of PAMM lesions in the acute phase. The reason that characteristic wedge-shaped PAMM lesions appear dark in infrared reflectance isn't completely understood, but may be related to the confocal tonal shift that occurs as a result of suppression of scattered light by the confocal aperture in the cSLO system. Photographers often see this tonal shift in real time while adjusting the focus of the cSLO. It is especially apparent with elevated lesions that fall outside the plane of focus, but can also occur when ocular tissues are slightly thickened and scatter reflected light.[9]

The SD-OCT finding in PAMM is typically described as a band-like hyperreflective plaque at the level of the inner nuclear layer that corresponds to the gray IR lesion. Several reports in the literature describe the value of the registration feature specific to Heidelberg Spectralis, which allows each OCT B-scan to be matched to its exact location on the accompanying NIR fundus image, facilitating accurate correlation of the retinal findings between each of these modalities.[1,10] With this system, baseline OCT scans can also be used as a reference for accurate registration of serial OCT imaging. The instrument is able to identify retinal landmarks in a reference image and place the scan pattern in the same location at each subsequent visit. In the case presented here, the PAMM lesion occurred in an area where baseline reference scans had been captured previously. This coincidence facilitated serial self documentation by the patient (Figure 6).

Fluorescein angiography has not proven to be useful in identifying PAMM lesions or ischemia of the deep capillary plexus.[2,11-13]

Discussion

This case provides unique documentation of the onset and evolution of PAMM lesions. The hyperreflective changes on SD-OCT and hypo-reflectivity on cSLO appeared simultaneously with the onset of the subjective scotoma on post-op day one. These changes were resolved by day ten, confirming the short duration of these findings presented in other cases.[1,13-15] A scotoma corresponding to the area of the lesion observed on cSLO and SD-OCT images was detectable by HVF testing at post-op day eight and remained at 60 weeks.

At the time this case occurred, PAMM was a newly described condition and there were no reports associated with uncomplicated cataract surgery. Since then, Creese and associates have published a series of ten cases of severe vison loss after cataract surgery with SD-OCT findings they describe as consistent with PAMM.[8] All patients had permanent vision loss with final acuity of less than 6/60. Three cases were diagnosed with a central retinal artery occlusion (CRAO), while the remaining seven were attributed to transient retinal artery occlusions.

The article included two "illustrative cases" from the series which demonstrated the clinical appearance of CRAO. Although there was demonstrated hyperreflectivity of the INL on OCT, it was not localized like often found in PAMM. The hyperreflectivity occurred both nasal and temporal to the fovea and appeared more consistent with the prominent middle limiting membrane sign (p-MLM) described by Chu and associates in 2013.[16] There was also increased thickness of the inner retinal layers on OCT. The accompanying IR images showed a subtle diffuse gray area often seen with increased retinal thickness in CRAO rather than the well circumscribed wedge-shaped lesions originally described as an important feature of PAMM (Figure 7). Given the differences in multimodal findings and visual outcome compared to this series, we believe our case represents a different process of transient retinal capillary ischemia after cataract surgery and is more representative of the original description of PAMM. Unlike the severe vision loss described in the Creese series, vision in this case eventually stabilized at 20/25 with a very mild persistent scotoma.

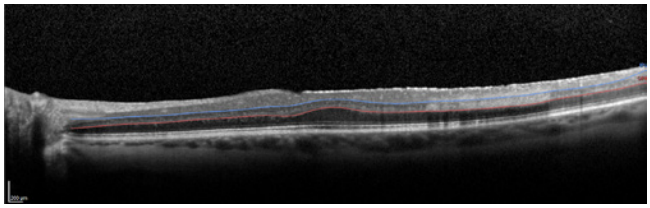

Figure 6: SD-OCT shows the classic area of hyperreflectivity extending from the inner plexiform layer (IPL) to the outer plexiform layer (OPL) found in PAMM. Note the shadowing that occurs posterior to the hyperreflectivity of the inner nuclear layer. In this case, the PAMM lesion coincidentally occurred in the same parafoveal location where baseline SD-OCT images had been previously captured.

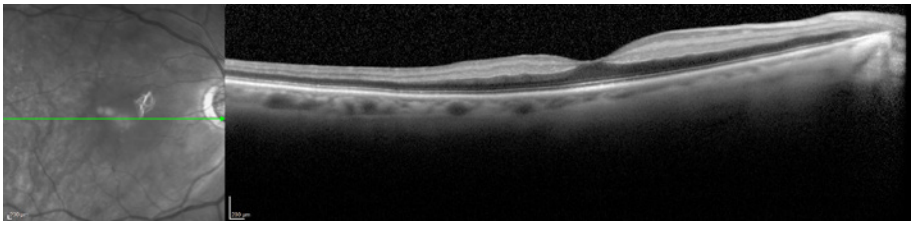

Figure 7: A typical example of a central retinal artery occlusion (CRAO) demonstrating hyperreflectivity of the inner nuclear layer throughout the macula along with increased thickening of the anterior retinal layers and a prominent middle limiting membrane (p-MLM) sign at the margin of the OPL. The accompanying IR image shows a diffuse gray area, rather than the sharply outlined dark gray lesions seen in PAMM. CRAO findings like this have sometimes been labelled as PAMM despite clear differences in appearance of both SD-OCT and IR images.

These differences illustrate the confusion that may result from the changing definition of PAMM since its initial description. PAMM is now considered a clinical finding rather than a disease. Referring to this finding as INL hyperreflectivity, rather than PAMM, seems more obvious and descriptive when referring to this OCT finding in isolation.

Conclusions

We believe this to be the first reported case of PAMM after uncomplicated cataract surgery. The etiology of PAMM in this case still remains to be elucidated. The patient does not have the vascular risk factors[1,17] that typically contribute to PAMM. However, ERM-associated cystoid macular edema and resultant traction on inner retinal vasculature may have played a role in the development of the condition. Additionally, it is worthwhile to consider contributory factors that have been linked to AMN as potentially playing similar roles in PAMM pathogenesis due to the prior identification of cases resembling PAMM as cases of AMN. Several case reports document systemic sympathomimetics playing a role in AMN pathogenesis,[18-21] and another report describes a case of AMN following an intranasal injection of epinephrine during rhinoplasty.[22] It is also possible that the microvascular stress induced by the ERM was exacerbated by the presence of epinephrine in the infusion bottle during surgery.

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