Evaluation of Paracentral Acute Middle Maculopathy with Multimodal Imaging in a Pregnant Patient

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ABSTRACT

Paracentral acute middle maculopathy is a relatively recently described, self limiting clinical entity which is characterized with changes in the middle retinal layers on optical coherence tomography. Many risk factors have been described such as dehydration, oral contraceptive use, caffeine and vasopressors. Paracentral acute middle maculopathy can accompany many retinal vascular diseases such as diabetic retinopathy, hypertensive retinopathy, Purtscher retinopathy, retinal artery and vein occlusion. Patients generally present with scotoma, however they may be asymptomatic as well. In this paper, paracentral acute middle maculopathy in an otherwise healthy 33-year old pregnant patient in third trimester of pregnancy was described with multimodal imaging including optical coherence tomography, near infrared reflectance and fundus autofluorescence.

Keywords: Scotoma, Pregnancy.

INTRODUCTION

Paracentral acute middle maculopathy (PAMM) was first described as a variant of acute macular neuroretinopathy by Sarraf et al. in 2013.[1] It typically affects middle layers of retina including inner nuclear layer and outer plexiform layer in accordance to its description. The patients with PAMM generally presents with acute paracentral scotoma and visual acuity is found to be normal or mildly affected.[1, 2] In the etiopathogenesis, several risk factors such as dehydration, oral contraceptive use, caffeine and vasopressor agents.[1, 3] In addition, it is seen in many retinal vascular diseases such as diabetic retinopathy, hypertensive retinopathy, sickle cell anemia-related retinopathy, Purtscher retinopathy and retinal artery and vein occlusions.[3]

CASE REPORT

A 33-years old woman presented with central scotoma occurred 2 days ago. In her history, there was no systemic or ocular disease other than pregnancy (gestational age: 27 weeks) and she was receiving prenatal vitamin preparation. In the examination, best-corrected visual acuity was 1.0 in both eyes. Intraocular pressure was measured as 12 mmHg in the right eye and 13 mmHg in the left eye. Fundus examination and dilated fundus examination were normal in both eyes. On optical coherence tomography, focal hyper-reflective area was observed at level of inner nuclear layer and outer plexiform layer in the right eye. In the same area, hypo-reflectance on near-infrared reflectance (NIR) image and hypo-autoflorescence on fundus auto-fluorescein (FAF) image were observed (Figure 1). As the patient was pregnant (gestational age: 27 weeks) fundus fluorescein angiography could not be performed; thus, follow-up was scheduled with diagnosis of paracentral acute middle maculopathy.

In control visit on month 1, it was found that scotoma complaint was improved and hyper-reflective area was faded on OCT. the hypo-reflectance are on NIR was almost disappeared with regression in hypo-autoflorescence on FAF (Figure 2)

In control visit on month 3, scotoma complaint was fully recovered. OCT, NIR and FAF images returned to normal Figure 3).

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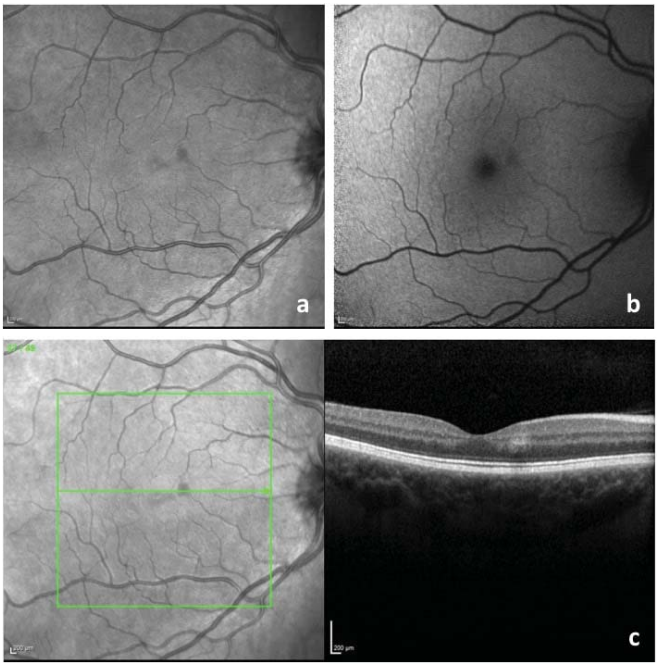

Figure 1: a) *Hypo-reflectance on near-infrared reflectance (NIR) imaging of right eye;* **b)** *hypo-autoflorescence on fundus auto-fluorescein (FAF) image at same region;* **c)** *focal hyper-reflective appearance at level of inner nuclear layer and outer plexiform later at juxtafoveal region on spectral domain-optical coherence tomography (SD-OCT).*

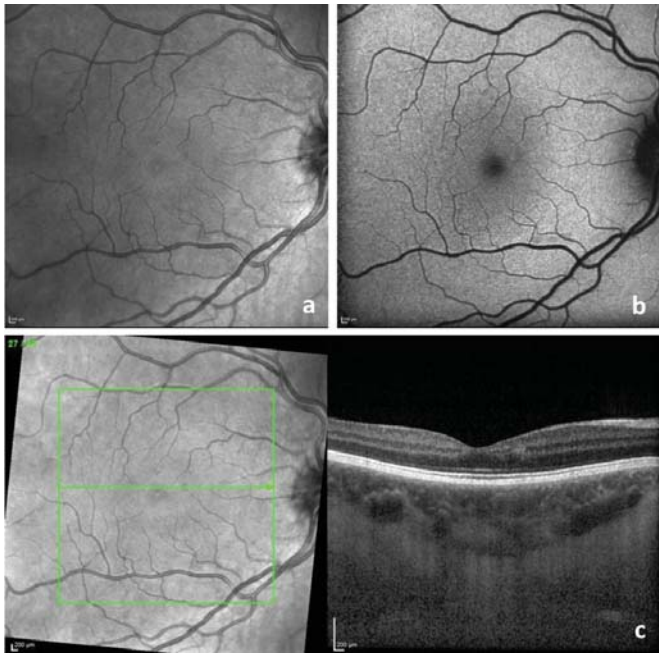

Figure 2: *month 1;* **a)** *decreased hypo-reflectance on NIR imaging;* **b)** *decreased hypo-autoflorescence on FAF imaging;* **c)** *faded hyper-reflective area observed on OCT.*

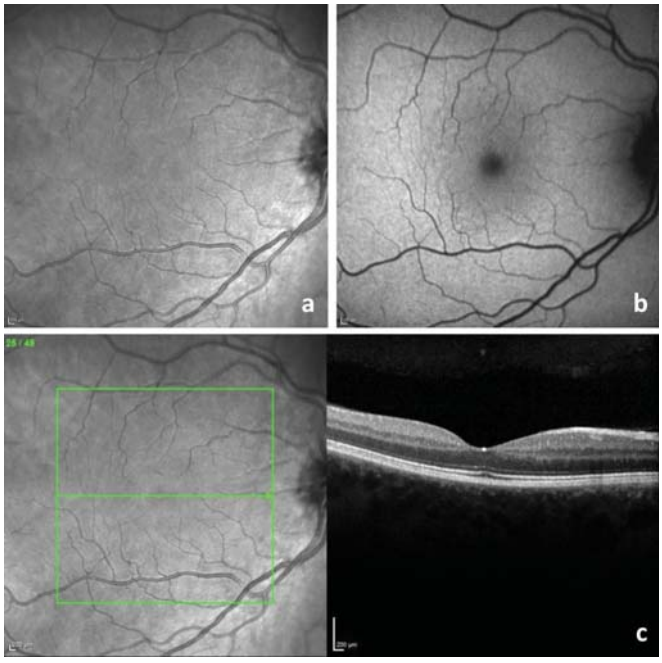

Figure 3: *month 3; Normal* **a)** *NIR* **b)** *FAF and* **c)** *OCT images.*

The patient gave written informed consent for use of photographs and documents for educational and scientific purposes (including scientific publications).

DISCUSSION

Paracentral acute middle maculopathy (PAMM) was first described as a variant of acute macular neuroretinopathy by Sarraf et al. using spectral domain-OCT in 2013.[1] In a study on postmortem donor human retina, Tan et al. identified 4 capillary plexuses including at nerve fiber layer, retinal ganglion cell layer, junction of inner plexiform and inner nuclear layers (superficial capillary plexus) and junction of deep inner nuclear layer and outer plexiform layer (deep capillary plexus).[4] PAMM results from intermediate and deep retinal capillary ischemia, which could be shown more clearly by introduction of optical coherence tomography angiography in clinical practice.[3,5] In the literature, two forms of paracentral acute middle maculopathy have been described: type 1 and type 2.[1] Type 1 PAMM includes cases in which structures above outer plexiform layer, superficial or intermediate capillary plexus, are involved while type 2 PAMM includes cases in which structures below outer plexiform layer, deep capillary plexus, are involved.

As it was the case in our patient, fundus examination can fail to confirm the diagnosis, at this point, multi-modal imaging, particularly near-infrared reflectance and spectral domain-OCT, can be diagnostic. There is no established treatment in paracentral acute middle maculopathy;

however, it is recommended to identify and treat risk factors such as dehydration, caffeine or epinephrine. It is though that pregnancy and oral contraceptive use can lead paracentral acute middle maculopathy by predisposing hyper- coagulability through hormonal changes resulting from a common mechanism in both entities.[5, 6]

In the literature, paracentral acute middle maculopathy was previously shown in 2 pregnant women at first and second trimester, respectively. Our patient is the first case in which PAMM was detected in the third trimester of pregnancy.[5,6] Consultation was ordered to exclude underlying causes such as diabetes mellitus, hypertension and thrombophilia; however, no pathology was detected. Inner nuclear layer atrophy was lacking in our patient, which is seen in majority of cases with PAMM during follow-up and is thought to be as a result of reperfusion injury.

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