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Optical Coherence Tomography Angiography of Acute Macular Neuroretinopathy/Paracentral Acute Middle Maculopathy

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Keywords

AMN; OCT-A; oct angiography; PAMM; acute macular neuroretinopathy; paracentral acute middle maculopathy

Acute macular neuroretinopathy (AMN) has been characterized as wedge-shaped superficial retinal defects in the macula, resulting in paracentral scotomas. AMN has been associated with vasoconstrictors and sympathomimetics.¹ In 2013, paracentral acute middle maculopathy (PAMM) was described as an AMN variant, seen as a hyperreflective band involving the middle retinal layers on spectral-domain optical coherence tomography (SD-OCT).¹ Type 1 PAMM lesions involve layers above the outer plexiform layer (OPL), reflecting superficial or intermediate capillary plexus occlusion; type 2 lesions involve layers below the OPL, affecting the deep capillary plexus.¹

Macular lesions in AMN/PAMM are best visualized on near-infrared reflectance (NIR) imaging, correlating with SD-OCT OPL hyperreflectivity^{2,3} and may be otherwise subclinical. Definitive evidence of the ischemic origin of PAMM has been lacking. Only one case report has demonstrated a transient retinal artery occlusion on fluorescein angiography (FA).⁴ OCT angiography (OCT-A) provides novel retinal vascular flow visualization and provides a unique opportunity to visualize the depth of a vascular defect.⁵ Here we report the noninvasive ability of OCT-A to visualize a capillary perfusion defect in PAMM/AMN.

REPORT OF A CASE

A pregnant 29 year-old female at 21 weeks gestational age presented for evaluation of a paracentral scotoma in her right eye. One day prior, she experienced a presyncopal episode after noticing the scotoma. She was treated with intravenous fluids for hypotension (blood pressure: 77/50 mmHg) and was slightly anemic. Besides her pregnancy, she had no other past medical or ocular history. Her only medication was a prenatal vitamin. She denied illicit drug use and excess caffeine consumption.

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On examination, her visual acuity with correction is 20/20-1 and 20/20 in the right and left eyes, respectively. Anterior and posterior segment exam was unremarkable. SD-OCT of the right eye revealed hyperreflectivity of the inner plexiform layer (IPL), inner nuclear layer (INL) and OPL with a corresponding ovoid-shaped dark gray lesion seen on NIR imaging paracentral to the fovea (Figure 1A–C). Given the pregnancy, FA was deferred. Instead, OCTA was performed and revealed a deep retinal capillary plexus perfusion defect that persisted over time, congruent with her persistent scotoma (Figure 2). Following observation, this lesion evolved to INL atrophy at 1-month suggestive of focal middle retinal infarction (Figure 1F). In collaboration with her obstetrician, efforts were made to avoid hypotension during her delivery.

DISCUSSION

The typical findings of a wedge-shaped lesion on NIR imaging and hyperreflective middle retinal layers on SD-OCT have consistently been demonstrated in AMN/PAMM, as well as other retinal vasculopathies, which are thought to reflect focal intraretinal nonperfusion.⁵ This case appears unique as a type 1 PAMM lesion, as prior cases were mostly seen in older males.¹ Previously described AMN/PAMM cases were related to hypertensive- or vasoconstrictive-inducing substances. In this case, the combined hypotension/anemia as well as hypercoagulable state of pregnancy may have contributed to the ischemic event.

OCT-A findings of capillary nonperfusion in this case correlated with subsequent INL atrophy, typical of type 1 PAMM lesions. OCT-A is a rapidly emerging technology, though at this time, knowledge remains limited regarding OCT-A implications. In our review of the literature, no previous reports of the use of OCT-A in AMN or PAMM are identified.

OCT-A may be a useful adjunct to the multimodal imaging of AMN/PAMM and may enhance visualization of the retinal microvasculature perfusion at a retinal layer level. The ability of OCT-A to differentiate laminar focal capillary defects that maybe undetected by FA is a potential significant advantage of OCT-A. As in this case, OCT-A may also be particularly helpful where conventional FA may not be preferred, such as pregnancy or allergy. Further studies are warranted to confirm these observations.

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Figure 1.

A–C. Initial presentation. Dark lesion (arrow) on NIR imaging in superotemporal macula (A,B) and corresponding focal hyperreflectivity (C, arrow). **D–F. One month later.** Resolution of lesion (arrow) on NIR imaging (D,E) and corresponding INL thinning (F, arrow).

Figure 2. OCT-A reveals outer retinal capillary nonperfusion (A, arrow) at location of lesion in affected eye and intact outer retinal capillary perfusion (C) in unaffected eye. Corresponding OCT (B, D) revealing OCT-A segmentation (between red and green lines).