

Paracentral Acute Middle Maculopathy and Acute Macular Neuroretinopathy

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ABSTRACT: Type 1 acute macular neuroretinopathy (AMN), also known as paracentral acute middle maculopathy (PAMM), is a novel form of AMN in which spectral-domain optical coherence tomography (SD-OCT) demonstrates a characteristic hyper-reflective band-like lesion at the level of the inner nuclear layer. This form of AMN has only recently been described in five patients who were predominantly older men with vasculopathy. Herein, we describe a young, healthy female patient with classic features of type 1 AMN or PAMM with SD-OCT.

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INTRODUCTION

Recently, two forms of acute macular neuroretinopathy (AMN) have been defined by Sarraf et al based on characteristic features with spectral-domain optical coherence tomography (SD-OCT).^{1,2} Type 1 AMN, also referred to as paracentral acute middle maculopathy (PAMM), was characterized by SD-OCT abnormalities at the level of the inner nuclear layer (INL), whereas type 2 AMN was associated with SD-OCT findings of outer macular involvement similar to that recently described by Fawzi et al.³

Whereas type 2 AMN patients fit the original description of young, otherwise healthy women,⁴ type 1 AMN patients were predominantly older men with vasculopathic risk factors, which raised the question of whether the latter form may represent a new disease entity.^{1,2} Herein, we describe the first case report, to our knowledge, of a type 1 AMN lesion in a young, otherwise healthy woman.

CASE REPORT

A 32-year-old woman was referred for a second opinion and complained of a wedge-shaped paracentral scotoma of 1-week duration affecting the left eye. Medical history was entirely unremarkable, and ocular history was remarkable only for bilateral LASIK surgery. The patient reported drinking two to three cups of coffee per day.

Visual acuity (VA) was 20/20 in the right eye and 20/80 in the left eye. Slit-lamp examination of the anterior segment was unremarkable. Ophthalmoscopic examination of the retina was significant for a parafoveal wedge-shaped lesion in the left eye (Figure 1, page S34) and was within normal limits in the right eye. An unrelated small chorioretinal scar was noted inferotemporal in the macula in the left eye. Near infrared reflectance (NIR) demonstrated a dark grey, wedge-shaped lesion paracentral in the left eye (Figure 2A, page S34). Registered SD-OCT showed a characteristic band-like hyper-reflective lesion at the level of the inner nuclear layer (INL) extending from the outer plexiform layer (OPL) to the inner plexiform layer in a paracentral location consistent with type 1 AMN or PAMM (Figure 2B, page S34).

The paracentral scotoma improved, and VA rapidly normalized to 20/20 in the left eye over the next month, with gradual resolution of the NIR

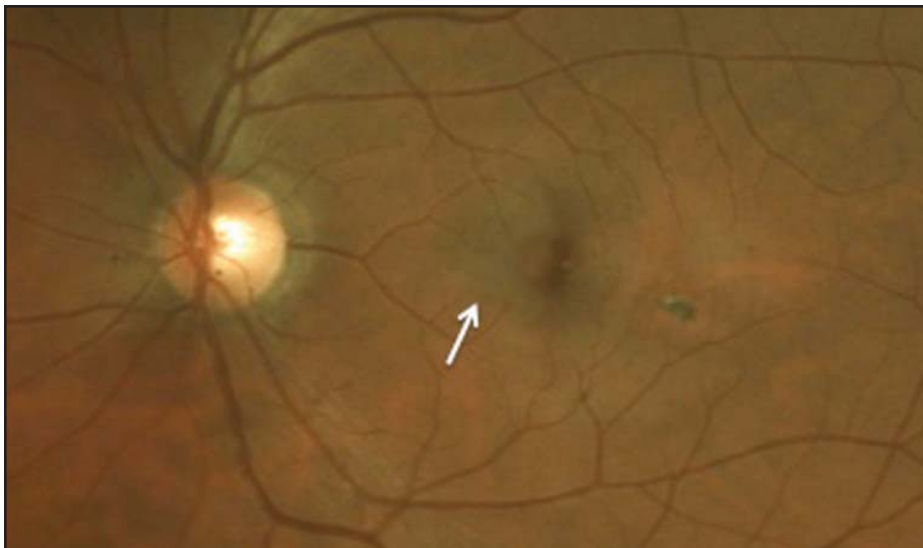


Figure 1. Optos color photograph of the left eye at presentation showing grey-white, wedge-shaped lesion (white arrow). Note old chorioretinal scar in temporal macula.

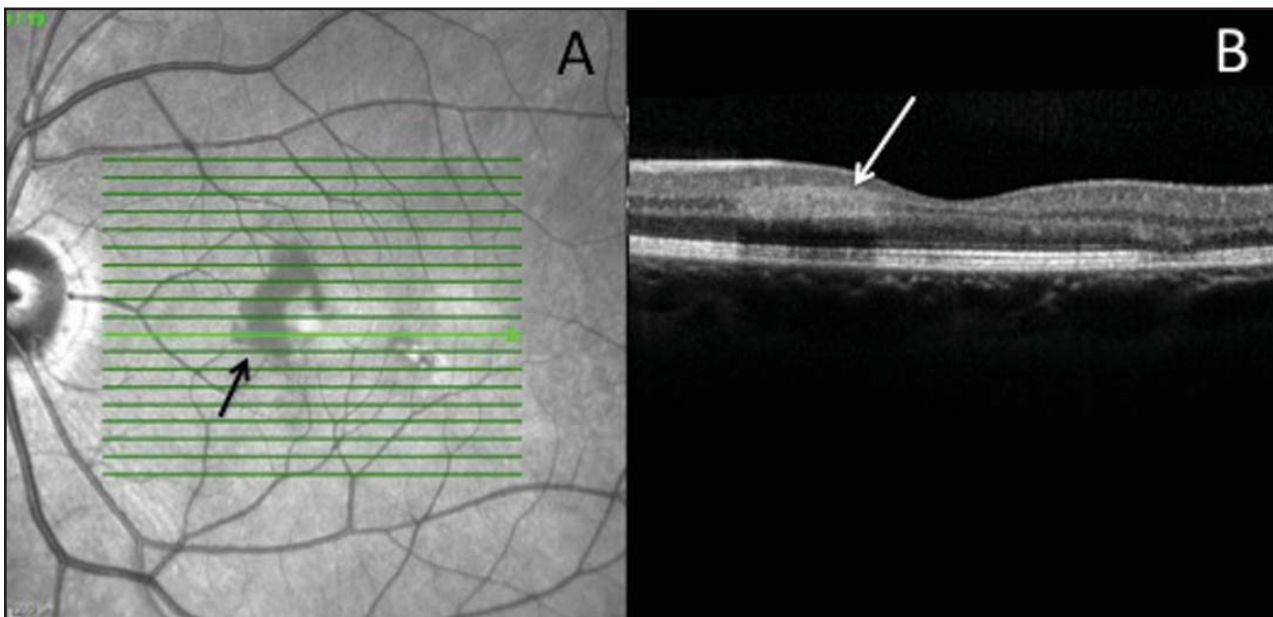


Figure 2. Near infrared reflectance (NIR) and spectral-domain optical coherence tomography (SD-OCT) images of the left eye at presentation. (A) NIR image shows a paracentral arcuate, wedge-shaped, dark grey lesion affecting the nasal fovea of the left eye. An unrelated small chorioretinal scar is present inferotemporal in the left eye. (B) SD-OCT registered with the NIR lesion shows a characteristic band-like hyper-reflective plaque at the level of the inner nuclear layer extending from the outer plexiform layer to the inner plexiform layer in a paracentral location (white arrow) consistent with paracentral acute middle maculopathy.

lesion. At the time of her last visit, 3 months later, SD-OCT showed resolution of the hyper-reflective INL lesion, with subsequent thinning of the INL and attenuation of the OPL band (Figure 3).

DISCUSSION

Type 1 AMN, also referred to as PAMM, is a novel form of AMN only recently described in the literature in five patients.¹ It is typified by a paracentral hyper-reflective band-like lesion at the level of the INL on SD-OCT, which leads to INL

thinning. Our case of type 1 AMN is unique because the patient was a young, otherwise healthy woman, which to our knowledge is a patient profile that has only been reported in type 2 AMN.

AMN has been associated with vasoconstrictors, such as caffeine and epinephrine, suggesting an underlying ischemic etiology.^{5,6} The pathogenesis of type 1 and type 2 AMN is postulated to be related to the anatomy of the retinal circulation.^{1,2} Whereas type 1 AMN and PAMM lesions are precisely aligned with the deep capillary plexus (DCP) of the macula,

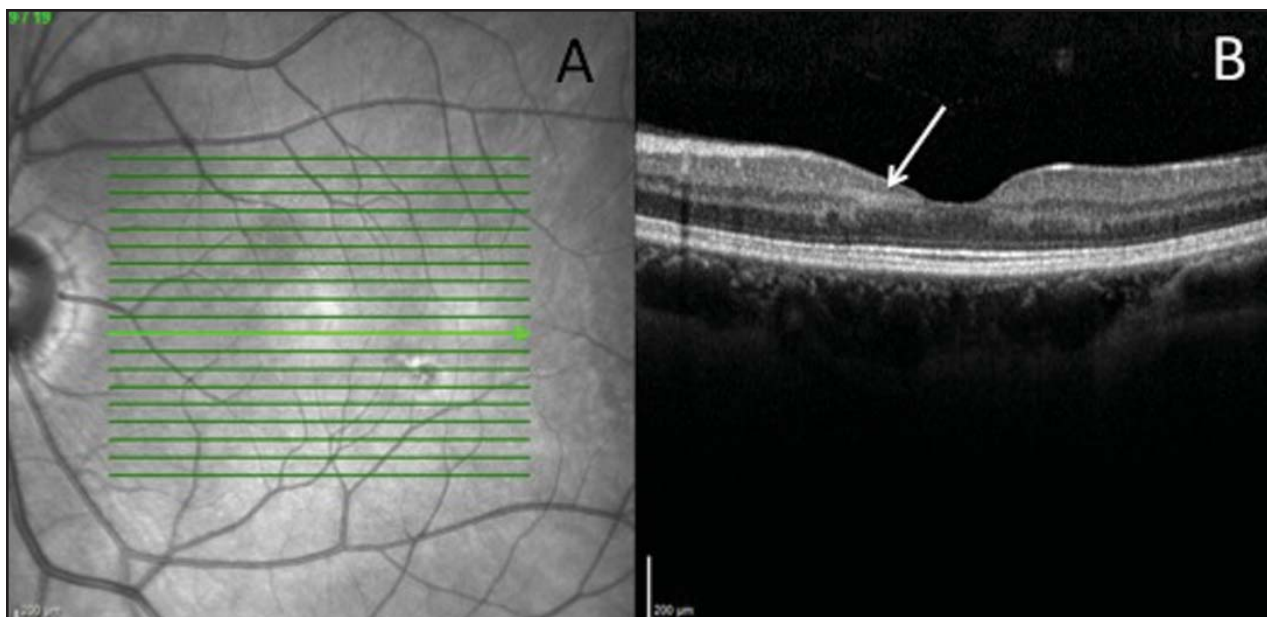


Figure 3. Near infrared reflectance (NIR) and spectral-domain optical coherence tomography (SD-OCT) images of the left eye 3 months after presentation. (A) NIR image shows resolution of the dark grey paracentral lesion. (B) SD-OCT shows subsequent thinning of the inner nuclear layer and attenuation of the outer plexiform layer band (white arrow).

type 2 AMN lesions colocalize with the outer portion of the DCP. The DCP contributes 10% of the oxygen supply to photoreceptors and may explain the loss of the ellipsoid zone and subsequent thinning of the outer nuclear layer in these type 2 cases.^{1,3}

This case report bridges the two sub-types of AMN recently proposed in the literature. Whereas type 1 AMN or PAMM was originally described in predominantly older men with vasculopathy, our case demonstrates that this form of AMN may have an identical demographic to the type 2 or classic AMN demographic of a young, otherwise healthy woman. As more index cases are studied in the future with advancing multimodal imaging systems, we may be able to determine whether type 1 AMN or PAMM and type 2 AMN represent single or multiple disease entities.

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