

PARACENTRAL ACUTE MIDDLE MACULOPATHY IN BIRDSHOT CHORIORETINOPATHY: A NOVEL ASSOCIATION

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Background/Purpose: To present a novel case of paracentral acute middle maculopathy in association with birdshot chorioretinopathy.

Methods: Case report.

Results: A patient presented with decreased vision and findings of uveitis, vasculitis and a paracentral scotoma. Multimodal imaging was consistent with paracentral acute middle maculopathy in the setting of a multifocal chorioretinopathy and a laboratory workup was consistent with birdshot chorioretinopathy.

Conclusion: Paracentral acute middle maculopathy is a newly described entity that may be associated with birdshot chorioretinopathy, and clinicians should be aware of it when patients present with new paracentral scotoma and signs of intraocular inflammation.

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Paracentral acute middle maculopathy (PAMM) is a newly recognized condition initially reported in 2013 as a variant of acute macular neuroretinopathy (AMN) with a proposed mechanism related to ischemia at the level of the superficial and deep retinal capillary plexi.¹ However, more recently, it has been described in association with other conditions of ischemic etiology.² We describe a novel case of PAMM in a patient with concurrent birdshot chorioretinopathy (BSCR).

Case Report

A 55-year-old white female presented for sudden onset sharply demarcated paracentral scotoma right eye, she reported a history of floaters in both eyes for the past year, cataract surgery in both eyes approximately 2 months before, and mild amblyopia left eye. Her

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review of systems was negative, her medical history was only significant for hypertension for which she took metoprolol and aspirin, and had not been using any over-the-counter medications. Visual acuities were 20/25 right eye and 20/50 left eye. Tenotomy and anterior segment examination were normal.

Funduscopy was significant for a well-demarcated, faint whitening of the macula just superotemporal to the foveal right eye (arrows) and also mild–moderate vitritis and attenuation of retinal arterioles in both eyes (Figure 1A); the periphery showed deep, faint white lesions (arrows) in a radial configuration (Figure 2A). Fluorescein angiography (early and late frames) showed some staining of the superotemporal arterioles and also both superior and inferior temporal venules, and also staining of leakage of the optic nerve without significant macular leakage (Figure 1, B and C). Indocyanine green angiography demonstrated multiple peripheral hypofluorescent lesions (arrows) in greater quantity than were apparent clinically or on fluorescein angiography (Figure 2B). Optical coherence tomography demonstrated a partial posterior vitreous detachment, some vitreous cells, and also a thickened and hyperreflective area involving the inner nuclear layer and outer plexiform layer (arrows) associated with posterior shadowing corresponding to the lesion seen on funduscopy (Figure 3, A and B). She was initially diagnosed with PAMM right eye and vitritis with retinal vasculitis both eyes. Oral prednisone was initiated. Laboratory testing was significant for positive HLA-A29. She was diagnosed with PAMM in the setting of BSCR. She was started on steroid sparing systemic immunomodulators and tapered off of prednisone; her PAMM lesion resolved with subtle focal atrophy (arrow) in the setting of epiretinal membrane (Figure 3C).

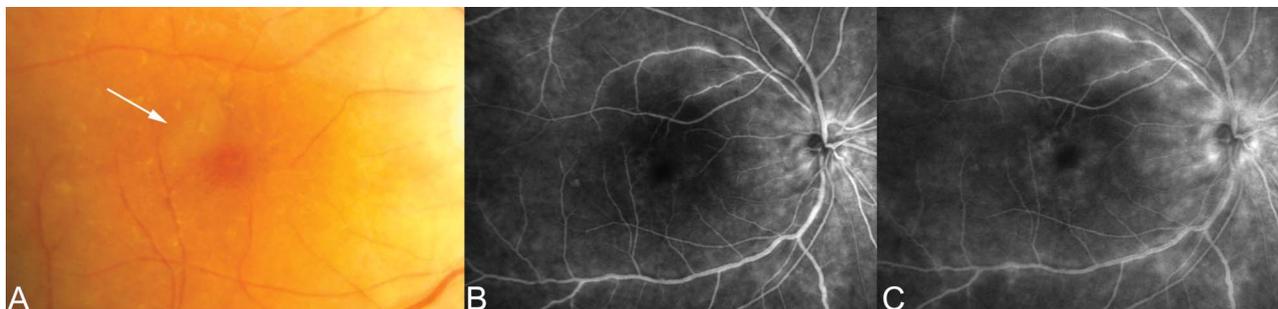


Fig. 1. Color and fluorescein angiography photographs of the right macula. **A.** Color fundus photograph of the right macula demonstrating focal whitening on the superotemporal margin of the fovea (arrow). **B.** Midphase fluorescein angiography of the right eye demonstrating staining of the vessels. **C.** Late-phase fluorescein angiography of the right eye demonstrating staining of the vessels without significant leakage.

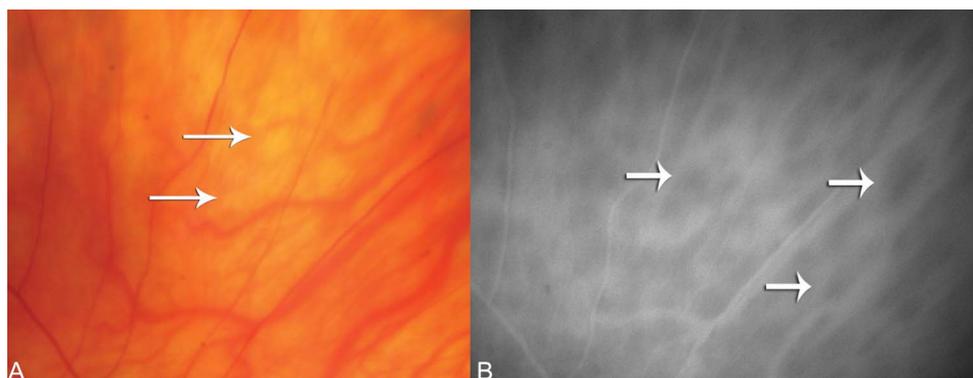


Fig. 2. **A.** Peripheral color and **(B)** indocyanine green angiography photographs with arrows indicating choroidal lesions.

Discussion

Paracentral acute middle maculopathy was initially described as an ischemic variant of AMN. In Saraff & Rahimy’s article identifying PAMM, PAMM was a unilateral condition, whereas AMN has been described as either unilateral or bilateral. Two subtypes of PAMM have been observed: Type 1 is located in the outer plexiform layer and inner nuclear layer, whereas Type 2 is deeper and involves the outer plexiform layer to the retinal pigment epithelium (RPE).¹ Both types result in a paracentral scotoma.¹ Type 1 is newly described, whereas Type 2 is consistent with previous reports of

AMN.³ Possible risk factors for PAMM in the setting of AMN that have been identified include caffeine, blood loss, viral prodrome, and toxemia of pregnancy.¹

Birdshot chorioretinopathy is described as bilateral inflammatory disease. Complications include vitritis, cystoid macular edema, epiretinal membrane, retinal neovascularization with or without vitreous hemorrhage, macular or peripapillary subretinal neovascularization, and optic atrophy.⁴ Acute macular neuroretinopathy or PAMM has not been previously reported as a complication in BSCR. Birdshot chorioretinopathy is believed to be due to a granulomatous inflammatory process affecting both the choroid and retinal vessels.⁵ Early

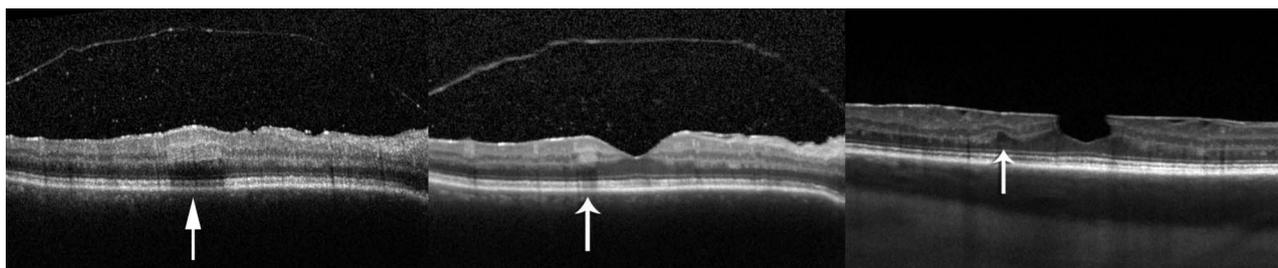


Fig. 3. Optical coherence tomography of the right macula. Left, optical coherence tomography line scan at initial presentation through the center of the lesion (arrow) showing thickening and hyperreflectivity at the level of inner nuclear and outer plexiform layers with posterior shadowing. Middle, optical coherence tomography line scan at initial presentation through the fovea showing the lesion (arrow) with similar characteristics. Right, optical coherence tomography line scan at follow-up showing resolution of thickening and hyperreflective lesion with interval development of inner nuclear and outer plexiform atrophy (arrow) and epiretinal membrane.

in the course of BSCR, the macula is often unaffected; however, optical coherence tomography of peripheral lesions demonstrates atrophy of the both the outer retina and choroid.⁶

We believe this presentation is consistent with Type 1 PAMM involving the superficial retinal plexus in the setting of concurrent BSCR. This is the first report of PAMM in the setting of BSCR; however, recent reports have described associations with nonischemic central retinal vein occlusion (CRVO),⁷ migraine,⁸ hypertensive retinopathy,⁸ prolonged direct orbital pressure,⁸ sickle cell disease,^{8,9} Purtscher retinopathy after motor vehicle collision,⁸ postviral/postvaccination syndrome.⁸ Bilateral PAMM was reported in a pregnant woman with evidence of retinal vasculitis due to Behcets, HLA typing was positive for both A29 and B51 subtypes.¹⁰ Given the vasculitic component of BSCR, it is plausible that this inflammatory setting could lead to transient occlusion of the superficial or deep retinal capillary plexi resulting. Given PAMM is a newly described entity; it is possible that this association is underrecognized. Further study is needed to better determine the incidence in BSCR; but for patients with new paracentral visual scotoma, clinicians should maintain a high level of suspicion given the subtle changes on fundoscopy seen in PAMM and be aware of the findings may present in a variety of clinical scenarios.

Key words: acute macular neuroretinopathy, birdshot chorioretinopathy, inflammation, OCT, paracentral acute middle maculopathy, uveitis, vasculitis.

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