Title: Paracentral Acute Middle Maculopathy in a Patient with Susac Syndrome

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Abstract
The pathogenesis of Paracentral Acute Middle Maculopathy (PAMM) is not fully understood, but is likely secondary to ischemia of the retinal capillaryplexuses. We present a case in a patient with Susac Syndrome.

I. Case History
Patient demographics: 46-year-old Caucasian male
Chief complaint
Central blind spot OS that appears to grow throughout the day x 4 days; also notes peripheral temporal vision OS is distorted and has floaters x 1.5 weeks
Ocular history
H/o multiple branch retinal artery occlusions OD and chronic embolus OD
Peripheral vision loss OS suggestive of BRAO but without embolus or thinning on ancillary testing 1 month prior
Medical history: Susac Syndrome, Hypertension, Premature Ventricular Complexes
Medications: Aggrenox, Metoprolol tartrate, Pantoprazole

II. Pertinent Findings
Clinical
Best corrected acuity: 20/20-1 OU with slight eccentric viewing OS
Preliminary: unremarkable aside from IN field constriction OD
Slit Lamp Exam: unremarkable OU
Tonometry (mmHg): 14 OD, 15 OS
Dilated Fundus Exam
C/D ratio of 0.50 round OD and 0.20 vertical/0.25 horizontal OS
Arterial plaque inferior to ONH OD
Large CWS adjacent to nasal ONH OS and poorly perfused artery/BRAO
Otherwise unremarkable OU
SD-OCT
Temporal/superonasal retinal thinning with intact foveal contour OD
Hyperreflective band at the level of the inner nuclear layer, intact foveal contour OS
Fundus photos and nerve fiber layer OCT done for documentation
Laboratory studies: History of unremarkable work-up
Radiology studies: MRI/MRA ordered; unremarkable, no evidence of encephalopathy

III. Differential Diagnosis
Leading
Branch retinal artery occlusion, Hypertensive retinopathy
Other causes of paracentral vision loss
IV. Diagnosis and Discussion
Paracentral Acute Middle Maculopathy (PAMM) is a condition diagnosable by spectral domain optical coherence tomography (SD-OCT). Originally thought to be part of the spectrum of acute macular neuroretinopathy, it is now recognized as a distinct entity\(^1,2\). Patients typically present complaining of a paracentral scotoma and with band-like hyperreflective intraretinal lesions present on the SD-OCT\(^1\). These lesions occur at the junction of the outer plexiform layer (OPL) and the inner nuclear layer (INL) and result in subsequent thinning of the INL\(^3\). It is thought that this condition is the result of ischemia of the intermediate and deep capillary plexuses and as supporting evidence, has been reported in association with various vascular disorders including vein or artery occlusions, diabetic retinopathy, sickle cell retinopathy, and Purtscher retinopathy\(^1,2\). Therefore, it is not surprising to encounter this finding in a patient with Susac Syndrome, which is typified by a triad of encephalopathy, branch retinal artery occlusions and hearing loss\(^4\). Brandt et al. used various types of retinal imaging to follow a patient through an acute phase of Susac Syndrome and identified four distinct “sections” within the acute lesion; one section labeled as hyperintense swelling of the inner plexiform layer (IPL) and the OPL, followed by loss of the INL, is consistent with PAMM\(^5\). Initially, SD-OCT and near-infrared reflectance (NIR) were found to be the best modes of imaging these lesions\(^3\). Recent studies have focused on using En Face OCT and OCT angiography to further characterize PAMM lesions and have provided additional evidence that the deep capillary plexus is affected in this condition\(^2\).

V. Treatment, management
There is currently no treatment for Paracentral Acute Middle Maculopathy\(^1\). Patients must be managed based on any associated risk factors such as the use of pressor agents or vasoconstrictors and any underlying vasculopathic conditions. Further investigation is needed to determine the underlying etiology and risk factors so that potential treatment can be developed in the future.

Bibliography
\(^1\)E. Rahimy et al. “Paracentral Acute Middle Maculopathy: What we knew then and what we know now.” Retina, the Journal of Retinal and Vitreous Diseases. 2015: 35(10) 1921-1930.

VI. Conclusion
Paracentral acute middle maculopathy is thought to be caused by ischemia of the retinal capillary plexuses, though this theory has yet to be proven. This case report provides further evidence of PAMM in association with other retinal vascular disorders.