Painless Scotoma in a Young Woman

Himanshu Banda, MD; Nicholas E. Engelbrecht, MD

Introduction:

A 27-year-old female presents with a two day history of a painless paracentral scotoma in the right eye. She describes seeing a spot in her vision just below fixation. She has a past medical history of multiple sclerosis (MS) and has been taking Gilenya® (fingolimod) for 4 months. She denies any pain with eye movement or history of ophthalmic manifestations of MS. She denied recent international travel, vaccinations, recent pregnancies, or changes in her medical status. A 12-point review-of-systems was otherwise negative.

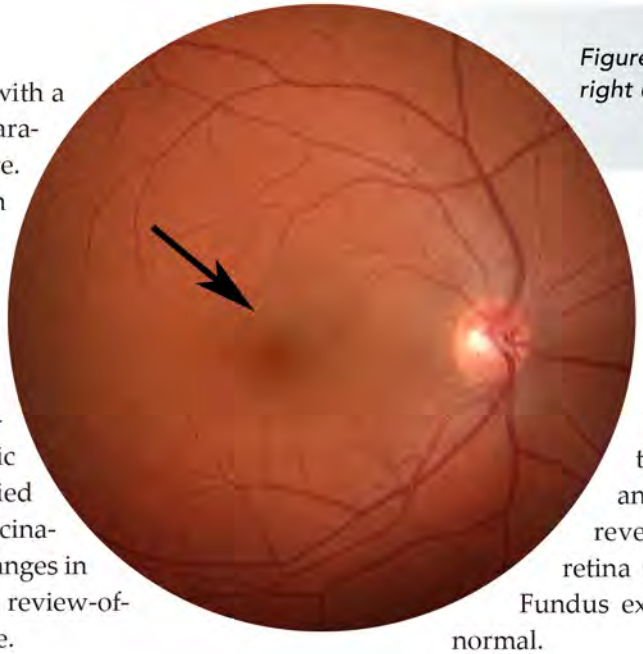

Figure 1: Color fundus photo of the right eye. Subtle hypopigmentation noted above the fovea.

Exam:

Best corrected visual acuity was 20/20 in both eyes. No relative afferent pupillary defect was noted. Confrontation to visual field and extraocular motility was intact. Anterior segment examination was otherwise unremarkable.

DFE revealed subtle hypopigmentary changes superior to the fovea, but was otherwise unremarkable (Figure 1). SD-OCT through the macula of the right eye revealed hyper-reflectivity of the ellipsoid layer and external limiting membrane (Figure 2). Infrared fundus photography revealed a petalloid lesion in the macula with the tip pointed toward the fovea (Figure 3). Multicolor photography also highlighted the petalloid irregularity (Figure 4). Fluorescein angiography (FA) of the right eye revealed an appropriate arm-to-retina time and no perfusion defects. Fundus examination of the left eye was normal.

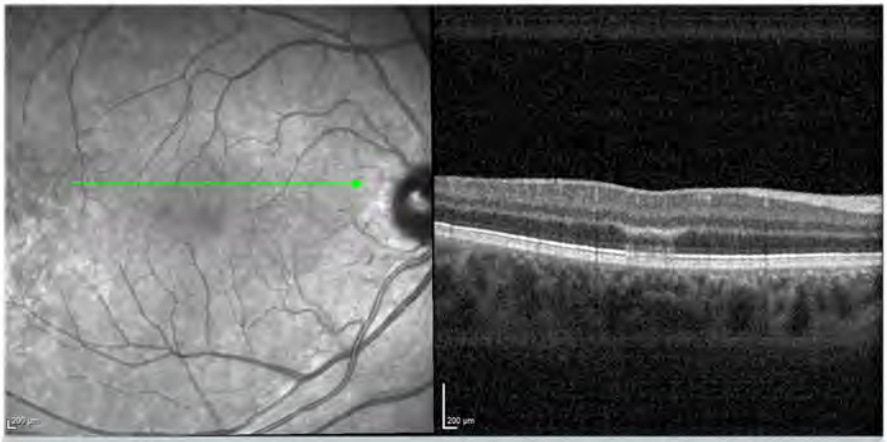

Figure 2: OCT of the right eye. Note hyper-reflectivity of outer retinal layers.

Discussion:

The presumed diagnosis in this case was Acute Macular Neuroretinopathy (AMN). AMN was first described by Bos and Duetman in 1975[1]. It is characterized by the sudden onset of paracentral scotomas in one eye (sometimes both >50%) and typically effects women in their 30s. The pathophysiology of AMN remains elusive, however some hypothesize a vascular etiology effecting the photoreceptor layer[2].

There are a multitude of risk factors associated with

AMN, including fever, oral contraceptives, intravenous contrast, intravenous ephedrine, caffeine, trauma, hypotensive shock. A variety of infectious viral agents have been implicated in AMN, including influenza, respiratory viruses and even chikungunya fever[3]. However, most cases have no known risk factors and are idiopathic. The patient in this case has a history of MS on fingolimod. Though this medication may cause cystoid macular edema, there are no known associations with AMN.

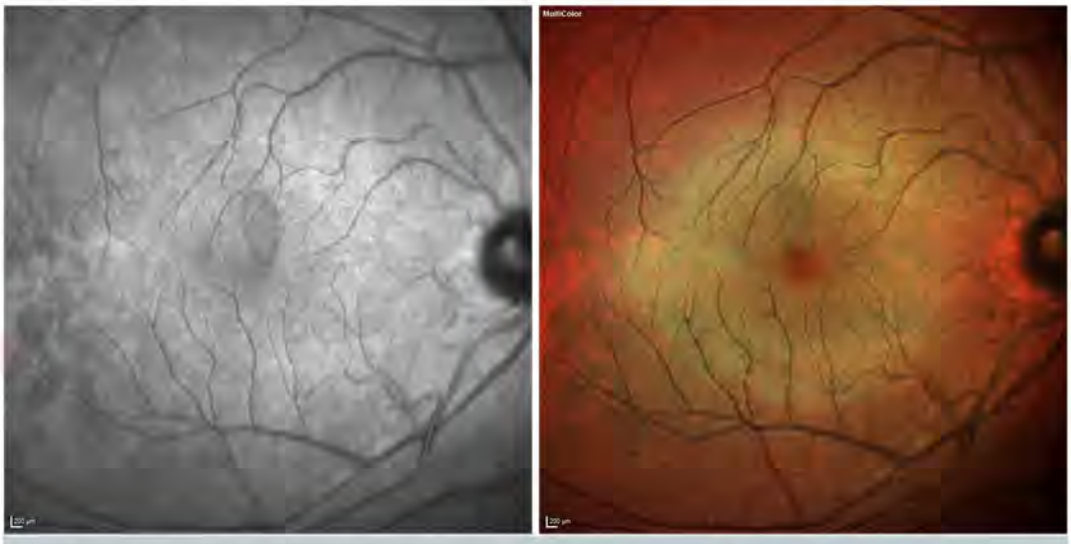

Figure 3 and Figure 4: Infrared and multicolor images of right eye. Petalloid lesions above fovea apparent compared to fundus photography.

Fundus findings in AMN can be subtle on examination and are not clearly defined on color photographs (Figure 1). The best modalities for highlighting retinal architecture of AMN are SD-OCT and infrared (IR) imaging[4]. SD-OCT usually demonstrates hyper-reflectivity of the outer retina, from the outer plexiform layer outward. IR and even multicolor photos clearly defined the area in question (Figure 3 & 4). FA and ICG angiograms are usually normal.

Prognosis for AMN is generally favorable, however patients may experience a permanent scotoma[5]. Our patient continues to have a scotoma on follow-up visits, though fainter compared to her initial presentation.

References:

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2. Sarraf D, Rahimy E, Fawzi AA, et al. Paracentral acute middle maculopathy: a new variant of acute macular neuroretinopathy associated with retinal capillary ischemia. JAMA Ophthalmol 2013;131:1275-87.

3. Pang CE, Navajas EV, Warner SJ, Heisler M, Sarunic MV. Acute Macular Neuroretinopathy Associated With Chikungunya Fever. Ophthalmic Surg Lasers Imaging Retina 2016;47:596-9.

4. Ozmert E, Kurt RA, Batioglu F. Enhanced visualization of acute macular neuroretinopathy by infrared imaging and spectral optical coherence tomography. Indian J Ophthalmol 2013;61:776-7.

5. Bhavsar KV, Lin S, Rahimy E, et al. Acute macular neuroretinopathy: A comprehensive review of the literature. Surv Ophthalmol 2016;61:538-65.

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