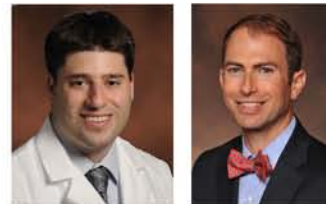




A 39-Year-Old Man with a Rapid Decrease in Vision

Nicholas D. Chinskey, MD; Bradley T. Smith, MD



Introduction:

The patient is a 39 year old man with no significant past medical history who presented with a rapid decrease in vision in the right eye. About one and a half weeks prior to presentation, the patient reported a viral like illness with headaches and myalgia. He was seen by his primary care doctor, who in addition to the viral illness, diagnosed with patient with folliculitis of his arm. He was treated with oral doxycycline for a five day course. About two to three days prior to presentation, the patient reported flashes and decreased vision in the right eye. At initial presentation, best corrected visual acuity was count fingers at one foot in the right eye, while 20/20 in the left. Pupils, confrontation visual fields and slit lamp examination were all normal. The fundus exam showed pallor of the macula with at least

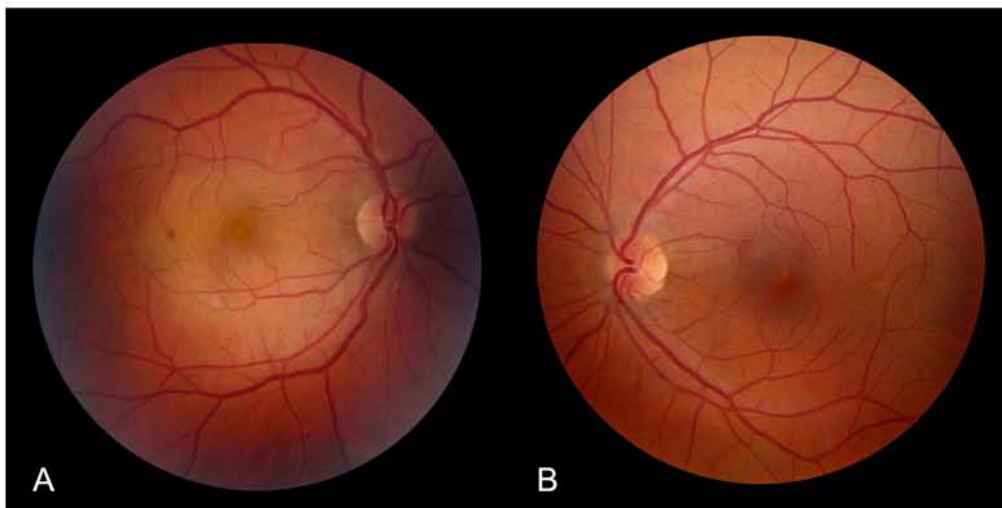


Figure 1: Fundus photography of the right eye (a) at the time of presentation shows pallor of the central macula with a single intra-retinal hemorrhage temporal to the fovea. The left eye (b) demonstrates a single pigment epithelial detachment (PED) temporal to the fovea.

one area of intra-retinal hemorrhage and some pigment mottling centrally (Figure 1a). The left eye showed a mild degree of pigment mottling centrally with a small pigment epithelial detachment (PED) temporal to the fovea (Figure 1b). Fluorescein angiography demonstrated late staining of nearly the entire right macula,



Figure 2: Fluorescein angiography of the right eye demonstrates early scattered hyper and hypo- florescence (a) with late staining (b). The left eye again shows evidence of a PED (c).

with a small area of pooling in the left eye at the PED (Figures 2a-2c). Optical coherence tomography (OCT) showed spotty irregularity of the ellipsoid region in the right eye without subretinal fluid while again demonstrating the PED in the left (Figure 3a-3b).

Over two weeks, the patient's vision improved to 20/400 in the right eye and then back to 20/20 at three months out.

At his last visit, there was significant pigment mottling of the central macula with resolution of the retinal whitening (Figure 4a-b). OCT showed an irregular outer retina and thickened retinal pigment epithelium, but return of the ellipsoid band (Figure 4c). The PED remained in the left eye (Figure 4d-e). The patient was tested for the Coxsackie virus and did have positive titers for Coxsackie B, type 5.

Discussion:

Unilateral acute idiopathic maculopathy (UAIM) is a rare condition that occurs in young adults leading to acute, sudden loss of central vision (often 20/200 or worse) that was first described by Yanuzzi et al. in 1991¹. It often occurs days after a viral like illness, with many patients with positive Coxsackie virus titers. Typically, patients present with a neurosensory retinal detachment with thickening of the retinal pigment epithelium (RPE) and occasional intraretinal hemorrhages and cells in the posterior vitreous¹⁻². Fluorescein angiography shows irregular, early hyper and

hypofluorescence, with late diffuse staining. Most patients have return of normal or near normal vision over the course of several weeks¹⁻².

Very few bilateral cases have been described. In these few cases, patients developed a classic UAIM lesion in one eye, with a similar lesion either inside, or just outside the macula, in the other²⁻³. Here we describe a patient whose imaging and clinical course match that of UAIM, while also developing an asymptomatic pigment epithelial detachment (PED) in the fellow eye.

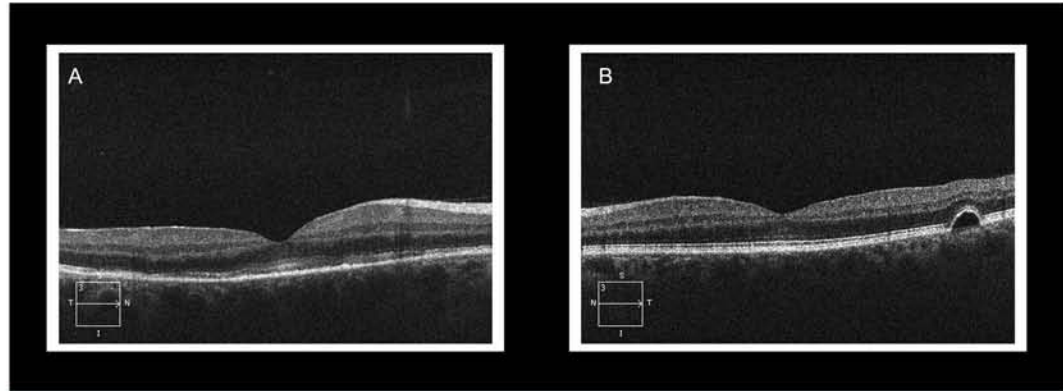


Figure 3: Optical coherence tomography (OCT) of the right eye (a) demonstrates an irregular outer retina with focal loss of the ellipsoid band. The left eye (b) shows a normal image with a temporal PED.

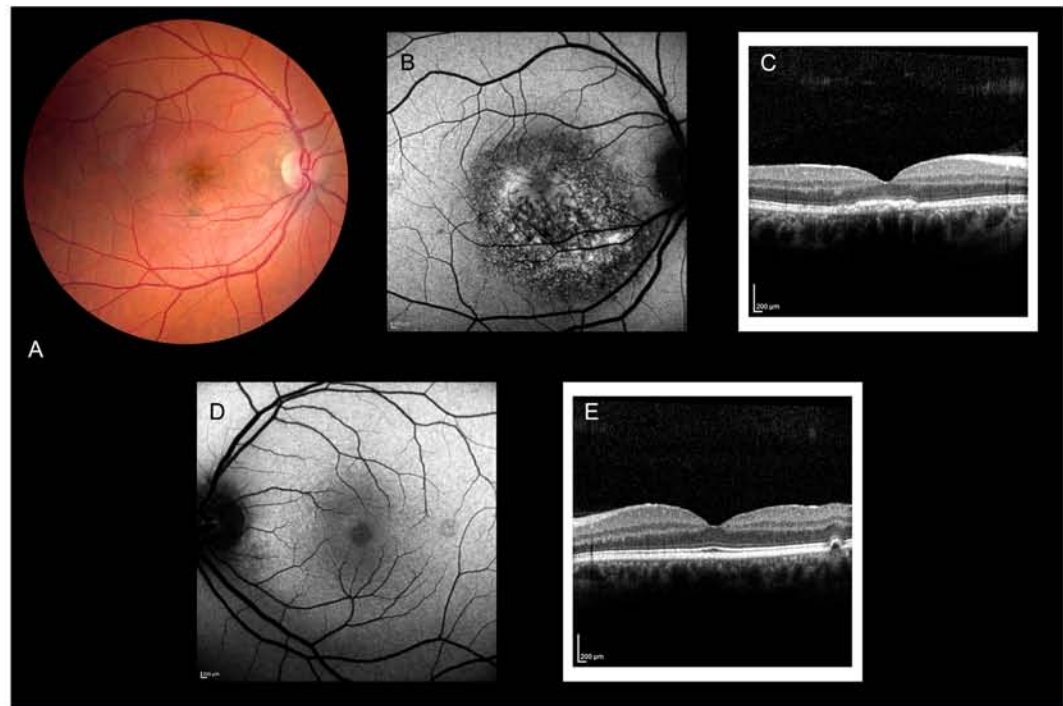


Figure 4: At three months follow-up, the fundus photo of the right eye show significant pigment mottling of the macula with resolution of retinal whitening (a). Autofluorescence demonstrates grossly irregular retinal pigment epithelium of the right eye (b). On OCT, there is return of the ellipsoid band with focal thickening of the retinal pigment epithelium (c). The left eye autofluorescence (d) is irregular at the site of the PED, which persists on OCT (e).

This case also demonstrates some of the best OCT imaging of early and late lesions in UAIM. Previous reports predated OCT or relied on time domain images. There have only been a few reports in the literature examining the findings with spectral domain quality^{4,5}. In this case, where modern day spectral domain OCT was available at all times points, there is patchy loss of the ellipsoid region with focal irregularity of the RPE in the early stage of the disease. There was no evidence of sub-retinal fluid in our case, which has been seen in other reported cases. Following resolution of symptoms, there is clear, focal thickening of the RPE with restoration of the ellipsoid band.

The cause of UAIM remains unknown. Although most authors agree that the disease is likely inflammatory in nature, the site of action (RPE vs choroid) is still debated.

References:

- 1) Yannuzzi LA, Jampol LM, Rabb MF, et al. Unilateral acute idiopathic maculopathy. *Arch Ophthalmol*. 1991 Oct;109(10):1411-6.
- 2) Freund KB, Yannuzzi LA, Barile GR, et al. The expanding clinical spectrum of unilateral acute idiopathic maculopathy. *Arch Ophthalmol*. 1996 May;114(5):555-9.
- 3) Ghazi NG, Daccache A, Conway BP. Acute idiopathic maculopathy: report of a bilateral case manifesting a macular hole. *Ophthalmology*. 2007 May;114(5):e1-6.
- 4) Xu H, Lin P. Unilateral recurrent acute idiopathic maculopathy. *Graefes Arch Clin Exp Ophthalmol*. 2011 Jun;249(6):941-4.
- 5) Milani P, Cacioppo V, Raimondi G, Scialdone A. Spectral domain OCT and autofluorescence imaging of unilateral acute idiopathic maculopathy. *Eur J Ophthalmol*. 2012 May-Jun;22(3):499-502.



Introduction:

The Retina Institute welcomes Yicheng Chen, MD and Abdallah Jeroudi, MD to our Fellowship Program.

