A 23-Year-Old Female Reports Seeing Objects "Shaped Like Jellyfish"

Gautam Vangipuram, MD; Sabin Dang, MD

Introduction:

A 23-year-old female presents to our clinic reporting issues with her vision for the last month. She reports seeing objects "shaped like jellyfish" in the center of her visual field. In addition, she reports a headache of variable intensity for the last month. All these symptoms were proceeded by a recent viral respiratory illness lasting for two weeks. Her past medical and ocular history is unremarkable.

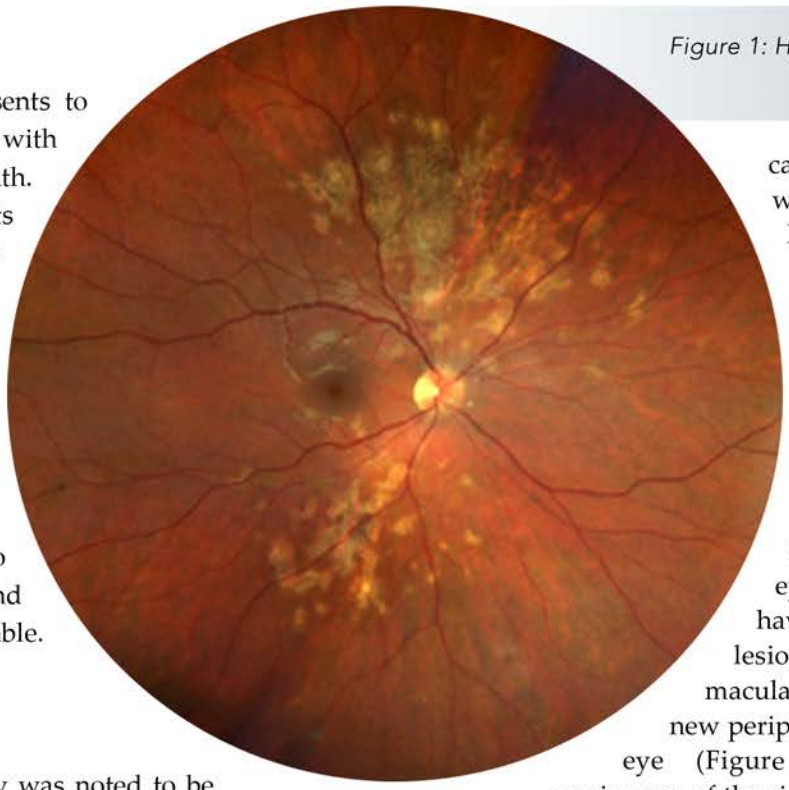

Figure 1: Hypopigmented lesions in the right eye.

Exam:

On exam, her visual acuity was noted to be 20/20 OU with normal intraocular pressures and no afferent pupillary defect. Her anterior segment exam was unremarkable and her vitreous was notable for rare cell in both eyes. Her posterior segment exam was notable for mild disc edema in the right eye and multiple hypopigmented lesions scattered through the posterior pole, sparing the macula, with variable stages of pigmentation (Figure 1). Her right eye was affected more than her left. Based on her presentation and exam findings with associated headache, a preliminary diagnosis of acute posterior multifocal placoid pigment epitheliopathy (APMPPE) was made. She was immediately sent from our clinic to the emergency room for head imaging to rule out concomitant cerebral vasculitis. Additionally, a laboratory workup was started to rule out common causes of ocular inflammation including tuberculosis, syphilis, and sarcoidosis which all came back normal. She was started on high dose IV steroids with neurology consultation.

Following a tapering of her steroids to oral 10mg prednisone, she was seen two weeks later with slight worsening of her vision in her right eye. She was found to have new posterior pole lesions now affecting the macula in her right eye and new peripheral lesions in her left eye (Figure 2). A fluorescein angiogram of the right eye was notable for early blockage with late staining of her chorioretinal lesions (Figure 3). OCT of the macula through the lesions was notable for outer retinal irregularity with no intraretinal or subretinal fluid (Figure 4). Fortunately, her visual acuity was preserved at 20/20 OU. Her oral steroids were increased back to 60mg. At her subsequent visits, she was noted to have stable exam findings but became increasingly intolerant of the side effects associated with her steroid course. Her steroids were reduced to 40 mg and on subsequent follow up a week later, she was noted to have new lesions in her both eyes (Figure 5). Her vision remained 20/20 OU and she was subsequently increased on her oral steroids and counseled to follow up with rheumatology to initiation of steroid sparing immunotherapy. Given her relapsing course of new chorioretinal inflammation shortly after cessation of steroids, she was given a diagnosis of ampiginous or relentless placoid choroiditis.

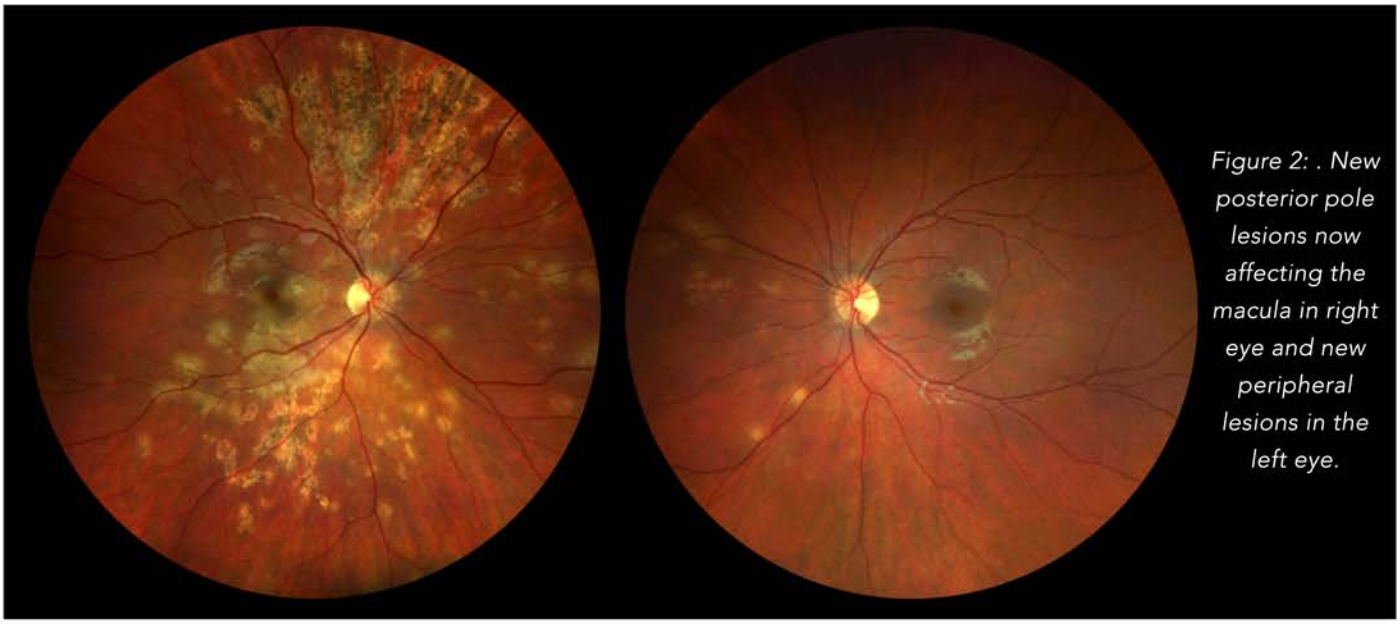

Figure 2: . New posterior pole lesions now affecting the macula in right eye and new peripheral lesions in the left eye.

Discussion:

APMPEE is a rare (0.15 cases per 100000 persons) bilateral ocular inflammatory syndrome affecting the choroid and retinal pigment epithelium that can eventually lead to outer retinal atrophy and vision loss.[1-2] It generally occurs in young healthy individuals and is often times proceeding by a viral prodrome. Generally, this disease is monophasic with spontaneous resolution in 1-2 months. However, inflammation associated with APMPPE can also be present in the central nervous system as cerebral vasculitis. Thus, any patient with suspected APMPPE and neurological symptoms should be evaluated with appropriate cerebral vascular imaging to rule out intracranial inflammation. When lesions recur or new lesions develop in spite of immunosuppressive therapy, as with our patient, a diagnosis of ampiginous choroiditis (relentless placoid choroiditis) should be suspected.[3-6] This name derives as a combination of another more serious ocular syndrome: serpiginous choroiditis, which presents as bilateral asynchronous chorioretinal inflammation emanating from the optic disc often leading to central vision loss sometimes despite immunosuppressive therapy.[7] Patients with a suspected diagnosis of ampiginous choroiditis should be thoroughly worked up for infectious

Figure 3: Fluorescein angiogram of the right eye was notable for early blockage with late staining of the chorio-retinal lesions.

etiologies such as tuberculosis and syphilis and consultation with a rheumatologist is recommended for initiation of long-term steroid sparing immunotherapy. The visual prognosis in this condition is guarded and variable depending on the degree and location of chorioretinal lesions and patients must be followed closely with multimodal imaging for the development of recurrent inflammation.

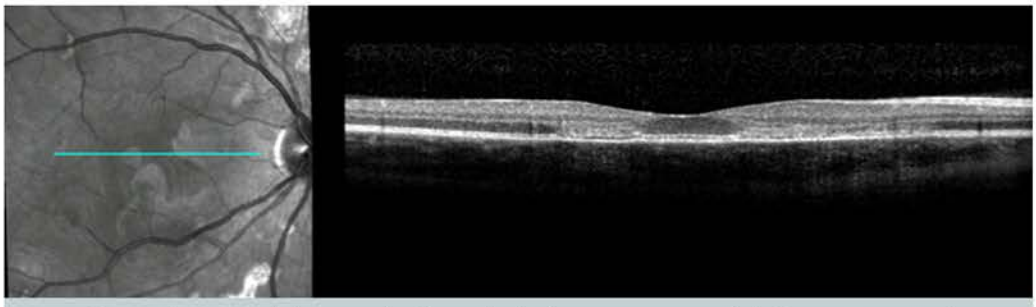

Figure 4: OCT of the macula through the lesions was notable for outer retinal irregularity with no intraretinal or subretinal fluid

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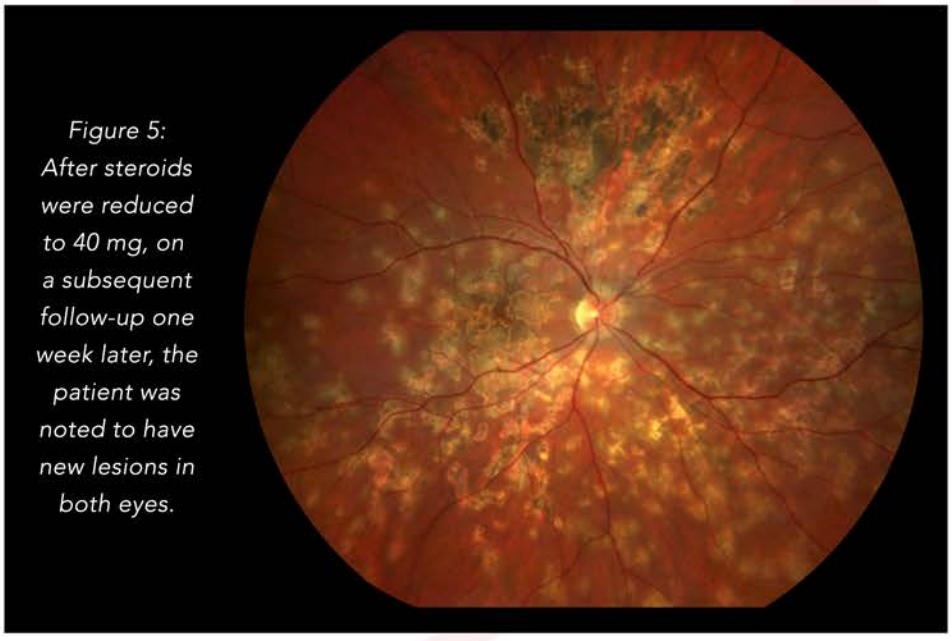

Figure 5: After steroids were reduced to 40 mg, on a subsequent follow-up one week later, the patient was noted to have new lesions in both eyes.

THE RETINA INSTITUTE

2201 Brentwood Blvd.
St. Louis, MO 63144

(314) 367-1181
tri-stl.com

Kevin J. Blinder, MD
Sabin Dang, MD
Alia K. Durrani, MD
Nicholas E. Engelbrecht, MD
M. Gilbert Grand, MD

Daniel P. Joseph, MD, PhD
Thomas K. Krummenacher, MD
Gaurav K. Shah, MD
Bradley T. Smith, MD