A 48-Year Old Female with Bilateral Vision Loss

Wen-Shi Shieh, MD; Matthew A. Thomas, MD

Introduction:

A 48-year old female presented with sudden onset of blurred vision in both eyes. Visual acuity (VA) was 20/100 in the right eye and 20/60 in the left eye. Fundus examination of both eyes revealed many cotton wool spots distributed in peripapillary fashion along with intraretinal hemorrhages and central edema (Figure 1). Fluorescein angiography demonstrated early blockage from the cotton wool spots, late capillary leakage in the perifoveal region, but otherwise normal perfusion and no evidence of optic nerve edema or vasculitis bilaterally (Figure 2). Optical coherence tomography (OCT) of the right eye showed superficial and intraretinal edema as well as subretinal fluid centrally with outer retinal disruption (Figure 3A). OCT of the left eye had similar findings.

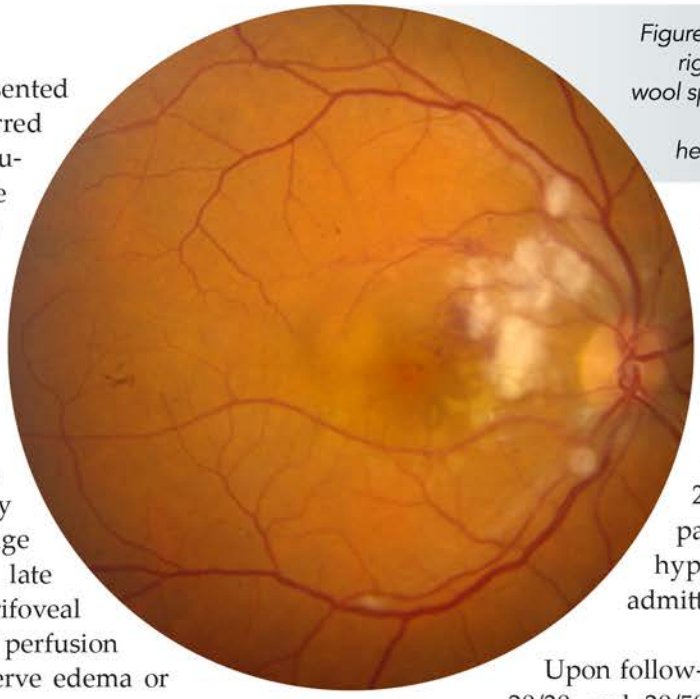
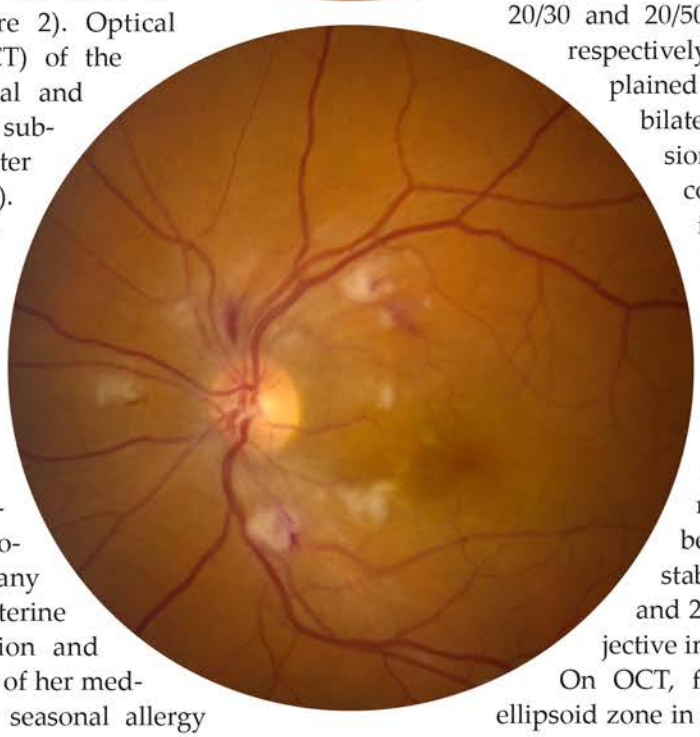


Figure 1: Fundus photographs of the right and left eye showing cotton wool spots distributed in peripapillary fashion along with intraretinal hemorrhages and central edema.

Clinical Course:

The patient was noted to have a Purtscher-like retinopathy. Her past medical history included hypertension, mild hemochromatosis (that did not require any phlebotomy), a history of uterine bleeding requiring a dilation and curettage procedure. Review of her medications was significant for seasonal allergy medications and recent discontinuation of oral contraceptives. On review of systems, the patient admitted to having worsening headaches for a 1-week duration. Blood pressure measurements were obtained in the office and was found to be elevated at 205/120. Consequently, the patient was sent to the ER for hypertensive emergency and admitted to the ICU for treatment.



Upon follow-up one month later, VA was 20/30 and 20/50 in the right and left eye, respectively, although the patient complained of scotomas in her vision bilaterally. The patient's hypertension was reported to be well-controlled on hydrochlorothiazide and lisinopril. OCT imaging showed resolution of macular edema and subretinal fluid, but paracentral areas of disruption in the ellipsoid zone and overlying thinning in both eyes (Figure 3B). Three months later, the patients' best corrected VA remained stable at 20/30 in the right eye and 20/50 in the left eye with subjective improvement of the scotomas. On OCT, further regeneration of the ellipsoid zone in previous areas of disruption are seen bilaterally but parafoveal areas of

retinal thinning persist (Figure 3C). Systemically, the patient's hypertension remains well-controlled on a single agent of hypertensive medication.

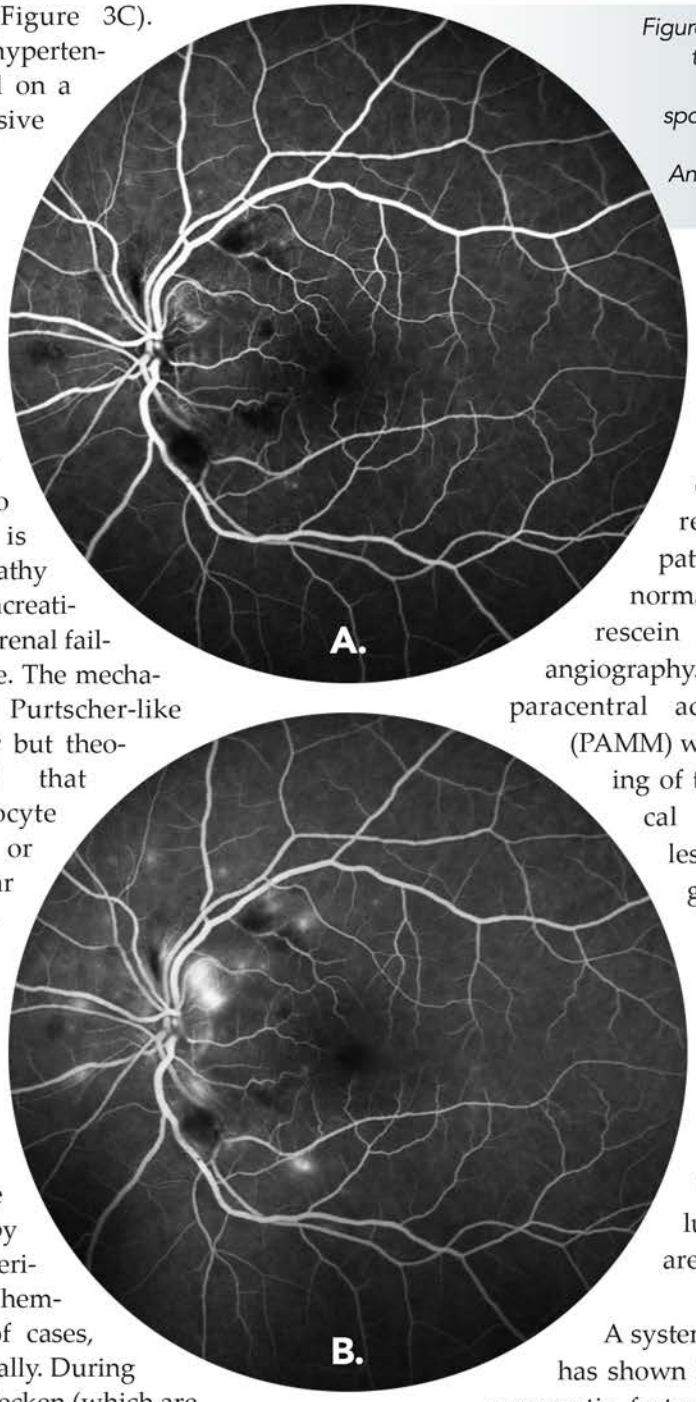


Figure 2: Fluorescein angiography of the left eye demonstrating early blockage from the cotton wool spots (A) and late capillary leakage in the perifoveal region (B). Angiographic imaging of the right eye showed similar findings.

Discussion:

Purtscher's retinopathy, first described in 1910 by Otmar Purtscher, is an occlusive microvasculopathy typically associated with trauma.[1] A similar clinical presentation due to non-traumatic etiologies is called Purtscher-like retinopathy and is often seen in acute pancreatitis, amniotic fluid embolism, renal failure, and autoimmune disease. The mechanism of Purtscher and Purtscher-like retinopathy remains unclear but theories have hypothesized that embolization due to leukocyte aggregates, fat, fibrin, or platelets leads to arteriolar occlusion and ischemia. Other proposed mechanisms include activation of the complement cascade and thus formation of C5a-induced leukocyte, platelet, and fibrin aggregates.[2]

Clinically, Purtscher-like retinopathy is characterized by cotton-wool spots in the posterior pole with few intraretinal hemorrhages. In the majority of cases, these findings present bilaterally. During the acute phases, Purtscher flecken (which are pathognomonic of the disease) can be observed, which are polygonal areas of inner retinal whitening between arterioles and venules that also exhibit a clear zone between the affected retina and an adjacent arteriole. Optic disc and retinal edema can also be present.[2,3]

Fluorescein angiography can demonstrate a spectrum of findings, including capillary nonperfusion, retinal and/or disc edema, perivascular staining, and late leakage, which may suggest the severity of the disease.[4] Interestingly, our patient's angiography demonstrated good perfusion and no evidence of nerve edema or vascular abnormalities. In a recent case report, Shahlaee et al. described a young woman with sudden-onset bilateral vision loss after a recent viral illness and a diagnosis of Purtscher-like retinopathy. Similar to our patient, the reported case had normal perfusion on baseline fluorescein angiography and OCT angiography. However, OCT evidence of paracentral acute middle maculopathy (PAMM) was observed on en face imaging of the middle retina as multifocal globular hyperreflective lesions, which the authors suggest are representative of distal ischemic events in smaller terminal retinal arterioles, precapillaries, and capillaries. The patient achieved full visual recovery over a 12-month follow up period and serial en fact OCT imaging showed resolution of these hyperreflective areas.[5]

A systematic review by Miguel et al. has shown that etiology can be a visual prognostic factor for cases of Purtscher-like retinopathy, with pancreatitis and trauma being associated with higher probability of vision improvement.[2] In a separate study, a significant visual recovery of at least 2 and 4 Snellen lines was seen in 50% and 23% of the eyes studied without any treatment.[6] While there are isolated case reports suggesting that high-dose, intravenous steroids may be beneficial, the treatment of Purtscher-like retinopathy largely consists of observation. However, a thorough history and systemic work-up is often warranted and management should be directed toward addressing the underlying cause.

References:

1. Agrawal A, McKibbin MA. Purtscher's and Purtscher-like retinopathies: a review. Surv of Ophthalmol. 2006; 51(2):129–136.

2. Miguel AIM, Henriques F, Azevedo LFR et al. Systematic review of Purtscher's and Purtscher-like retinopathies. Eye (Lond) 2013;27:1–13.

3. Proença Pina J, Ssi-Yan-Kai K, de Monchy I, Charpentier B, Offret H, Labetoulle M. Purtscher-like retinopathy: case report and review of the literature. J Fr Ophtalmol. 2008 Jun;31(6 Pt 1):609-13.

4. Kelley JS. Purtscher's retinopathy related to chest compression by safety belts. Fluorescein angiographic findings. Am J Ophthalmol. 1972 Aug; 74(2):278-83.

5. Shahlaee A, Sridhar J, Rahimy E, Shieh WS, Ho AC. Paracentral Acute Middle Maculopathy Associated with Postviral Purtscher-like Retinopathy. Retin Cases Brief Rep. 2017 Jan 12. [Epub ahead of print]

6. Agrawal A, McKibbin MA. Purtscher's retinopathy: epidemiology, clinical features and outcome. Br J Ophthalmol. 2007 Nov; 91(11): 1456–1459.

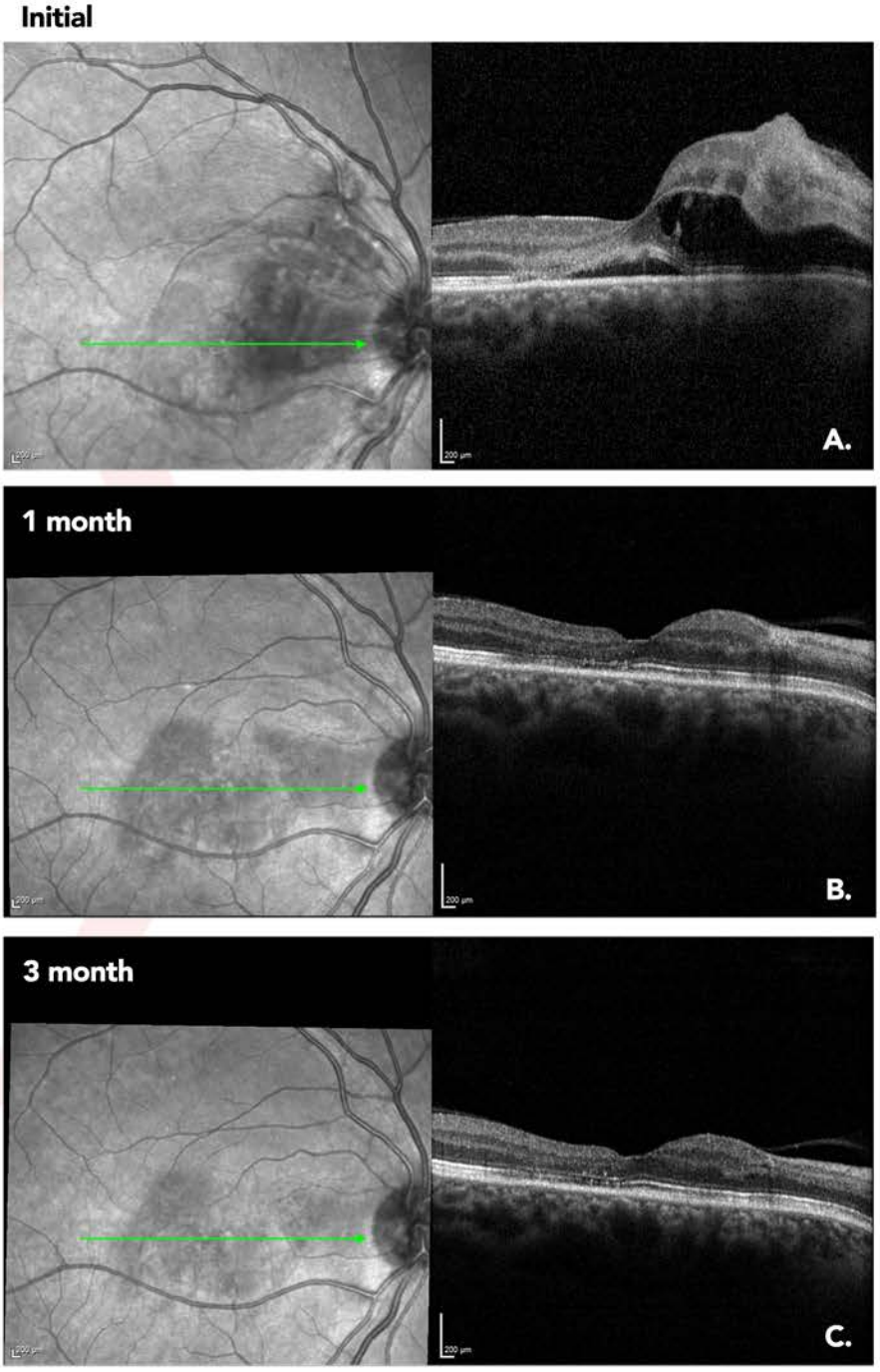


Figure 3. (A) Superficial and intraretinal edema with subretinal fluid centrally and outer retinal disruption on initial OCT of the right eye. (B) One-month follow up: Resolution of macular edema and subretinal fluid; paracentral areas of disruption in the ellipsoid zone and overlying retinal thinning. (C) Regeneration of the ellipsoid zone in previous areas of disruption with persistent foci of retinal atrophy at three months follow-up.

THE RETINA INSTITUTE
(314) 367-1181
tri-stl.com

Kevin J. Blinder, MD
Nicholas E. Engelbrecht, MD
M. Gilbert Grand, MD
Daniel P. Joseph, MD, PhD
Thomas K. Krummenacher, MD
Richard J. Rothman, MD
Gaurav K. Shah, MD
Bradley T. Smith, MD
Matthew A. Thomas, MD