A 27-Year-Old Female with a Blind Spot in Both Eyes

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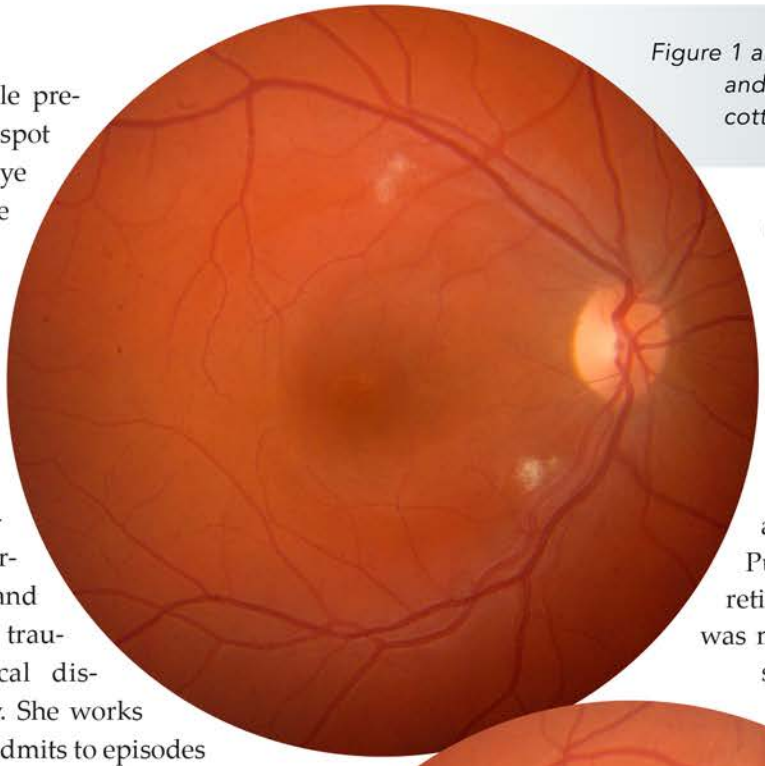
Introduction:

A 27-year-old female presented with a blind spot in both eyes, right eye more than left. She had an episode of pancreatitis one month prior to presentation and felt that her visual symptoms occurred around the time of her diagnosis. She is otherwise healthy and denies any recent trauma, rheumatological diseases, or pregnancy. She works as a bartender and admits to episodes of binge drinking.



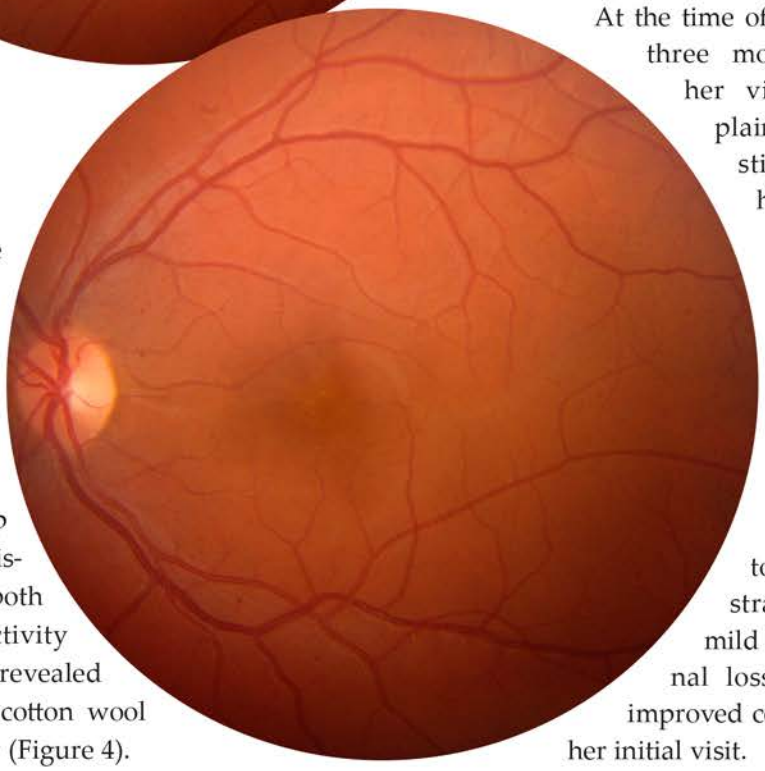


Figure 1 and 2: Fundus photo of the right and left eye, respectively. Note the cotton wool spots in the right eye.

Exam:

Her visual acuity measured 20/50 in the right eye and 20/30 in the left. Intraocular pressure and anterior segment examination was unremarkable. Posterior segment examination revealed central RPE changes in both eyes with multiple cotton wool spots in the right eye (Figure 1 and 2). Her discs were sharp and there was no APD. OCT revealed disruption of the outer retinal layers in both eyes without edema or hyperreflectivity (Figure 3). A fluorescein angiogram revealed areas of blockage correlating with the cotton wool spots but otherwise showed normal flow (Figure 4).

Clinical Course:

Given her recent diagnosis of pancreatitis and the posterior pole findings the patient was diagnosed with Purtscher-like retinopathy. As her visual acuity was only mildly impaired and treatment options for Purtscher and Purtscher-like retinopathy are limited the decision was made to observe the patient and see her back in several months. At the time of her return three months later her visual complaints, while still present, had improved and the cotton wool spots had resolved. Her OCT continued to demonstrate some mild outer retinal loss that was improved compared to her initial visit.

Discussion:

Originally described in 1910 in a patient who sustained severe head trauma, Purtscher retinopathy results in loss of vision and characteristic retinal whitening along with intraretinal hemorrhages in the posterior pole. Although originally described following blunt trauma, Purtscher-like retinopathy has been seen in cases of pancreatitis, fat embolism, renal failure, and connective tissue disorders.[2]

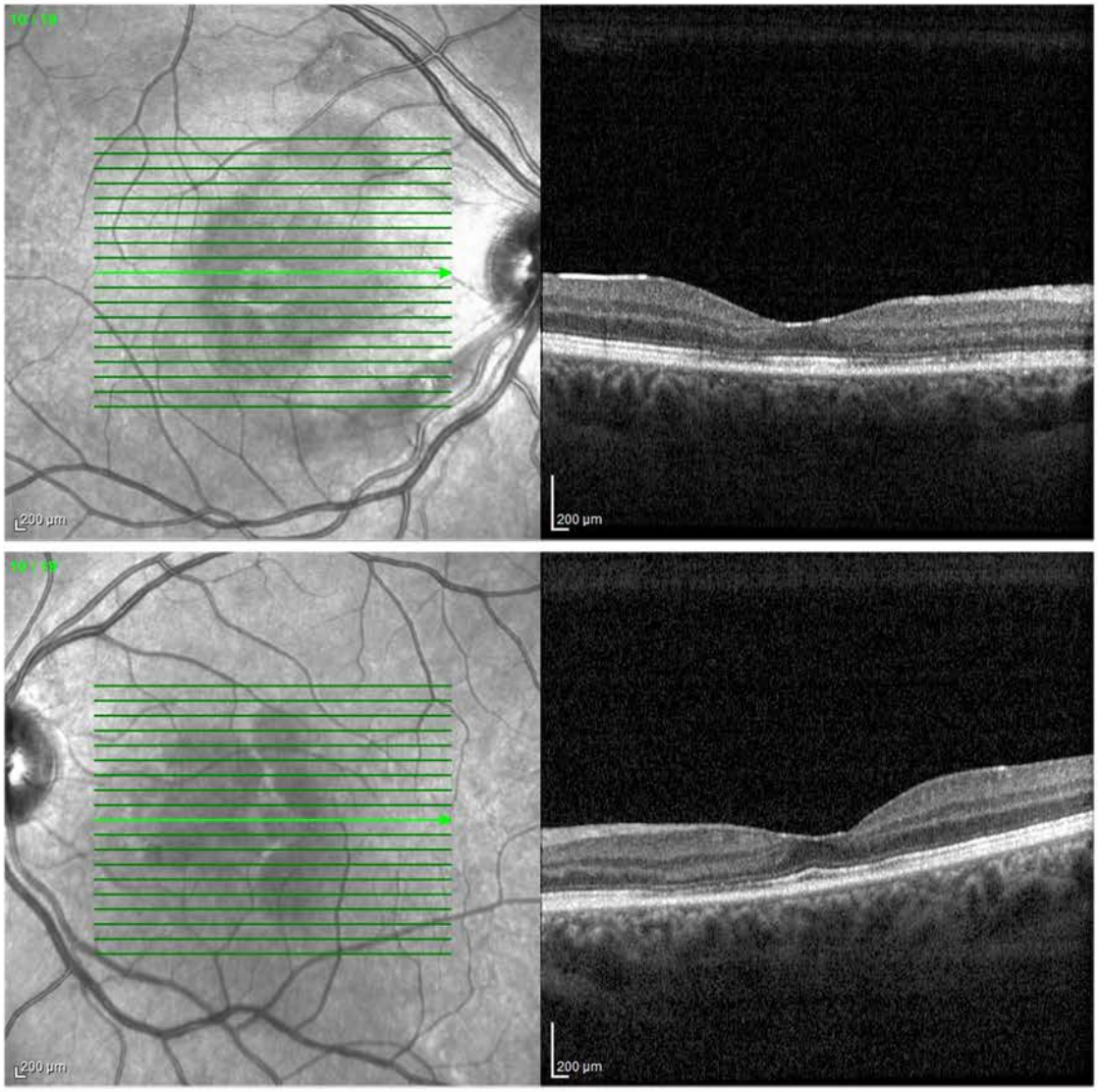


Figure 3: OCT of the right and left eye. There is outer retinal disruption in both eyes.

The etiology of the retinal findings in Purtcher and Purtscher-like retinopathy may be related to leukocyte aggregation by activated complement factor 5, resulting in occlusion of retinal capillaries. Histopathology has demonstrated fibrin positive occluding material in retinal and choroidal vessels, areas of retinal edema and loss of photoreceptors with an abrupt transition to normal retina.[1] It is possible that embolization may be caused by other etiologies such as fat, air, or platelets. Regardless of the etiology of the occlusion, the histopathology suggests that micro infarctions involving retinal capillaries supplying the nerve fiber layer and choriocappilaris are the cause of the damage. Similar findings can occur in conditions as varied as blunt trauma, acute pancreatitis, and collagen vascular diseases.

Patients typically present with unilateral or bilateral loss of vision ranging from mild impairment to severe visual dysfunction. Central, paracentral, or arcuate scotomas have all been described. The onset of visual symptoms typically occurs within 24-48 hours following the acute injury or illness. Fundoscopic exam reveals multiple, discreet areas of retinal whitening along with cotton wool spots and intraretinal hemorrhages.[4,6] Hemorrhage, when they occur, are usually minimal and flame shaped.[1,7] Disc edema may be present as well. The discrete areas of retinal whitening have been described as polygonal in shape and located between arterioles and venules. The size of these lesions are variable and can be between a quarter to several disc areas.[1] A pseudo cherry red spot may be present if the whitening occurs around the fovea. Swelling of the nerve fiber layer corresponding to cotton wool spots,

intraretinal and subretinal fluid, and inner retinal hyperreflectivity consistent with a paracentral acute middle maculopathy (PAMM) lesion may be seen on OCT.[3,5] Fluorescein angiography may show masking of the choroidal fluorescence by the retinal whitening or hemorrhage, and nonperfusion of the retinal capillary bed may be seen.[1] Late leakage from ischemic retinal vessels may be seen as well.

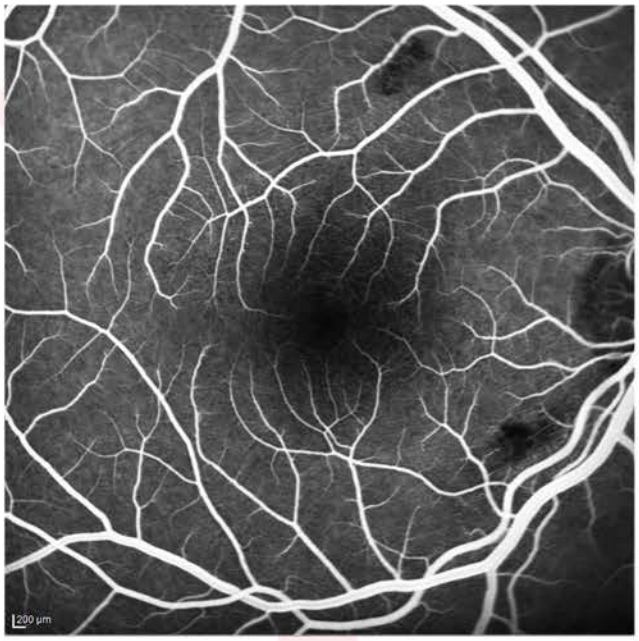


Figure 4: FA of the right eye. Note the blocking from the cotton wool spots without evidence of leakage or vascular obstruction.

Visual outcome is dependent on the degree of damage and amount of capillary nonperfusion of the macula and nerve. There is no standard treatment although there have been isolated case reports of successful treatment using high dose intravenous steroids and hyperbaric oxygen. Treatment of the underlying systemic condition is of course recommended. In most cases without treatment the retinal lesions resolve spontaneously leaving behind some mottling of the pigment epithelium, disc pallor, or sheathing of the retinal vessels.

References:

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