Purtscher Retinopathy Following Subarachnoid Hemorrhage and Femur Fracture

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Abstract

Purtscher retinopathy is a rare microvasculopathy that occurs following trauma. Clinical presentation consists of cotton wool spots, retinal hemorrhages, and Purtscher flecken within the posterior pole. While similar findings have been associated with a heterogenous group of systemic disorders, these non-traumatic etiologies are classified as Purtscher-like retinopathy. Although the pathogenesis remains to be fully understood, clinical and experimental evidence implicate an occlusive vasculopathy of the precapillary arterioles. Clinical presentation usually resolves within months following trauma; however, visual prognosis is variable. There are presently no evidence-based treatments available for Purtscher retinopathy.

KEY WORDS

Purtscher retinopathy, traumatic retinopathy, occlusive microvasculopathy, long bone fracture

INTRODUCTION

Purtscher retinopathy was first described in 1910 by Ottmar Purtscher after he observed bilateral patches of retinal whitening and retinal hemorrhages in patients who sustained severe cranial trauma.[1-5] The condition has since been classically defined as a sequelae of head trauma, chest compression injury, or long bone fracture in the absence of direct ocular trauma.[1-4] In the setting of trauma, a diagnosis is made clinically based on the presence of peripapillary cotton wool spots, retinal hemorrhages, and pathognomonic Purtscher flecken.[6] Similar funduscopic findings have been observed with various systemic etiologies including renal failure, pancreatitis, and autoimmune disease.[1, 3, 4] When findings present in the absence of trauma, a diagnosis of Purtscher-like retinopathy is given.[1, 5]

CASE DESCRIPTION

A 58-year-old African American male presented for a comprehensive eye examination with complaints of reduced distance vision OU without correction after breaking his spectacles a month prior. He denied a history of ocular surgeries or ocular trauma and had no family history of eye diseases. His last ocular examination (a year prior) was remarkable for a pterygium in the right eye and refractive error OU. His medical history was notable for hypertension, hypertriglyceridemia, tobacco use (½ PPD for 20 years), depression, and prostate cancer (status post prostatectomy and external beam radiotherapy in 2010). He was not actively taking any ocular or systemic medications and was controlling vascular risk factors through diet and exercise.

Entering visual acuities without correction were 20/70 with the right eye and 20/70 with the left eye. He was best corrected to 20/30+1 with the right eye and 20/20-2 with the left eye, stable to the year prior. The right eye was able to pinhole to 20/20. Pupils were equal, round, and reactive to light without an afferent pupillary defect OD and OS. Confrontation visual fields were full to finger count with each eye. Extraocular muscles demonstrated full range of motion OU. Anterior segment evaluation was remarkable for complexion-associated melanosis of the bulbar conjunctiva OU and a nasal pterygium encroaching the visual axis OD. Both eyes had trace nuclear

sclerosis and vitreous syneresis. Intraocular pressures measured via Goldmann applanation were 14mmHg OD and 12mmHg OS. As demonstrated in Figures 1 and 2, dilated fundus examination revealed cotton-wool spots OU that were primarily localized peripapillary. A flame hemorrhage was present in the superior temporal arcade of the right eye and inferior to the optic nerve head in the left eye. Both optic nerves were pink with distinct margins. The retinal arteries demonstrated mild attenuation OU. Drusen was present throughout the arcades in both eyes. The macula of each eye was clear and without macular edema. The retinal periphery was intact without breaks OU.

Figure 1: *Right posterior pole on initial presentation.*

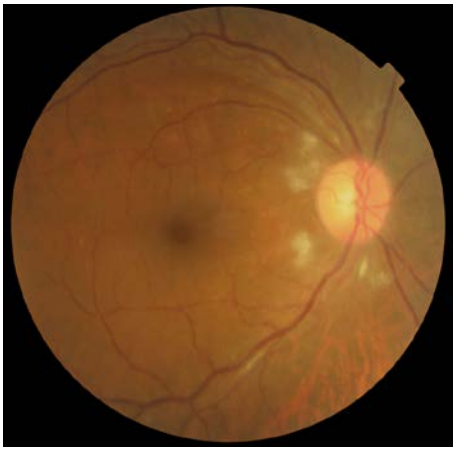
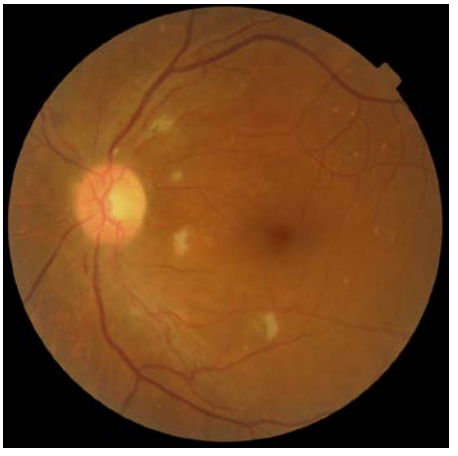


Figure 2: *Left posterior pole on initial presentation.*



Given these findings, in-office blood pressure was measured to be 135/87. Upon further questioning, the patient admitted that his glasses had broken a month prior due to being struck by an automobile while riding his bicycle. Following the accident, he was admitted to the hospital where he was found to have a small subarachnoid hemorrhage along the left anterior frontal lobe and a right femur fracture. He was taken to the operating room on the same day where he underwent uncomplicated cephallomedullary nail for the fracture.

Considering the patient's history, a diagnosis of bilateral Purtscher retinopathy was made. He returned to the clinic three months following the initial visit; visual acuities remained stable and dilated fundus examination revealed resolution of the cotton-wool spots and retinal hemorrhages previously noted (Figures 3 and 4).

Figure 3: *Right posterior pole 3 months after initial presentation.*

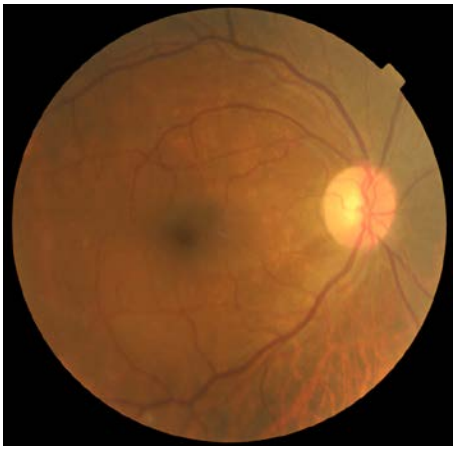
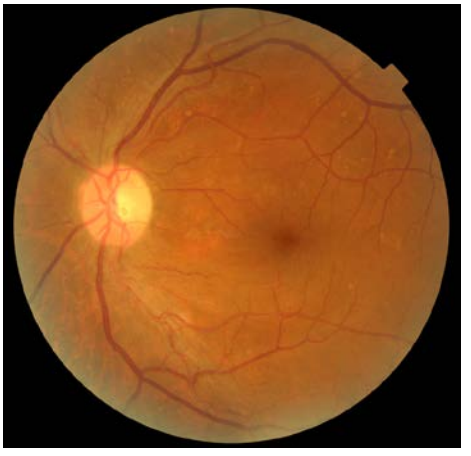


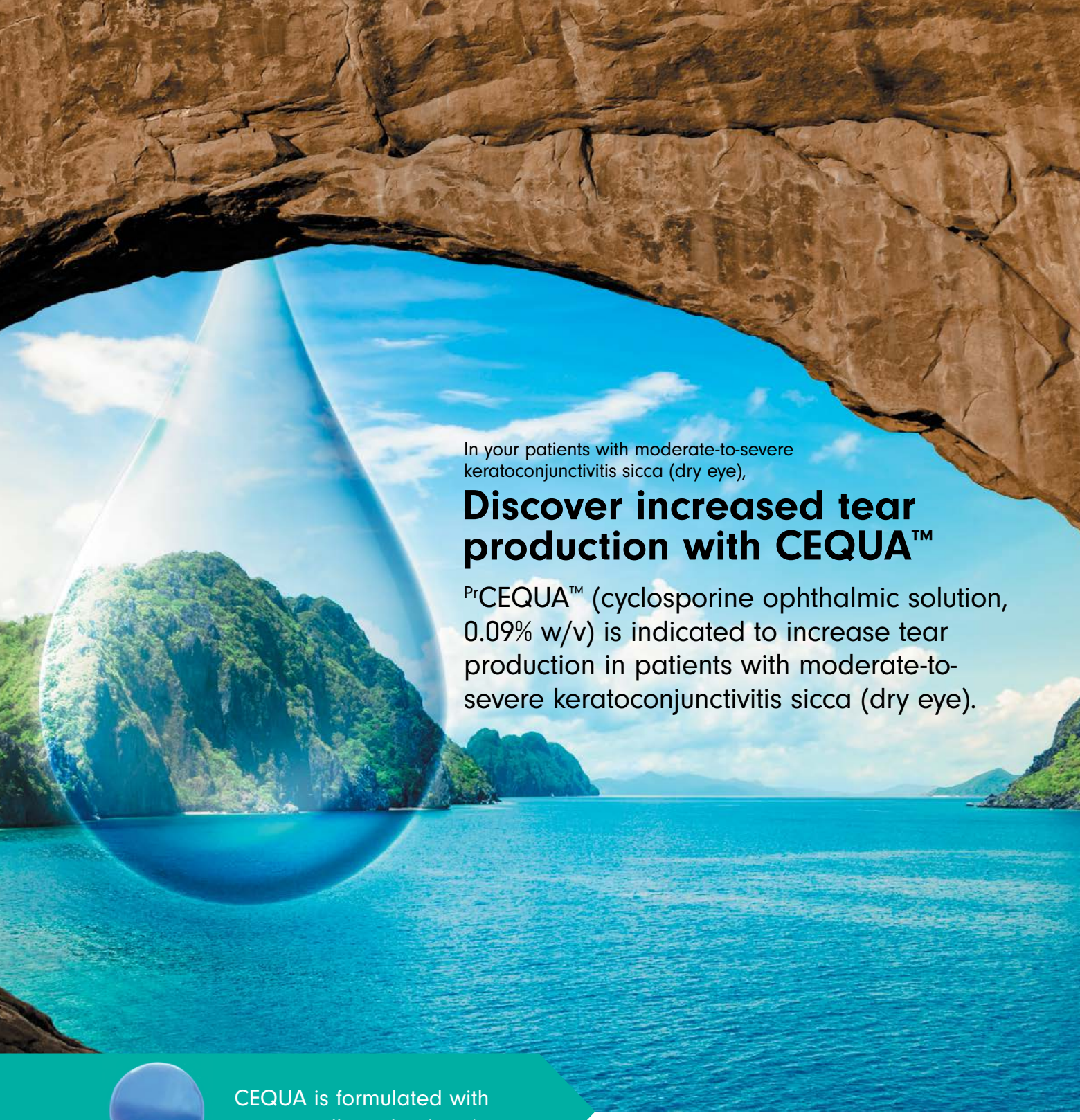
Figure 4: *Left posterior pole 3 months after initial presentation.*



DISCUSSION

Characteristic acute ophthalmoscopic features of Purtscher retinopathy include cotton-wool spots and intraretinal hemorrhages localized to the posterior pole, often peripapillary.[1,3-5,7] Although not observed in our patient, pathognomonic Purtscher flecken are present in roughly 50% of cases.[1,3,6] These are defined as areas of inner retinal whitening localized between arterioles and venules with a characteristic 50μm of uninvolved retina surrounding the vessels.[1,3] This zone of clear retina anatomically corresponds to the capillary-free zone adjacent to retinal arteries and precapillary arterioles, suggesting that Purtscher flecken are a result of occlusion of the precapillary arterioles.[5] The flecken are polygonal in shape and vary in size.[5] Flame hemorrhages are the most common type of retinal hemorrhages, but are often minimal.[5] Less frequently, macular edema and optic nerve edema may additionally be noted.[1] Funduscopic findings have been observed as early as immediately following trauma and may initially progress for the first several days.[3,4] Approximately 60% of cases occur bilaterally; however, asymmetry in presentation may be noted.[1,3,5,8]

Although the diagnosis of Purtscher retinopathy is clinical, ancillary testing is supportive.[5] Intravenous fluorescein angiography findings are variable. Mild cases may demonstrate leakage from the retinal vasculature, while arteriolar occlusion may be evident in severe cases.[1,7] Choroidal fluorescence may be blocked in the setting of flecken or hemorrhage.[5] Capillary nonperfusion, disc leakage, and venous staining have also been documented.[3] Choroidal hypofluorescence on indocyanine green angiography has been reported, suggesting involvement of the choroidal vasculature.[5,9] Hyperreflectivity of the inner plexiform, inner nuclear, and outer plexiform layers is demonstrated



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on optical coherence tomography of Purtscher flecken, indicative of ischemia to the deep capillary plexus.[4] Ancillary testing may thus be used to assess and confirm the presence of these findings.

Visual acuity upon initial presentation may range from 20/20 to hand motion.[3] Visual field testing may demonstrate central or paracentral scotomas in some cases.[3] Though acute retinal signs of Purtscher retinopathy often spontaneously resolve within 1-3 months after the precipitating injury,[5] visual prognosis is variable. Agrawal et al. performed a prospective observational study of 24 eyes in 15 different patients with Purtscher retinopathy (n=12) and Purtscher-like retinopathy (n=3).[6] Of the eyes that received no treatment (n=22), 50% showed an improvement in visual acuity of at least 2 Snellen lines at follow-up, and 23% (n=5) improved at least 4 Snellen lines.[6] A systematic review of Purtscher and Purtscher-like retinopathy performed by Miguel et al. found that, of 22 cases for which 2-month follow-up data were available, 40% exhibited resolution of retinal findings, 64% developed optic atrophy, 23% had retinal pigment epithelium mottling, 14% demonstrated retinal thinning, and 4% had narrowed retinal arteries.[1] Those authors additionally reported that neither Purtscher flecken, choroidal hypoperfusion, nor optic edema was a prognostic factor for visual improvement.[1] Despite the several limitations to this study and review, their findings broadly demonstrate that while visual recovery is not uncommon, permanent sequelae following resolution may lead to varying degrees of vision loss.

The incidence of Purtscher and Purtscher-like retinopathy has been documented to be 0.24 persons per million per year; although this figure is likely an underestimation given that many may be asymptomatic[1, 6] Chuang et al. examined 100 patients following long bone or pelvic fracture and found cotton wool spots and retinal hemorrhages in four patients; however, only one patient was visually symptomatic.[10] It is accepted that the condition is likely underdiagnosed and underreported, and the true incidence may be higher.

The pathogenesis of Purtscher retinopathy remains disputed. Purtscher initially postulated that the retinal findings resulted from lymph extravasation of the retinal vessels due to an abrupt increase in intracranial pressure resulting from head trauma.[5] The discovery that non-traumatic systemic etiologies could induce a Purtscher-like retinopathy has since led to alternative theories.[11] Activation of the complement system and intravascular coagulation are established phenomena following both trauma and acute pancreatitis.[11] Since these conditions are associated with the development of Purtscher and Purtscher-like retinopathy, respectively, it has been speculated that the pathogenesis is linked to complement-mediated occlusion of the small retinal arterioles secondary to the embolization of C5a-induced leukocyte aggregates.[5, 11] This pathway may additionally induce indirect vascular occlusion via complement-mediated endothelial injury, prompting the coagulation cascade.[5] The theory of embolic vascular occlusion was trialed in an experimental study involving the injection of fibrin clots into the ophthalmic artery of animal models, which subsequently led to the development of a Purtscher-like retinopathy.[11]

Air and fat embolization have additionally been implicated as sources of microvascular occlusion in Purtscher retinopathy.[4] Fat embolization is a known complication of long bone fracture.[5,12] In the setting of trauma or orthopedic surgery, fat embolization occurs as a result of intramedullary fat released into the venous circulation.[5,12] In the presence of a patent foramen ovale or pulmonary arteriovenous shunt, it may embolize systemically.[5,12,13] In some instances, fat embolism syndrome (FES) may consequently ensue. Although fat embolization appears to be an inevitable sequela of long bone fractures, FES is a less common complication.[5] FES is defined as a combination of hematological, neurological, respiratory, and cutaneous symptoms, and is fatal in 20% of cases.[3,12,13] Retrospective and prospective studies investigating orthopedic and trauma surgeries have reported that the incidence of FES ranges from 1-19%, and this discrepancy is largely due to ongoing debate over criteria for diagnosis.[12] Up to 60% of patients with FES display retinal findings.[3,4]

More recently, Harrison et al. used computer fluid dynamics simulations of the posterior pole retinal vasculature to evaluate the effects of blood flow rate and vessel bifurcation angles on wall shear stress.[14] As vascular equilibrium is maintained through endothelial cell-mediated autoregulation, the authors theorized that fluctuations in wall shear stress throughout the vasculature, particularly at bifurcations, prompt a chain of rheological events leading to vascular endothelial dysregulation in Purtscher retinopathy[14]

Currently, the theory of microembolization resulting in arteriole occlusion is still the most accepted etiology.[1,14] Clinical, experimental, and histopathological evidence supports this theory.[4,11,14] Alternatively, it is possible that findings may stem from a complex combination of physiological events rather than an isolated mechanism.[14]

Though a conservative observation-only approach is often associated with favorable outcomes, isolated case reports have suggested that treatment with high-dose intravenous steroids may have some benefits, including a hastened visual recovery[1,2,5,6] Conversely, the analysis of 24 cases of Purtscher and Purtscher-like retinopathy performed by Miguel et al. found no statistically significant differences in visual outcome among patients who received corticosteroids compared to those who were simply observed.[1] The use of corticosteroids remains controversial and controlled randomized trials are warranted to both demonstrate an advantage over observation and establish guidelines for management[6,8]

CONCLUSION

Much remains to be understood regarding Purtscher retinopathy. The similar clinical presentation of Purtscher-like retinopathy raises the question of whether these conditions are separate entities, or rather have an overlap in their pathophysiological mechanisms. Obtaining a thorough case history is pertinent in these patients to differentiate between the two. In the absence of trauma, a systemic work-up is indicated to determine the underlying etiology. Presently, most of the available data are from clinical case reports. These case reviews are limited by the lack of a standardized methodology, small sample sizes, and inclusion of non-traumatic etiologies. The rarity of this condition imposes an additional impediment to prospective studies. While there are currently no evidence-based treatments, future advancements in understanding the pathogenesis may allow for avenues of therapeutic intervention. In patients who sustain vision loss, low vision services should be considered to address functional complaints. ●

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