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Case Report

Atypical Purtscher's Retinopathy with Coexisting Central Retinal Vein Occlusion: A Case Report

Jakson Johnson*, Kannan Freyaldenhoven, Camila Albo and Jeong-Hyeon Sohn

Department of Ophthalmology, The University of Texas Health Science Center at San Antonio, San Antonio, Texas, United States of America

***Corresponding Author:** Jakson Johnson, Department of Ophthalmology, The University of Texas Health Science Center at San Antonio, San Antonio, Texas, United States of America.

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Abstract

Purtscher retinopathy's (PR) is a rare microvasculopathy often linked to trauma, presenting with sudden vision loss and characteristic retinal findings. We report an unusual case of PR in a 43-year-old female with hypertension, complicated by prior central retinal vein occlusion (CRVO) and the absence of a clear traumatic etiology. The patient initially presented with progressive vision loss, disc edema, cotton wool spots, dilated venules, and retinal hemorrhages, initially diagnosed as CRVO. Following a hypertensive emergency, fundoscopic exam revealed Purtscher flecken, confirming PR. Etiological considerations included embolic events, hypertensive crisis, and the preceding CRVO. Treatment involved topical and systemic corticosteroids, intravitreal bevacizumab, and triamcinolone acetonide. While clinical findings improved with treatment, visual acuity stabilized at 20/150. This case uniquely highlights PR's occurrence without typical trauma, its overlap with CRVO, and the challenges in determining etiology. The combination of steroids and bevacizumab demonstrated anatomical benefits, though visual recovery was limited. This report underscores the importance of considering PR in atypical presentations and the need for further research to optimize treatment strategies for this rare condition.

Keywords: *Purtscher's Retinopathy; Purtscher-Like Retinopathy; Central Retinal Vein Occlusion; Purtscher Flecken; Macular Edema*

Abbreviations

PR: Purtscher's Retinopathy; CRVO: Central Retinal Vein Occlusion; NLP: No Light Perception; NSC: Nuclear Sclerotic Cataract; DFE: Dilated Fundus Examination; DBH(s): Dot-Blot Hemorrhage(s); NVI: Neovascularization of the Iris; NVD: Neovascularization of the Disc; BP: Blood Pressure; HR: Heart Rate; ER: Emergency Room; AT3: Anti-Thrombin 3; ESR: Erythrocyte Sedimentation Rate; ACE: Acetylcholinesterase; PO: Per Os (Oral)

Introduction

Purtscher's retinopathy is a rare ophthalmic condition that is typically seen in patients with a history of trauma or other systemic conditions, including head and chest trauma, long bone fractures, and acute pancreatitis [1]. The condition has varied presentations, ranging from minimal loss of visual acuity to hand movements in one or both eyes [2]. The loss of visual acuity may be accompanied by visual field loss, manifesting as central, paracentral or arc-shaped blind spots. When post-traumatic, visual symptoms are often delayed by 24-48 hours from the inciting event [1]. While the precise mechanism of visual impairment is unclear, current evidence suggests an

embolic pathophysiology [3]. Fundoscopic exam frequently demonstrates cotton-wool spots and intraretinal hemorrhages, but Purtscher flecken (areas of retinal whitening surrounded by a clear area of retinal arterioles), are pathognomonic [1]. Herein, we report the case of a patient with Purtscher's retinopathy that occurred in a 43-year-old female with no single apparent inciting factor.

Case Description

A 43-year-old obese female with a history of hypertension and right eye blindness secondary to possible central retinal vein occlusion (CRVO) three years ago presented to the emergency department due to painless, progressive vision loss in her left eye (01/13). At the time of presentation, the patient's HR was 115, and BP was 150/99. The patient described the vision loss as beginning centrally five days prior after getting dust in her eye while fixing her cabinets. The patient denied any history of trauma and could not identify any other contributing factors. Ophthalmic examination revealed visual acuity of 20/200, pinhole 20/50 in the left eye, and no light perception (NLP) in the right. Pressure, peripheral field, EOM, and pupillary exam of the left eye were all normal, and no external defects were noted. A slit lamp exam revealed trace nuclear sclerotic cataract (NSC). Dilated fundus examination (DFE) revealed disc edema, cotton wool spots on the disc and macula, dilated venules, and macular and peripheral dot-blot hemorrhages (DBHs). Based on this presentation and previous history of venous occlusion in the right eye, the patient was diagnosed with CRVO of the left eye, sent home, and told to follow up in two to three weeks.

Nine days later (01/22), the patient returned to the ER with complaints of headaches, insomnia, hot flashes, and increased vision loss in the left eye with associated pain. This time, the patient arrived in a state of hypertensive emergency (193/110). Visual acuity assessed in the ER was unchanged from the last visit, and she was not seen by ophthalmology. The patient was treated for hypertensive emergency and discharged.

Eight days later (01/30), the patient is seen in the ophthalmology clinic. At this visit, DFE showed white, vessel-sparing retinal lesions around the optic nerve and macula (Purtscher flecken) and mid-periphery DBHs which were consistent with Purtscher retinopathy (Figure 1). The exam also showed neovascularization of the iris (NVI) and cells in the anterior chamber. Visual acuity had deteriorated to 20/400 in the left eye, and a central field deficit was noted. Lab studies and autoimmunity panels were normal aside from increased AT3 (130), platelets (520), and ESR (88), and decreased ACE (<10), indicative of nothing other than non-specific systemic inflammation. Fundus photos showed multiple intraretinal dots, blot, and flame hemorrhages in all quadrants in the left eye, along with macular edema. The patient was started with one percent prednisolone eye drops tapering program, starting with four drops daily and reducing by one drop per week for four weeks. After three days of steroid drop treatment, the central field defect resolved, and visual acuity improved to 20/200. The patient then received a 1.25 mg Avastin injection in the left eye the following day (02/02/2024) and was further started on a daily prednisone PO tapering program starting at 60 mg daily and reducing by 10 mg per week for four weeks, then reducing by five mg every two weeks for four weeks then continuing on 25 mg daily until vision recovered.

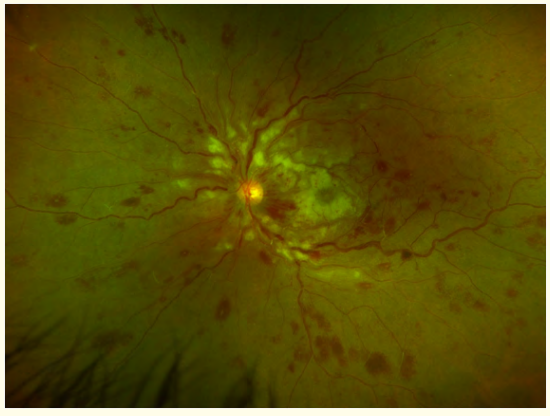

Figure 1: *Fundus photo of the left eye at the time of diagnosis showing multiple Purtscher flecken surrounding the optic nerve and macula, characteristically sparing the ocular vessels. Multiple dot-blot hemorrhages extending to the mid-periphery and engorgement of retinal veins are also visualized.*

Following one week of treatment (02/06), neovascularization of the disc (NVD) began to regress and Purtscher flecken in the periphery were no longer noted, although they remained present in the macula (Figure 2). At this visit, the patient was noted to have a mildly elevated lipase, raising pancreatitis as a possible cause of Purtscher-like retinopathy. Over the next four weeks of tapered steroid therapy, there was consistent, significant regression of Purtscher flecken and macular edema. However, visual acuity showed no stable improvement (03/05). Eight weeks after beginning oral steroid treatment (Figure 3), the patient received a 40 mg Triamcinolone Acetonide injection (04/17). An additional six weeks of low-dose oral prednisone therapy following this injection resulted in mild improvement of visual acuity to 20/150 OS and stabilized macular edema (999 -> 283) (05/22). The patient was ultimately lost to follow-up after completing the steroid taper. Throughout the course of treatment, the patient was screened for numerous autoimmune and infectious causes of her presentation, but none were identified.

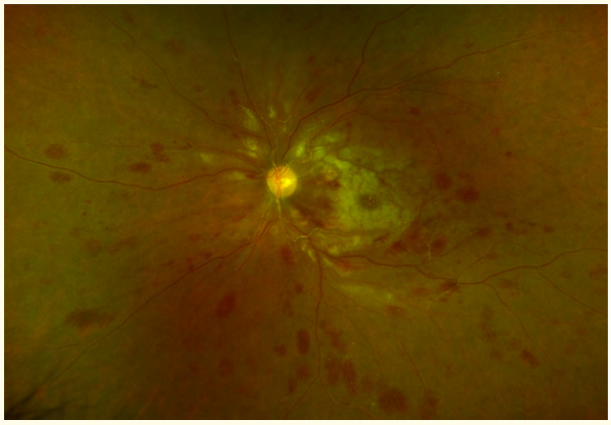

Figure 2: *Fundus photo of the left eye 3 weeks after beginning topical steroid therapy, 17 days post Avastin injection. Shows regression of Purtscher flecken and DBHs. Retinal vessels are no longer engorged.*

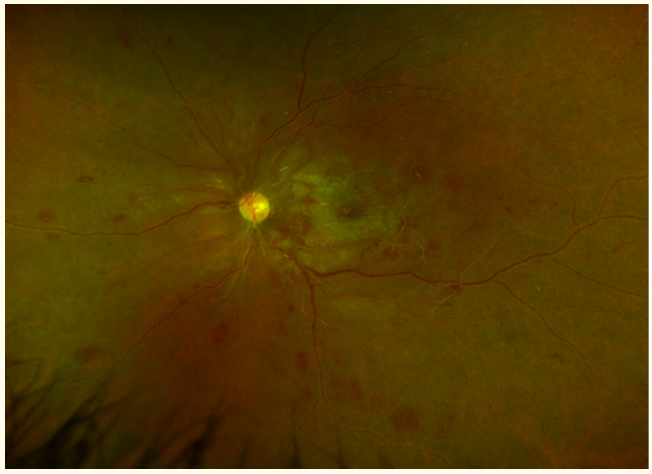

Figure 3: *Fundus photo taken 12 weeks after diagnosis following treatment with topical and oral steroids and avastin injection, showing significantly regressed Purtscher flecken and DBHs.*

Results and Discussion

As mentioned in a systematic review by Miguel., *et al.* the diagnosis of Purtscher's retinopathy may be confirmed by meeting three of the five criteria: Purtscher flecken, retinal hemorrhages, cotton-wool spots, plausible explanatory etiology, and complementary investigation compatible with the diagnosis [4]. While our case adequately meets the first three criteria, its etiology is less clear [4]. Currently, numerous hypotheses exist to explain the pathophysiology of this condition, including lymph extravasation from retinal vessels during a sudden increase in intracranial pressure, arteriolar spasm, and retinal arteriolar embolism [3]. Embolic sources are varied, ranging from air and fat emboli to fibrin clots and leukocyte aggregates [3].

In this case, an embolic cause could not be ruled out, given the patient's history and initial exam findings. Vessel occlusion caused by fibrin clots has been studied in animal models and is known to produce classic findings of Purtscher's Retinopathy including flame-shaped hemorrhages and cotton wool spots, as was evidenced in this patient [5]. Interestingly, the literature implicates embolic occlusion of precapillary arterioles as the culprit behind Purtscher's cases; this contrasts with our case in which findings characteristic of venous occlusion are present [6]. Other cases have associated Purtscher's with acute pancreatitis. The proposed pathology in these instances involves the release of activated proteases following pancreatic inflammation and subsequent activation of complement, leading to coagulation and leukoembolization of retinal arterioles [7]. However, this possibility is less likely given that symptoms of Purtscher's developed prior to mild elevation in lipase levels.

This patient's hypertensive emergency represents another possible cause for our patient's presentation. Reports have frequently implicated trauma as an etiology of Purtscher's, with direct blows to the head causing an increase in intracranial pressure and subsequent lymph extravasation and formation of Purtscher's flecken. While our patient lacked any history of head trauma, they did experience a hypertensive emergency to a BP of 193/110, marked disc edema, and headache, findings which are consistent with increased intracranial pressure. Furthermore, the development of Purtscher's flecken became evident after the episode of hypertensive emergency, which further supports this hypothesis. Nonetheless, the question still remains of whether this patient's increased intracranial pressure was acute enough to be the cause of their presentation.

Additionally, this patient initially presented with findings consistent with CRVO. While it is possible that Purtscher's retinopathy was present at the time of initial evaluation and simply missed on the diagnostic exam, it is also possible that CRVO may have been the initial insult that spurred the development of Purtscher's coinciding with further visual decline over the following week. To our knowledge, there are no other reports of Purtscher's retinopathy with coexisting CRVO which makes it difficult to assess the relation between the two conditions. Regardless, it is highly likely that CRVO played some role in this patient's visual changes and response to treatment and we believe it is important to note as a potential inciting factor in the absence of a clear alternative explanation.

Currently, there are no clear guidelines for the treatment of this condition; physicians are divided in that some advise no treatment while others have suggested the use of corticosteroids, Traditional Chinese Medicine, and hyperbaric oxygen for this condition. One systematic review focused on Purtscher's treatments found that corticosteroids were most commonly used among included studies; however, patients saw improvement in eyesight after six months whether or not any form of treatment was received [2]. In this case, both a topical Prednisolone acetate one percent taper and oral Prednisone taper were employed. Although characteristic exam findings of Purtscher's resolved throughout the course, visual acuity remained poor throughout the treatment period, which contrasts with other cases of Purtscher's that spontaneously resolved without treatment. This may be explained by the presence of other ocular comorbidities or the late detection of the condition, leading to more prolonged visual impairment. The patient also had a number of factors indicating severe disease and poor visual prognosis, namely, potential macular infarction, long duration of acute retinal changes, optic disc swelling, and involvement of the outer retina [9].

Previous cases of Purtscher's with coexisting macular edema have shown improvement after treatment with anti-VEGF therapy (bevacizumab) [8]. For instance, one case report of a patient with Systemic Inflammatory Response Syndrome (SIRS) associated Purtscher's demonstrated additive improvement in associated macular edema when intravitreal bevacizumab was given following a course of systemic corticosteroids [10]. Our case corroborates these findings, as the Avastin injection in conjunction with the systemic steroid course resulted in regression in both macular edema and funduscopic findings of PR. Further treatment with intravitreal Triamcinolone Acetonide was also given in this patient after no measurable improvement in visual acuity with two months of oral steroid therapy (stable at 20/200). The patient's visual acuity did improve (20/150) six weeks after the administration of intravitreal steroids, though it is difficult to establish a causative relationship. One report of stem cell transplant-associated Purtscher's Retinopathy by Castillo., *et al.* found dramatic improvement following prompt steroid injection [11]. The delay in offering this treatment in our case may have precluded more robust recovery.

Conclusion

Purtscher's Retinopathy is a rare ocular condition most often associated with trauma and systemic conditions but may present with overlapping ocular comorbidities, as in this case. The absence of a clear inciting event and the co-occurrence of CRVO make this case particularly unique and valuable for understanding the spectrum of Purtscher's retinopathy. Prompt treatment with steroids (injectable, systemic, and drops) and the possible addition of intravitreal bevacizumab may result in clinical improvement when supportive treatment fails. Further case reports and studies are needed to elucidate the complex interplay of factors contributing to Purtscher's retinopathy and to refine treatment protocols.

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