

# Asymmetric Purtscher-Like Retinopathy Caused by a Hypertensive Emergency and Undiagnosed Type 2 Diabetes

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## Abstract

**Purpose:** To report a case of Purtscher-like retinopathy (PLR) in the setting of a hypertensive emergency and undiagnosed type 2 diabetes mellitus (DM). **Methods:** A case was analyzed. **Results:** A 29-year-old, obese, hypertensive man presented with a 10-day history of progressive vision loss in the left eye with no history of trauma. The visual acuity was 20/25 OD and hand motions OS. A fundus examination showed dilated, tortuous veins; dot-blot and flame hemorrhaging; numerous cotton-wool spots; and polygonal areas of retinal whitening consistent with Purtscher flecken in the right eye and a vitreous hemorrhage in the left eye. The blood pressure was 226/125 mm Hg, and the hemoglobin A<sub>1c</sub> was 11.6%. The patient's presentation was concerning for a hypertensive emergency and type 2 DM as the etiology for the ocular findings. **Conclusions:** The presence of one condition in association with PLR does not exclude the presence of another concurrent etiology.

## Keywords

systemic conditions and the eye, Purtscher-like retinopathy, Purtscher flecken, hypertensive emergency

## Introduction

Purtscher retinopathy was first described in 1910 in a man who fell off a tree and sustained head trauma associated with visual decline.<sup>1</sup> In the initial report, the fundus appearance showed multiple retinal white patches, retinal hemorrhaging, and disc edema. Purtscher retinopathy has since been characterized as a chorioretinopathy associated with cotton-wool spots, minimal intraretinal hemorrhaging, and Purtscher flecken (polygonal areas of retinal whitening between retinal arterioles and venules) typically restricted to the peripapillary region and posterior pole.

Purtscher-like retinopathy (PLR) refers to a similar presentation in the fundus but is not associated with compression trauma. Reported causes of nontraumatic PLR include acute pancreatitis, fat embolization, nephrotic syndrome, connective tissue disorders, cryoglobulinemia, hemolytic uremic syndrome, thrombotic thrombocytopenic purpura, and malignant hypertension.<sup>2,3</sup> New-onset diabetes without hypertension has also been associated with PLR features.<sup>4</sup>

Here, we describe a patient who presented with progressive unilateral vision loss, uncontrolled hypertension, and newly diagnosed type 2 diabetes mellitus (DM) with no history of trauma and was found to have PLR. This case serves as a reminder that the diagnosis of one contributory etiology of PLR does not exclude the coexistence of a second condition associated with PLR.

## Case Report

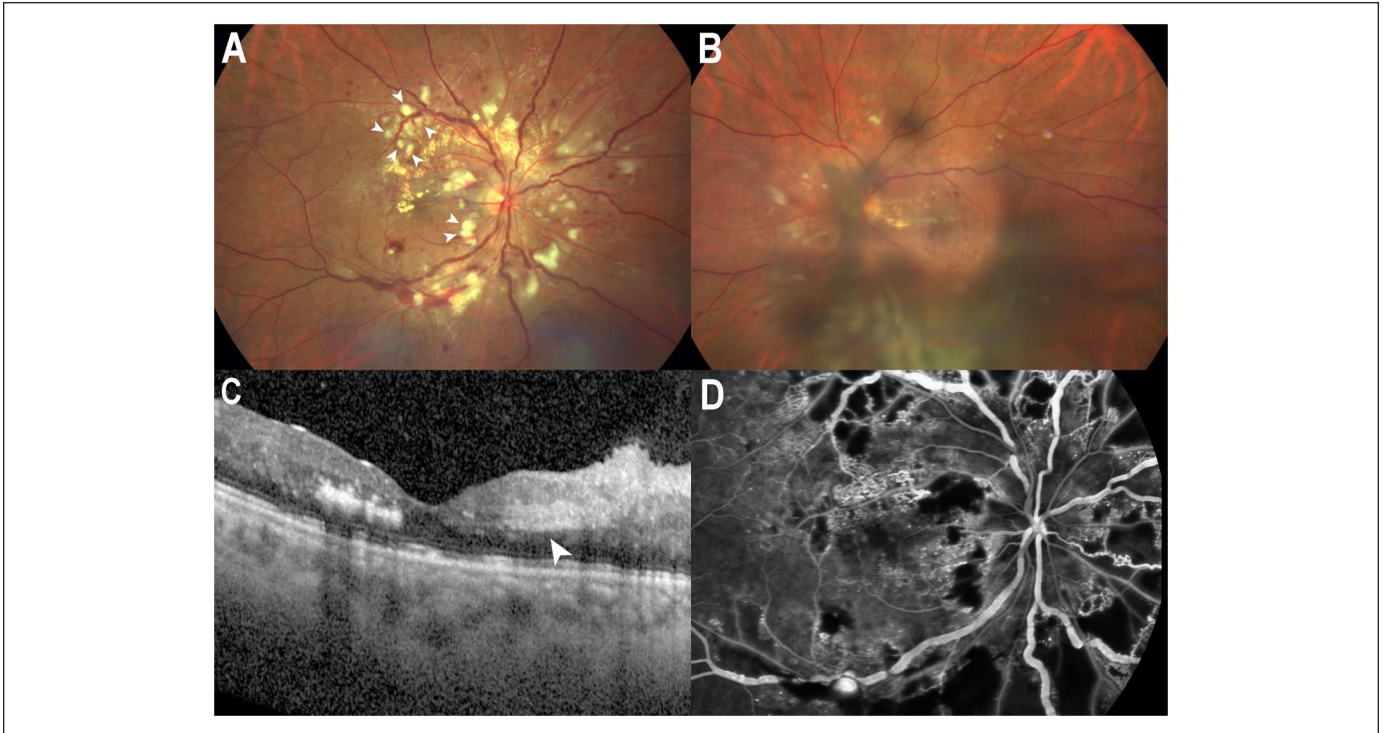
A 29-year-old obese hypertensive man presented to the emergency department with a history of rapidly progressive vision loss in the left eye. He denied eye pain and headaches. The patient had been informed he had hypertension but did not know his blood pressure values and was not taking antihypertensive medication. There was no history of trauma or other known comorbidities.

The initial examination showed a corrected visual acuity (VA) of 20/25 OD and hand motions OS with an intraocular pressure of 13 mm Hg in each eye. Examination of the right eye showed dilated and tortuous retinal veins, a dot-blot and flame hemorrhage, numerous cotton-wool spots in the posterior pole, and multiple areas of polygonal retinal whitening, also in the posterior pole, which largely spared the retina immediately adjacent to the arterioles, consistent with Purtscher flecken (Figure 1A). Examination of the left eye showed a vitreous hemorrhage obscuring some fundus detail; however, a few

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**Figure 1.** (A) Ultra-widefield fundus photograph of the right eye shows dilated and tortuous veins, cotton-wool spots, intraretinal hemorrhage, and areas of polygonal retinal whitening largely sparing the retina immediately adjacent to the arterioles, consistent with Purtscher flecken (arrows), all concentrated in the posterior pole, mimicking Purtscher retinopathy. (B) Ultra-widefield fundus photograph of the left eye shows vitreous hemorrhage obscuring parts of the posterior pole and inferior fundus. Cotton-wool spots and intraretinal heme are present along with lipid exudate in the papillomacular bundle. There is no visible retinal venous dilation or beading. (C) Optical coherence tomography of the right eye shows intraretinal lipid exudate and a focal area of hyperreflectivity in the inner nuclear layer, consistent with paracentral acute middle maculopathy (arrow). (D) Midphase fluorescein angiography of the right eye shows narrow retinal arterioles, dilated retinal veins, intraretinal microvascular abnormalities, venous beading, and marked retinal capillary nonperfusion.

cotton-wool spots, dot-blot heme, and lipid exudate were observed without venous tortuosity (Figure 1B).

Optical coherence tomography (OCT) of the right eye showed juxtafoveal lipid exudate and a focal area of hyperreflectivity in the inner nuclear layer consistent with paracentral acute middle maculopathy (PAMM) (Figure 1C, arrow). Fluorescein angiography (FA) showed extensive areas of retinal capillary nonperfusion, intraretinal microvascular abnormalities, and venous beading in the right eye (Figure 1D). B-scan ultrasonography of the left eye showed an attached retina.

The blood pressure on presentation was 226/125 mm Hg, the body mass index was 33.9 kg/m<sup>2</sup>, and the hemoglobin A<sub>1c</sub> (HbA<sub>1c</sub>) was 11.6%. The diagnosis was PLR in the right eye associated with a hypertensive emergency and concurrent newly diagnosed uncontrolled type 2 DM. Given the hypertensive emergency, the patient was treated with an intravenous beta blocker and admitted to the hospital. The patient's hyperglycemia was treated with insulin. A workup for secondary causes of hypertension was negative.

Two weeks after presentation, the patient received intravitreal bevacizumab (Avastin, Genentech) in the left eye. Six weeks after presentation, the VA in the left eye improved to 20/32 and was unchanged in the right eye at 20/25. The blood

pressure was 149/96 mm Hg. A follow-up ultra-widefield fundus photograph of the right eye showed fewer cotton-wool spots and Purtscher flecken as well as less intraretinal hemorrhaging (Figure 2A). Follow-up OCT of the right eye showed less retinal thickening, less lipid exudate, and improved PAMM (Figure 2B). The patient was scheduled for another intravitreal injection in the left eye 4 weeks later.

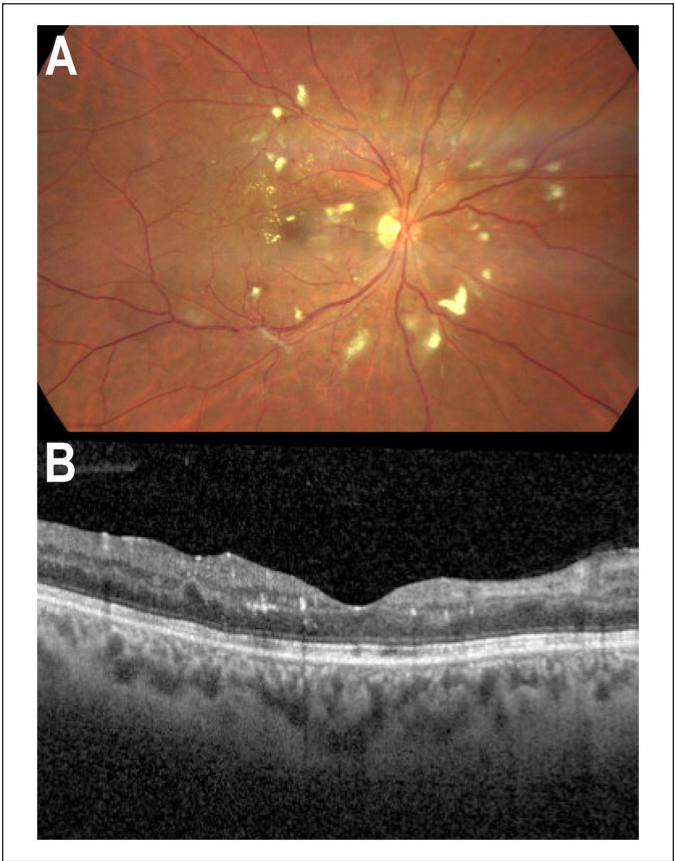
## Conclusions

This patient presented with progressive unilateral vision loss in the left eye without trauma or systemic symptoms and was subsequently found to have PLR in the right eye. Because of the vitreous hemorrhage in the left eye on presentation, a diagnosis of bilateral, or symmetric, PLR could not be confirmed. Concern was appropriately directed at the underlying conditions of the hypertensive emergency and uncontrolled type 2 DM.

A systematic review of Purtscher retinopathy and PLR<sup>5</sup> found that the most frequently encountered ophthalmoscopic signs in PLR were cotton-wool spots (seen in 93% of cases) followed by intraretinal hemorrhage (65%) and Purtscher flecken (63%). Purtscher flecken are pathognomonic findings and can be described as polygonal areas of retinal whitening that occur in

the inner retina, between retinal arterioles and venules. Cotton-wool spots are hypothesized to develop as a result of obstruction of a retinal arteriole and are commonly observed in DM and systemic hypertension. Purtscher flecken can be distinguished from cotton-wool spots by their appearance, anatomic location, and retinal imaging features (Table 1).<sup>6,7</sup>

The OCT findings in our case showed areas of hyper-reflectivity of the inner nuclear layers (ie, PAMM) (Figure 1C),



**Figure 2.** (A) Ultra-widefield fundus photograph of the right eye shows fewer cotton-wool spots, Purtscher flecken, and intraretinal hemorrhage. (B) Optical coherence tomography of the right eye shows less retinal thickening, less lipid exudate, and improved paracentral acute middle maculopathy.

indicating ischemia in the inner retinal circulation, which is characteristic of PLR.<sup>8,9</sup> Midphase FA of the right eye showed marked retinal capillary nonperfusion underlying areas of Purtscher flecken (Figure 1D), another finding characteristic of PLR.<sup>10</sup>

The improvement at 6 weeks in the right eye (ie, fewer cotton-wool spots and Purtscher flecken as well as less intraretinal hemorrhaging) (Figure 2A) is consistent with results of a systematic review of Purtscher retinopathy and PLR, which reported that normalization of the retinal appearance on ophthalmoscopic examination was present in 40% of patients at the 2-month follow-up.<sup>5</sup>

Several hypotheses have been proposed for the pathogenesis of Purtscher retinopathy and PLR. One such hypothesis is that activation of the complement cascade, in particular complement component C5, leads to complement-induced leukoembolization of precapillary arterioles. This mechanism is supported by the association of PLR with acute pancreatitis and autoimmune and vasculitic diseases.<sup>2,6,11</sup> Angiospasm caused by an acute rise in venous pressure from compressive trauma has also been hypothesized.<sup>2</sup>

In our case, endothelial damage associated with our patient’s hypertensive emergency could have initiated the vascular inflammation, vascular remodeling, and shift toward a proinflammatory and prothrombotic state that promoted development of an occlusive vasculopathy.<sup>12</sup> An additional factor in our case might have been the elevated blood glucose from uncontrolled type 2 DM. When the endothelium is exposed to hyperglycemia, intracellular events promote endothelial dysfunction.<sup>13</sup> Subsequent activation of leukocytes and leukocyte adhesion may have promoted capillary obstruction and damage to the inner retinal capillary bed.

Our case is noteworthy because of the simultaneous occurrence of 2 acute medical conditions at the time of presentation; that is, a hypertensive emergency and newly diagnosed type 2 diabetes with a markedly elevated HbA<sub>1c</sub>. Although each condition has been separately associated with the development of PLR, to our knowledge they have not been previously reported in the same patient as simultaneous etiologies for PLR at the time of presentation.<sup>3,4</sup> The documented bidirectional relationship between a hypertensive crisis and DM, where DM amplifies the cardiovascular risk associated with a

**Table 1.** Retinal Findings: Purtscher Flecken vs Cotton-Wool Spots.

Characteristic	Purtscher Flecken	Cotton-Wool Spots
Appearance	Polygonal areas of retinal whitening	Fluffy white patches with indistinct borders; may obscure underlying vessels
Anatomic location	Inner retina between retinal arterioles and venules	Within retinal nerve fiber layer
Pathogenesis	Occlusion of retinal capillary bed	Focal infarcts lead to accumulation of neuronal debris
OCT	Hyperreflectivity involving inner plexiform and inner nuclear and outer plexiform layers; band-like hyperreflective lesion at inner nuclear layer indicating PAMM	Focal thickening and hyperreflectivity of retinal nerve fiber layer
FA	Hypofluorescent areas corresponding to areas of capillary nonperfusion	May reveal areas of capillary nonperfusion adjacent to cotton-wool spots

Abbreviations: FA, fluorescein angiography; OCT, optical coherence tomography; PAMM, paracentral acute middle maculopathy.



hypertensive crisis, and conversely the fact that elevated blood pressure increases the risk for microvascular and macrovascular complications of diabetes, may be associated with persistent neurohormonal and inflammatory activation.<sup>14</sup> This suggests that a combination of metabolic and hemodynamic factors could have contributed to endothelial dysfunction and the development of PLR in our patient.

Our patient did not present with decreased VA in the right eye. Although PLR is typically associated with decreased vision at presentation, the mean VA can vary at presentation and may be 20/20.<sup>6,15</sup> In a case associated with coil embolization and balloon occlusion of an intracavernous carotid aneurysm,<sup>16</sup> it was hypothesized that microemboli from a thrombosed aneurysm caused multifocal areas of retinal infarction that gave the appearance of PLR as well as Horner syndrome; the presenting VA in that case was 20/20 in both eyes.

In conclusion, we present a unique case of PLR in the setting of both a hypertensive emergency and newly diagnosed, uncontrolled type 2 DM. With treatment of these underlying etiologies, the characteristic fundoscopic and OCT findings of PLR improved. This case serves as a reminder that more than one concurrent etiology of a PLR can indeed be present.

### Ethical Approval

This case report was conducted in accordance with Declaration of Helsinki. The collection and evaluation of all protected patient health information was performed in a US Health Insurance Portability and Accountability Act-compliant manner.

### Statement of Informed Consent

Informed consent was obtained from the patient for publication of all photographs and clinical information included herein.

### Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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