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## Purtscher Like Retinopathy Secondary to HELLP Syndrome: A Case Report

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### Abstract:

**Background:** Purtscher like retinopathy is a rare ocular condition with a clinical appearance similar to Purtscher retinopathy, without any history of trauma. It presents with acute vision loss in the presence of some underlying disease with retinal hemorrhages present within the posterior pole of the eye on ophthalmic examination. Mainstay of management is corticosteroid therapy with clinical improvement in 1-3 months. Presented here is a case of a pregnant lady with Purtscher like retinopathy.

**Case Presentation:** A 28-year lady gravida 2 para 1 was referred to our tertiary care ophthalmology service with chief complaints of sudden loss of vision in both eyes. She was a known case of pregnancy induced hypertension at 32+1 weeks and was on 3 weekly follow up with her gynecologist when she was found to have a blood pressure of 210/130 mmHg. She was admitted to the high dependency unit (HDU). On account of her preeclampsia, her pregnancy was terminated at 35 weeks and she had an uneventful delivery of a healthy baby boy. On ocular exam she had a visual acuity of 6/46 in both eyes unaided. On fundus exam she had flame shaped hemorrhages, peripapillary cotton wool spots and macular star in both eyes. Ultrawide field fundus fluorescein angiography showed hypofluorescence in areas of purtscher flecken. Based on examination findings and clinical presentation she was diagnosed as a case of Purtscher Like Retinopathy. On serial exams her retinal thickening resolved and her visual acuity improved to 6/9 unaided in both eyes at 4 weeks.

**Conclusions:** Purtscher like retinopathy secondary to HELLP syndrome is quite a rare disease with a clinical diagnosis. Examination findings such as Purtscher flecken are highly characteristic of the disease. Presence of associated PuR with acute conditions such as acute pancreatitis may be associated with worsening clinical outcomes. Further study is recommended to identify any such associations between PuR and clinical outcome of other associated illnesses. The condition itself however appears to be self-limiting and patients often recover from visual loss without any particular ophthalmic intervention.

**Keywords:** Purtscher Retinopathy, Purtscher Like Retinopathy, Optical Coherence Tomography, Flame-shaped hemorrhages, Macular star

### BACKGROUND

The first known case of Purtscher retinopathy (PR) was described in 1910 by Omar Purtscher, an Austrian ophthalmologist, in a middle-aged man who developed vision loss after falling off a tree and developing head trauma. Ophthalmic examination revealed retinal hemorrhages in the posterior pole of both eyes. Despite initial vision loss, patient fully recovered vision<sup>1</sup>.

Purtscher Like Retinopathy (PuR) is the term used to refer to the condition with a clinical appearance similar to

Purtscher retinopathy but without any history of trauma.

While the exact cause of either is not known it is believed that embolic occlusion of ophthalmologic vessels may play a role in its pathogenesis<sup>2,4</sup>.

Accurate determination of its incidence has not yet been made, but according to current estimates its incidence lies at 0.24 persons per million per year<sup>7</sup>. Purtscher like retinopathy has been reported to occur with a variety of conditions such as acute pancreatitis, child birth, lymphoproliferative disorders, chest compression, fat

embolization, Valsalva maneuver, after retrobulbar anesthesia, chronic renal insufficiency and others<sup>3-5</sup>.

Clinical presentation most often involves loss of visual acuity with variation in its severity depending on the patient. Fundoscopic examination reveals retinal hemorrhages, optic disc swelling and cotton wool spots present within the posterior pole of the eye. These changes present within 24-48 hours of onset of the causative illness and often resolve spontaneously in 1-3 months<sup>2,6</sup>.

Presented here is a case of Purtscher like retinopathy secondary to HELLP Syndrome.

#### **Ethics Declaration:**

Ethical approval and consent to participate: Written informed consent was obtained from the patient for publication of this case report and accompanying data. It was approved by the Institutional Review Board for Clinical Research committee of Pak-Emirates Military Hospital. All procedures were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

#### **CASE PRESENTATION**

A 28-year lady gravida 2 para 1 was referred to our tertiary care ophthalmology service with chief complaints of sudden loss of vision in both eyes for the last 3 weeks. She was 35 weeks pregnant when she experienced sudden painless visual loss in both eyes, neither associated with floaters nor with flashes of light. She was a known case of pregnancy induced hypertension at 32+1 weeks and was on 3 weekly follow up with her gynecologist when she was found to have a blood pressure of 210/130 mmHg. She was admitted to the high dependency unit (HDU) for further management as a case of preeclampsia. On baseline investigations she was found to have low platelet levels ( $67,000/\text{mm}^3$  [reference range – 150,000 – 400,000/ $\text{mm}^3$ ]) and raised ALT levels (500 IU - reference range <46). On account of her preeclampsia, her pregnancy was terminated at 35 weeks and she had an uneventful delivery of a healthy baby boy.

In order to assess her vision loss, further investigations were carried out. Her blood complete picture, chest x-ray,

prothrombin time, partial thromboplastin time, renal function tests and ultrasound-kidney, ureter, bladder were normal. Anti-nuclear antibody and anti-double stranded deoxyribonucleic acid (DNA) antibodies were negative. Her erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and D-dimer levels were all normal.

On ocular exam she had a visual acuity of 6/46 in both eyes unaided, not improving by pinhole or refraction. Color vision was 17/17 bilaterally. Her pupils were equally reactive to light and there was no relative afferent pupillary defect. Anterior segment exam of both eyes was unremarkable. On fundus exam she had flame shaped hemorrhages, peripapillary cotton wool spots and macular star visible in both eyes as shown in figures 1 and 2.

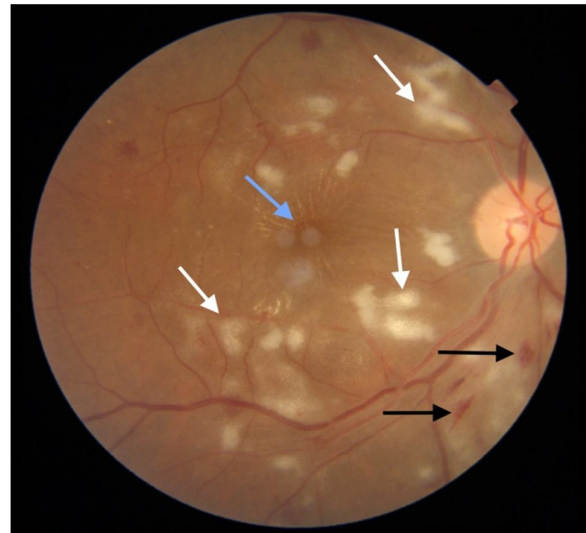


Figure 1: Fundus exam of right eye showing cotton wool spots (white arrows), retinal hemorrhages (black arrows) and macular star (blue arrow)

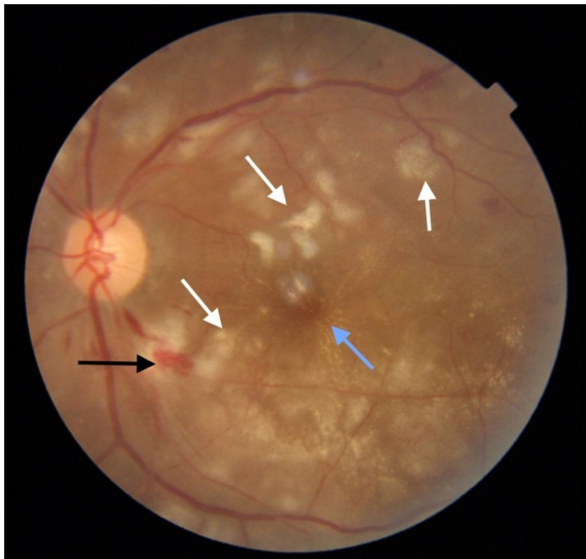


Figure 2: Fundus exam of left eye showing cotton wool spots (white arrows), retinal hemorrhages (black arrows) and macular star (blue arrow)

Optic discs had intact borders with cup to disc ratio of 0.3, peripapillary whitish flecken (Purtscher Flecken) and a normal retinal vasculature bilaterally visible in figures 3 and 4. Her optical coherence tomography macula showed a central macular thickness of 340 mm the right while that of 356 mm in the left (figure 3 and 4).

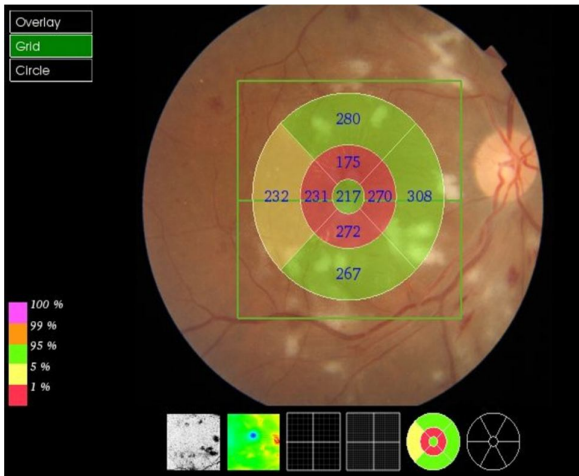


Figure 3: Optical Coherence Tomography macula right eye

Figure 4: Optical Coherence Tomography macula left eye  
 Ultrawide field fundus fluorescein angiography showed hypofluorescence in areas of Purtscher flecken, cotton wool spots and hemorrhages due to blocked choroidal fluorescence. Few areas of punctate hyper fluorescence are seen in extra foveal areas of right eye and supero-

nasal quadrant of left eye signifying leakage visible in figures 5 and 6 in the right and left eyes respectively. Based on the history, clinical exam and supportive investigations she was diagnosed to be a case of bilateral Purtscher like Retinopathy secondary to HELLP Syndrome. On serial exams her retinal thickening resolved and her visual acuity improved to 6/9 unaided in both eyes at 4 weeks.

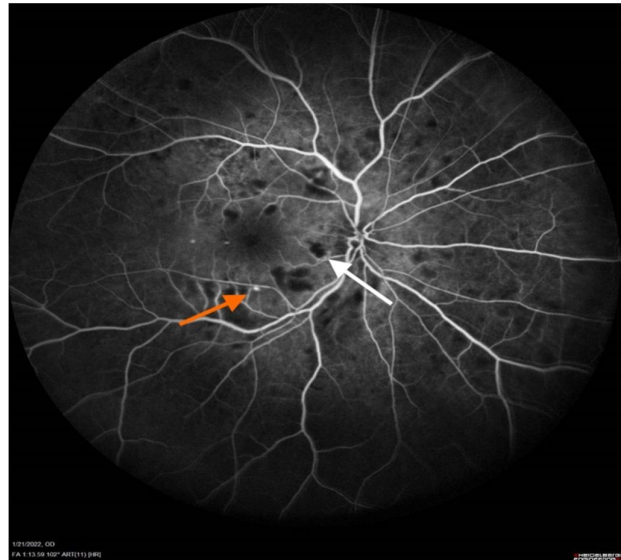


Figure 5: Fundus Fluorescein Angiography of right eye showing hypofluorescence in areas of Purtscher flecken (white arrows), cotton wool spots (yellow arrows)



Figure 6: Fundus Fluorescein Angiography of left eye showing hypofluorescence in areas of Purtscher flecken (white arrows), cotton wool spots (yellow arrows)

## DISCUSSION

Purtscher retinopathy is a rare condition which occurs primarily due to trauma to the head. Purtscher like retinopathy has a similar clinical and examination appearance to Purtscher retinopathy, however the inciting event is an underlying systemic illness. In either case, patients present with acute vision loss that may be uni - or bilateral. The severity of the vision loss can vary to minimal vision impairment to only being able to see hand movements. Visual field loss can present as multiple scotomata while peripheral vision is often preserved<sup>2</sup>.

While the diagnosis of Purtscher like retinopathy is clinical there are certain examination findings that can point one in the right direction. The most frequent finding on fundoscopy is cotton wool spots. Retinal hemorrhage and Purtscher flecken are also seen<sup>7</sup>. Purtscher flecken are pathognomic of PuR and are defined as areas of inner retinal whitening on fundoscopic examination with a clear demarcation between affected and normal retina. This sparing of perivascular retina is very characteristic of the disease but it may be absent in upto 50% of cases<sup>8</sup>. If this whitening is present around the fovea, it can cause the appearance of a pseudo cherry red spot which can cause confusion and misdiagnosis as central retinal artery occlusion<sup>2</sup>.

The exact mechanism of Purtscher and Purtscher like retinopathy is not known. The characteristic examination findings are often confined to the interior retina, between the retinal arterioles and venules. This, coupled with the fact that bilateral eye involvement is common and occlusion of retinal vessels is found on fluorescein angiography, suggests that the underlying pathogenesis is embolic in nature<sup>2</sup>.

Studies have been done to determine the frequency of conditions which cause associated Purtscher like retinopathy. According to a systemic review the most common cause of Purtscher retinopathy is acute trauma. The leading cause of Purtscher like retinopathy is acute pancreatitis<sup>7</sup>. Whether or not there is a statistically significant relation is subject to further research as the condition itself is quite rare and studies may lack adequate statistical power. In patients of acute

pancreatitis, presentation of Purtscher like retinopathy is associated with poor outcome and indicate imminent multiorgan failure<sup>9</sup>.

Currently there are no definite treatment guidelines for Purtscher and Purtscher like retinopathy. Management of underlying condition causing the vision loss can cause clinical improvement<sup>2</sup>. According to a systemic review in China, treatment options used in different cases include glucocorticoid therapy, traditional Chinese medicine therapy, glucocorticoid integrative medicine therapy and integrative medicine therapy. Regardless of whether patients received treatment or not, visual acuity improved within 1-3 months with no significant differences between treatment and non-treatment groups<sup>10</sup>. Since the proposed pathogenesis is due to embolic occlusion of retinal vessels, hyperbaric oxygen therapy may improve the condition but no published reports have been found where hyperbaric oxygen has been used for treatment of Purtscher and Purtscher like retinopathy. No studies have been reported which provide definitive evidence of the benefit of treatment.

## CONCLUSION

Purtscher like retinopathy secondary to HELLP syndrome is quite a rare entity and diagnosis is clinical. Examination findings such as retinal hemorrhage and Purtscher flecken are highly characteristic of the disease and should aid in diagnosis. Presence of associated PuR (Purtscher and Purtscher-like retinopathies) with acute conditions such as acute pancreatitis may be associated with worsening clinical outcomes. Further study is recommended to identify any such associations between PuR and clinical outcome of other associated illnesses. The condition itself however appears to be self-limiting and patients often recover from visual loss without any particular ophthalmic intervention.

## Abbreviations:

HDU: High Dependency Unit

HELLP: Hemolysis Elevated Liver Enzymes Low Platelets

PuR: Purtscher Like Retinopathy

PR: Purtscher Retinopathy

ALT: Alanine transaminase

IU: International Units

DNA: Deoxyribonucleic acid

ESR: Erythrocyte sedimentation rate

CRP: C reactive protein

OCT: Optical coherence tomography

FFA: Fundus fluorescein angiography

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#### DATA AVAILABILITY

All data are included in this article.

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#### Ethics declarations

#### Conflict of interest

Taimoor A Khan, Talha Liaqat, Mohammad A Mehboob, Asfandyar Khan, Ahsan Mukhtar. all authors have no conflicts of interest that are directly relevant to the content of this review.

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