Open Access Library Journal
2024, Volume 11, e12445
ISSN Online: 2333-9721
ISSN Print: 2333-9705

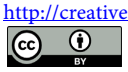
Purtscher-Like Retinopathy in Hemolytic Uremic Syndrome: A Case Report

Leon Zhang, Joshua Meyerov, Michael Dong, Nandini Singh

Department of Ophthalmology, The Alfred Hospital, Melbourne, Australia
Email: leonzjzhang@gmail.com

How to cite this paper: Zhang, L., Meyerov, J., Dong, M. and Singh, N. (2024) Purtscher-Like Retinopathy in Hemolytic Uremic Syndrome: A Case Report. *Open Access Library Journal*, **11**: e12445.
https://doi.org/10.4236/oalib.1112445

Received: October 9, 2024
Accepted: November 23, 2024
Published: November 26, 2024



Abstract

Purtscher-like retinopathies (PLRs) are occlusive retinal microangiopathies arising from non-traumatic etiologies with characteristic fundus findings. We present a case of PLR secondary to typical hemolytic uremic syndrome (HUS) that achieved an excellent visual outcome with treatment of the systemic disease alone. This report highlights that PLR should be considered as a differential for retinopathy in the setting of systemic illness. Furthermore, the treatment of the precipitant cause should be prioritized, with ongoing monitoring of visual function and screening for chronic ischemic complications.

Subject Areas

Hematology, Ophthalmology

Keywords

Purtscher-Like Retinopathy, Purtscher Retinopathy, Hemolytic Uremic Syndrome, Microangiopathic Hemolytic Anemia, Case Report

1. Introduction

Purtscher-like retinopathies (PLRs) are occlusive retinal microangiopathies with a broad array of non-traumatic etiologies [1]. PLR often presents characteristic ischemic fundus changes that correspond to variable degrees of visual loss [1] [2]. Despite the lack of standardized treatment guidelines, reversal of the precipitant cause and ongoing monitoring for ischemic complications are critical for optimizing visual outcomes.

2. Case Presentation

A 27-year-old female with HUS was referred for assessment of acute visual

disturbances of blurriness, visual hallucinations and intermittent diplopia. Over the preceding two weeks, she had been managed in the intensive care unit with anuric renal failure (eGFR 9, Cr 530) requiring hemodialysis, thrombocytopenia (platelet count nadir 59) and metabolic encephalopathy.

Visual acuity was 20/20 bilaterally, with no ophthalmoplegia or pain in eye movements. Intraocular pressures were within normal limits. Anterior segment examination was unremarkable. Dilated fundus exam demonstrated bilateral peripapillary and left inferior macular cotton wool spots (CWSs), more prominent in the left eye, and two small hemorrhages near the superotemporal arcade in the right eye (Figure 1). There were no central macular lesions in either eye. In the context of her systemic disease, the fundus findings were consistent with PLR. The visual hallucinations were attributed to the severe encephalopathy.

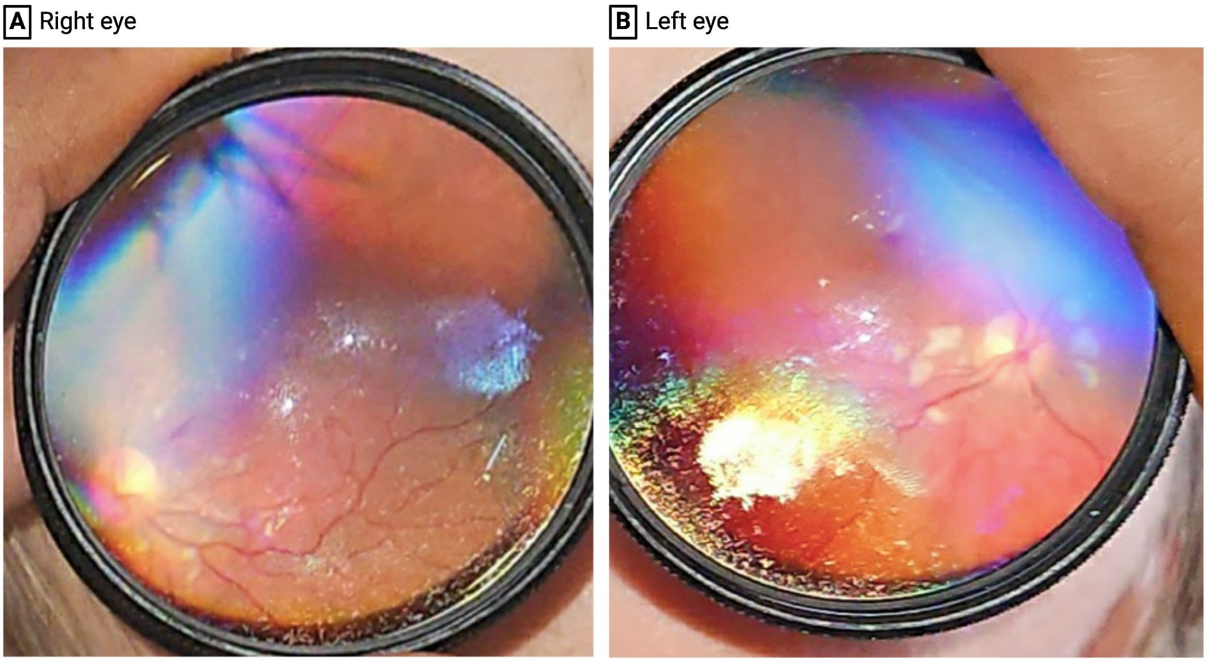


Figure 1. Binocular indirect ophthalmoscopy of both eyes.

The treatment of her systemic microangiopathic disease was prioritized, with serial ophthalmology reviews to monitor visual function and retinopathy. The patient had already received two doses of eculizumab (anti-C5 monoclonal antibody therapy), followed by plasmapheresis (PLEX), given her acutely worsening neurological status. Following five rounds of PLEX, she improved both clinically and biochemically. Six weeks post-discharge, all visual symptoms had resolved, and visual acuity remained preserved. Color fundus imaging demonstrated isolated CWSs (Figure 2(A)), with corresponding superficial retinal plexus capillary drop-out on OCT angiography. Optical coherence tomography (OCT) demonstrated focal inner retinal hyperreflectivity and thickening, corresponding to the CWSs, and trace intraretinal fluid (Figure 2(C), Figure 2(E)). Fluorescein angiography, however, did not demonstrate any significant retinal ischemia or vascular leakage.

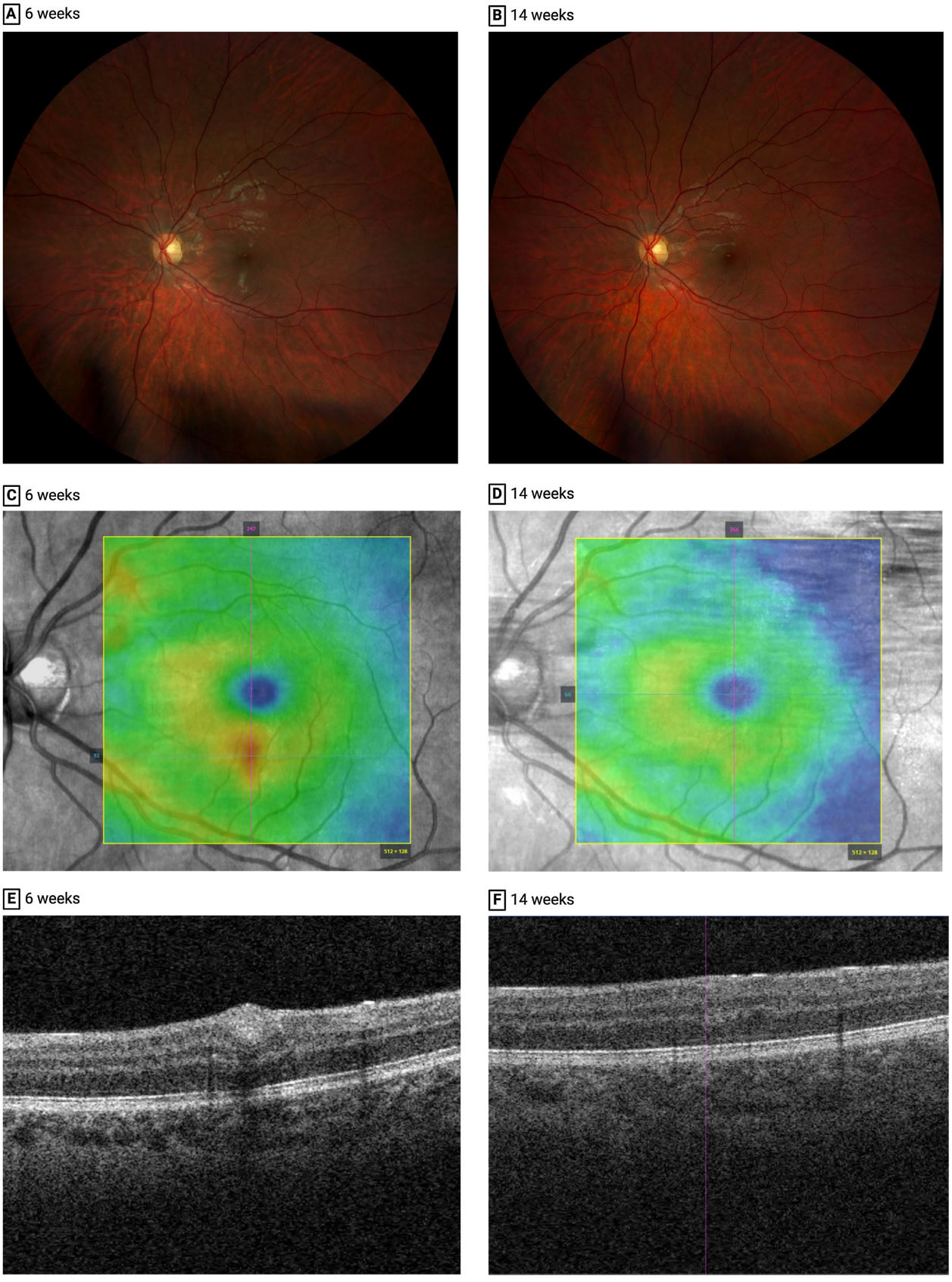


Figure 2. Images of the left eye over time. ((A) (B)) Color fundus images; (C) - (F) Optical coherence tomography.

Serial fundus and OCT imaging showed resolving cotton wool spots and corresponding inner retinal hyperreflectivity (**Figure 2(B)**, **Figure 2(D)**, **Figure 2(F)**), with otherwise normal optic disc and macula anatomy.

3. Discussion

PLR has a broad array of precipitant causes, including thrombotic microangiopathies, such as HUS [1]. Retinopathy is presumed to arise from precapillary occlusion with complement-mediated leukoaggregates, causing retinal tissue ischemia [2]. Fundus features include CWSs located in the posterior pole, sometimes with zones of clearing between retinal arterioles, termed Purtscher Flecken [2]. As in this case, intraretinal hemorrhages may also occur [2]. Vision loss is possible, ranging from minimal to profound impairment [2]. Our patient's excellent visual acuity can be explained by the central macula-sparing location of the retinal lesions.

Often, visual loss in PLR is self-limiting, with complete or partial resolution of retinopathy within weeks from onset. The risk of ischemic sequelae is higher with more severe visual impairment, longer duration of retinopathy and if there are other inflammatory or vascular risk factors [3].

Despite the lack of standardized treatment guidelines for PLR/PR, it is evident from this case that treating the inciting cause is critical for optimizing visual outcomes. There is some evidence for using high-dose steroids and hyperbaric oxygen therapy in the acute setting, however, the reported outcomes are variable [4] [5]. All patients should also be monitored closely for ischemic complications, including neovascularization. In such cases, anti-angiogenic agents and laser photocoagulation have been utilized to suppress the ischemic drive [6].

4. Conclusion

This report documents a patient with PLR in the setting of HUS who achieved an excellent visual outcome with the treatment of the systemic disease alone. Clinicians should consider PLR as a differential for retinopathy in the setting of systemic illness. Treatment of the precipitant cause should be prioritized, with ongoing monitoring of visual function and screening for chronic ischemic complications in the long term.

Conflicts of Interest

The authors declare no conflicts of interest.

References

[1] Agrawal, A. and McKibbin, M.A. (2006) Purtscher's and Purtscher-Like Retinopathies: A Review. *Survey of Ophthalmology*, **51**, 129-136. https://doi.org/10.1016/j.survophthal.2005.12.003

[2] Miguel, A.I.M., Henriques, F., Azevedo, L.F.R., Loureiro, A.J.R. and Maberley, D.A.L. (2012) Systematic Review of Purtscher's and Purtscher-Like Retinopathies. *Eye*, **27**, 1-13. https://doi.org/10.1038/eye.2012.222

[3] Lee, D.H., Hwang, C.K., Cukras, C.A., Wiley, H.E., Malik, F.F., Chew, E.Y., *et al.* (2020) Two Cases of Severe Purtscher-Like Retinopathy Demonstrating Recurrence and Progression to Neovascularization and Vitreous Hemorrhage. *American Journal of Ophthalmology Case Reports*, **18**, Article ID: 100664. https://doi.org/10.1016/j.ajoc.2020.100664

[4] Wu, C., Dai, R., Dong, F. and Wang, Q. (2014) Purtscher-Like Retinopathy in Systemic Lupus Erythematosus. *American Journal of Ophthalmology*, **158**, 1335-1341.E1. https://doi.org/10.1016/j.ajo.2014.09.001

[5] Lee, C., Hsieh, T., Chu, C., Hsu, Y., Wang, J., Wang, J., *et al.* (2023) Hyperbaric Oxygen Therapy as Rescue Therapy for Pediatric Frosted Branch Angiitis with Purtscher-Like Retinopathy: A Case Report. *Frontiers in Medicine*, **10**, Article 1119623. https://doi.org/10.3389/fmed.2023.1119623

[6] Meng, L., Yu, Q., Zhao, X., Chen, L., Wang, Y., Zhang, W., *et al.* (2023) Purtscher-Like Retinopathy in Systemic Lupus Erythematosus: Clinical Features, Risk Factors and Prognosis. *QJM*: *An International Journal of Medicine*, **116**, 923-932. https://doi.org/10.1093/qjmed/hcad204