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Severe proliferative retinopathy in a patient with sickle cell trait

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1. Case

A 21-year-old female with past medical history significant for sickle cell trait (SCT) and autoimmune hepatitis presented with floaters and blurry vision in her left eye. Visual acuity was 20/25 OU. Fundus examination of the left eye demonstrated a dense pre-retinal hemorrhage that spared the premacular bursa and extensive mid-peripheral fibrovascular proliferation with a macula-sparing tractional retinal detachment (Fig. 1). The right eye was asymptomatic, and had less extensive peripheral ischemia with several areas of early retinal neovascularization without traction (Fig. 2). The patient would undergo pars plana vitrectomy (PPV) in the left eye with a subsequent need for repeat PPV two years later for recurrent proliferative vitreoretinopathy (PVR) and tractional retinal detachment. The right eye was managed with panretinal photocoagulation (PRP) with a good therapeutic response.

2. Discussion

To the best of our knowledge this represents the first case of severe proliferative sickle cell retinopathy (PSCR) (Goldberg classification Stage V) in a SCT patient. Patients with sickle cell disorders, including sickle cell disease, sickle cell-hemoglobin C and sickle-thalassemia, are at increased risk for sight-threatening retinal disease which necessitates, at minimum, yearly screening. In contradistinction, SCT is often considered benign, resulting in undiagnosed PSCR as seen with our patient who had not undergone dilated ophthalmological evaluation in the five years prior to presentation. Nagpal and colleagues reported seven cases of PSCR in SCT patients, all of whom had underlying systemic pathology (tuberculosis, syphilis, sarcoidosis, diabetes). Similarly, it is possible that the diagnosis of autoimmune hepatitis one year prior to the evaluation of our patient may have contributed to the severity of her retinopathy. The authors hypothesized that the concomitant disease promotes red blood cell sickling, making the combination of disorders more pathogenic than either alone.[1] To this point, Nawaiseh et al. found an increased risk of PSCR in patients with higher white blood cell counts where Nia et al. found no increased risk of retinopathy in otherwise healthy SCT patients.[2,3]

3. Conclusion

Once thought of as benign, there is mounting evidence to suggest that SCT can give rise to severe PSCR in the setting of concomitant systemic disease. It may be prudent to evaluate patients with SCT yearly with a dilated fundus examination to evaluate for signs of retinopathy, particularly in the setting of concomitant systemic disease. The presence of retinopathy should prompt initiation of a comprehensive medical evaluation to rule out the presence of underlying systemic disease, namely inflammatory, infectious and hyperglycemic disorders.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

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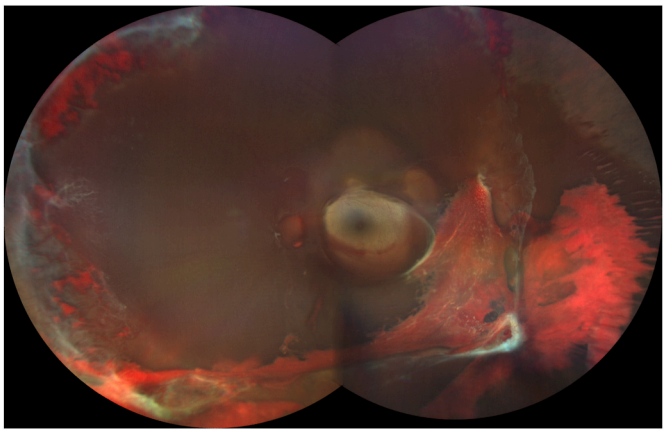

Fig. 1. Color fundus photo of the left eye demonstrating pre-retinal hemorrhage that spares the premacular bursa and mid-peripheral fibrovascular proliferation with macula-sparing tractional retinal detachment inferiorly and nasally. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

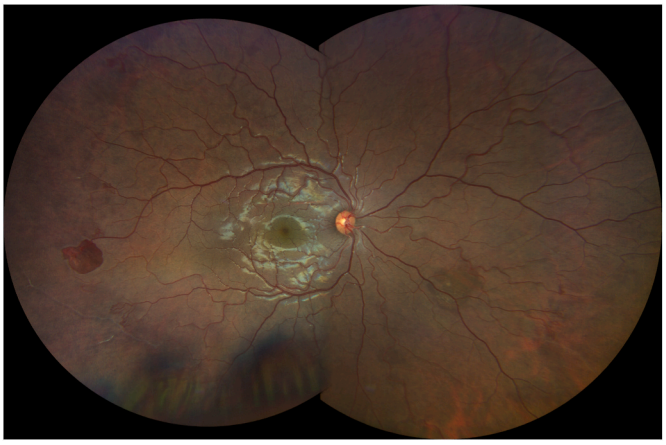

Fig. 2. Color fundus photo of the right eye demonstrating extensive peripheral ischemia with several areas of early retinal neovascularization without traction. There is a small peripheral preretinal hemorrhage temporally. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

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3. Nawaiseh M, Roto A, Nawaiseh Y, et al. Risk factors associated with sickle cell retinopathy: findings from the Cooperative Study of Sickle Cell Disease. *Int J Retina Vitreous*. 2022 Sep 22;8(1):68.