

Case Report

Bilateral Symmetrical Serous Retinal Detachment as a Complication of HELLP Syndrome

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Abstract

The Incidence of Serous Retinal Detachment (SRD) in HELLP Syndrome is rare. Till now, there are just few published case reports regarding this ocular finding. There are two hypotheses that try to explain its pathophysiology. The first hypothesis assumes that the SRD is as a consequence of choroidal ischemia with blood retinal barrier damage. The second assumes a disturbance of clotting cascade with consequent thrombotic genesis in the choroidal vasculature. A bilateral affection with a near mirror image distribution of serous retinal detachment between both eyes, that follows the anatomical distribution of choroidal vasculature, is first to be described in our case.

Case presentation: A 34 year old primigravid caucasian woman with non remarkable medical history experienced a sudden bilateral serous retinal detachment at the first postpartum day after caesarean delivery of twins. The general clinic picture matched to HELLP Syndrome. Visual acuity was at first postpartum day 0.4 in the right and 0.3 in the left eye. We observe a bilateral SRD in this patient with slight pigment epithelium thickening in both eyes. At the 7th and 8th day, an improvement of visual acuity was observed in correlation with a decline of SRD and RPE thickening.

Conclusions: To understand the pathophysiology and the correlation between HELLP-Syndrome and SRD is the Interdisciplinary work up essential. Studying more cases is required for understanding the pathophysiology of this rare syndrome.

Case Presentation

HELLP Syndrome (Hemolysis, Elevated Liver enzymes and Low Platelet count) is reported in 0.2% to 0.6% of all pregnancies [1] and is associated with significant maternal and fetal morbidity and mortality [2,3]. The exact frequency of ocular complications remains unknown as only few reports are available possibly because ocular findings may be undiagnosed while patients stay in the intensive care unit due to the potentially fatal nature of the disease.

Here, we report a 34 year old primigravid caucasian woman, who experienced sudden, painless bilateral blurring of vision at the first post-operative day following caesarean delivery of twins at 35+4 weeks of pregnancy. At first postpartum day, she developed a systemic clinical picture consistent with HELLP Syndrome. The first ophthalmologic examination was conducted in the intensive care unit on the third day postpartum. Her Best Corrected Visual Acuity (BCVA) was 0.4 and 0.2 in the right and left eye, respectively (decimal). Anterior segment examination was unremarkable. Fundus examination revealed bilateral symmetric Serous Retinal Detachment (SRD) affecting the posterior pole. Signs of hypertensive retinopathy, such as disc edema, cotton wool spots, retinal hemorrhages, arterial narrowing, arteriovenous crossing abnormalities (arteriovenous nicking), vascular wall changes

were not found. We recommended observation and no treatment was given regarding ophthalmological findings. At the 7th day postpartum, her BCVA had spontaneously improved to 1.0 and 0.8. A panoramic fundus photo then showed a bilateral SRD with a nearly symmetrical distribution in both eyes, centered superotemporal to the optic disc (Figure 1). Optical coherence tomography using Spectral is HRA-OCT device (Heidelberg Engineering, Heidelberg, Germany) showed slight Pigment Epithelium (RPE) thickening and SRD in both eyes with decline in the course of follow up examinations (Figure 2). Fluorescein angiography showed scattered areas of choroidal non-filling in the early phase, however no leakage in late phases was observed (Figure 3). Complete reattachment of the retina took place at 20th day postpartum with full visual recovery.

Discussion

HELLP Syndrome was first described in 1982 [2]. Seven years later Burke et al. [4] published the first case of bilateral SRD in HELLP-syndrome. So far, only few cases of serous retinal detachment in HELLP syndrome have been reported. SRD could be unilateral or bilateral, with variable clinical significance and severity.

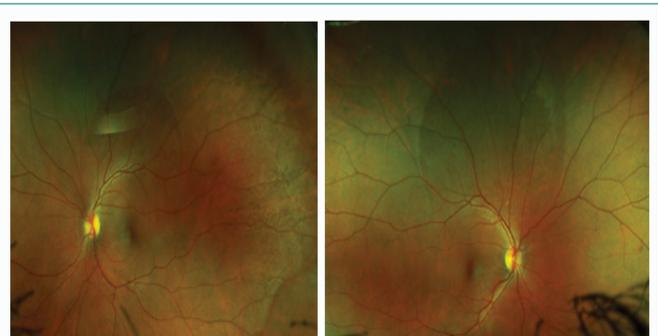


Figure 1: Note the nearly mirror image distribution of SRD superotemporal to the optic disc in both Right eye (on the right side) and left eye (on the left side).

Citation: Sakhnyny T, Gekeler F, Leschke S, Alnahrawy O. Bilateral Symmetrical Serous Retinal Detachment as a Complication of HELLP Syndrome. *Am J Clin Case Rep.* 2021;2(6):1041.

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Publisher Name: Medtext Publications LLC

Manuscript compiled: Aug 06th, 2021

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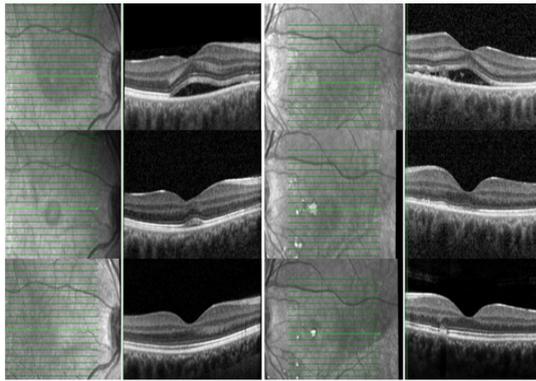


Figure 2: Shows OCT images of the right "A" and left "B" eyes on follow up visits at 7, 16 and 42 days post partum. The subretinal fluid disappears gradually and the retinal pigment epithelium thickening is getting slowly to its normal appearance on OCT.

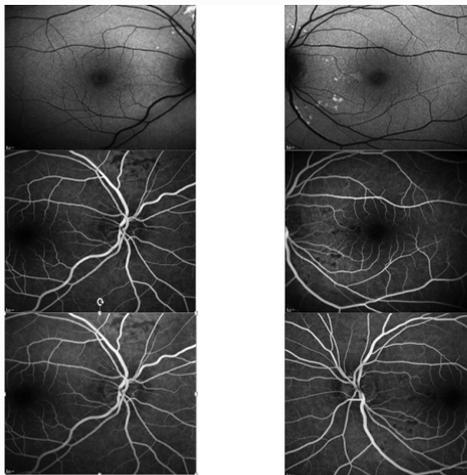


Figure 3: Fundus Autofluorescence (top) and Fluorescein Angiography (early and late phase): (A) right (B) left Eye: shows a peripapillary scattered areas of choroidal non-filling in the early phase (middle), however, no leakage in late phases.

While several theories trying to explain the pathophysiology of this ocular complication of HELLP, it remains not fully understood. Hayreh supposed a choroidal ischemia secondary to an intense arteriolar vasospasm. Vasoconstrictor agents like Angiotensin 2, leak freely from the choriocapillaris into choroidal interstitial fluid which cause vasoconstriction and ischemia with blood-retinal barrier damage. In consequence arise lesions in the RPE which cause the accumulation of sub-retinal fluid [5]. Cogan hypothesized a pathologic clotting mechanism where a consequent sequestration of platelets out of the circulation and thrombocytopenia result in a sudden deceleration and patchy occlusion of circulation between short posterior ciliary arteries and choroidal circulation [6].

To our knowledge, our patient's finding of bilateral symmetrical affection with a nearly symmetrical distribution of SRD has not been reported before. Symmetry could be a unique feature of this ocular manifestation of HELLP, where the localization of SRD follows a specific anatomical distribution of the affected vasculature and thus making the involvement of short ciliary arteries evident in the pathology. This partially supports Cogan's theory [5]. This ocular complication differs in general from the isolated hypertensive choroidopathy in cardiovascular patients that shows rather no SRD, while Elsching's spots and patchy choroidal filling defects in angiography are seen. The symmetry feature could also differentiate this disease from central Serous Chorioretinopathy (CSR).

Resolution of SRD depends on the pumping and metabolic function of RPE [4]. Our case showed rapid recovery of ophthalmological findings within 3 weeks excluding a pronounced RPE or choroidal damage.

Ophthalmologist should be aware of other ocular complications of pregnancy like cortical blindness, Purtscher-like retinopathy, central retinal vein occlusion, central serous Chorioretinopathy and the possibility of SRD as a differential diagnosis.

Currently the only effective treatment is prompt delivery. Regarding ocular findings, a close observation and reassurance were sufficient in our case.

Conclusions

To understand the pathophysiology and the correlation between HELLP-Syndrome and SRD is the Interdisciplinary work up essential. Studying more cases is required for understanding the pathophysiology of this rare syndrome.

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