Serous retinal detachment and choroidal thickening secondary to HELLP syndrome

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Background

HELLP syndrome is a disorder of pregnancy closely related to the hypertensive disorders pre-eclampsia and eclampsia. It is characterised by **H**aemolysis, **E**levated **L**iver enzymes and **L**ow **P**latelets.
Serous retinal detachment is a rare but well documented complication of HELLP syndrome/pre-eclampsia with an incidence of approximately 1% in this patient group.
Most cases reports of serous RD in HELLP/pre-eclampsia document resolution of subretinal fluid (SRF) and choroidal thickening 4-8 weeks post-partum, usually with a good visual prognosis.

This report describes a patient with HELLP syndrome presenting with bilateral serous RD and choroidal thickening. Serial OCT scans demonstrated the time course of resolution of choroidal thickening and SRF.

Case report

History:

A 26-year-old primiparous woman presented to the eye department 1 day post-partum with bilateral vision loss.
She had given birth via normal vaginal delivery the previous day. She had been taken to theatre for removal of retained placenta and on returning from theatre, report vision loss (left >right eye).
Her pregnancy had been unremarkable and she has no medical history of note.
Antenatally and peripartum, blood pressure was normal except for a single reading of 150/80 for which no treatment was given.
Blood tests on the day of delivery showed: Haemoglobin 73g/L
Platelets 46 x 10^9/L
Alanine transaminase 45 U/L
Albumin 22g/L.
On this basis a presumptive diagnosis of HELLP syndrome was made by her obstetric physicians.

On examination:

Visual acuity CF right CF left
Anterior segment NAD
No intraocular inflammation.
Fundoscopy: Large bilateral serous retinal detachments (Figure 1)

Diagnosis:

She was diagnosed with serous RD secondary to HELLP syndrome

Management:

Treatment of HELLP syndrome is supportive. The patient remained an inpatient under the obstetric team for one week before discharge and subsequent outpatient ophthalmology follow-up.

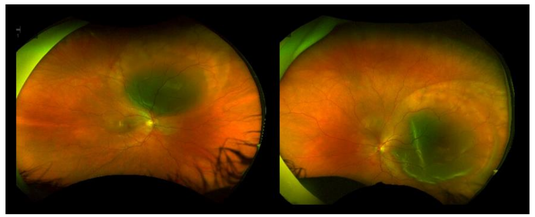


Figure 1 – fundus imaging day 1 post-partum showing bilateral serous RD

On day 2 post-partum, there was noted to have been a rapid and dramatic reduction in SRF in both eyes (Figure 2)

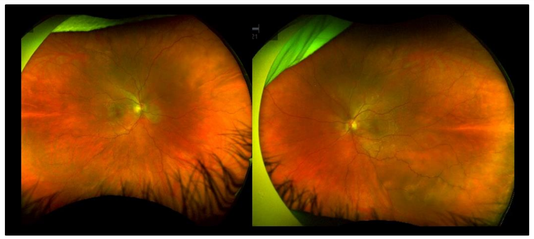


Figure 2 – fundus imaging day 2 post-partum rapid and dramatic reduction in SRF in both eyes

OCT on day 2 post-partum revealed bilateral choroidal thickening with sub-foveal choroidal thickness (SFCT) of 550 microns right and 560 microns left (Figure 3).

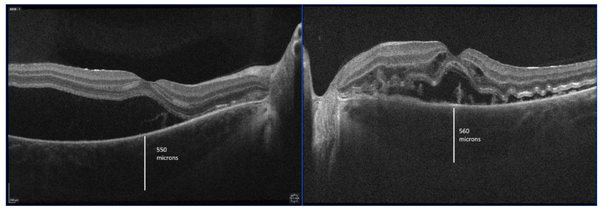


Figure 3 – OCT scans day 2 post-partum showing SRF and choroidal thickening.

Quantification of SRF

Manual segmentation of OCT scans using Zeiss FORUM controls allowed quantification of subretinal fluid over time [NB: day 1 post-partum is semi-quantitative as some areas of SRF were outside the area captured by the OCT scan

There was a rapid and dramatic reduction in SRF in the first 48 hours post-partum, followed by slower and complete resolution over the following 5 weeks. Choroidal thickness returned to within normal limits more slowly than the resolution of SRF (SFCT 270 microns right eye, 310 microns left eye at day 34 post-partum) (Figure 4).

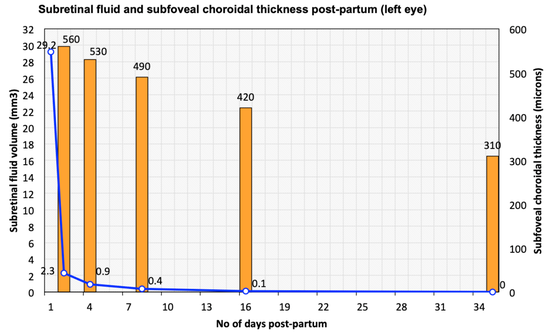


Figure 4 – choroidal thickness and SRF resolution over time

Visual acuity slowly returned to normal over 8 months of follow-up:

Post partum	Right VA	Left VA
Day 1	CF	CF
1 week	6/15	6/12
1 month	6/12	6/9
8 months	6/9	6/6

Discussion

One unresolved question in this case is why large amounts of SRF can be cleared rapidly while a much smaller amount takes a number of weeks to resolve.

We postulate the following hypothesis. This hypothesis is consistent with the observations made in this case and with what is currently understood from experimental models of RD:

HELLP syndrome leading to choroidal ischaemia and vascular damage

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Choriocapillaris hyperpermeability and RPE pump and barrier dysfunction
+ low albumin (low oncotic pressure)
+ increased choroidal hydrostatic pressure (valsalva manoeuvre in labour)

↓

Fluid ingress into subretinal space and RD formation

↓

Post-partum resolution of precipitating factors
(e.g. reduced hormonal effects on choroidal vasculature, rising serum albumin, no raised hydrostatic pressure from Valsalva manoeuvre)

↓

RPE barrier remains compromised in early post-partum phase – this would allow rapid, passive diffusion of SRF out of the subretinal space, as observed day 1- day 2.

↓

RPE barrier integrity restored early, slowing the rapid, passive egress of SRF

↓

Residual SRF to be removed by relatively slower active mechanisms.
(The RPE pump is powerful, capable of pumping the equivalent of the total ocular volume per day (2.4ml/day). In this case, SRF resolution between day 4 and day 34 was much slower than this rate suggesting that RPE pump function may be impaired for some time after ischaemic insult.)

Conclusion

- This case shows an uncommon cause of vision loss in the peri-partum period and highlights HELLP syndrome as a cause of serous detachment in this patient group.
- This is the first time that observations have been made on the time course of resolution of SRF and choroidal thickening in serous detachment secondary to HELLP syndrome.
- Such observations are useful in hypothesis-generation about the mechanisms of retinal detachment formation and resolution. Our hypothesis is speculative and further clinical and pre-clinical investigation is needed to understand the formation and resolution of serous RD in HELLP/pre-eclampsia.