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Case Report

Bilateral Exudative Retinal Detachment: A Complication of HELLP Syndrome

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Background:

The HELLP syndrome is a life threatening condition which is noticed primarily during pregnancy. The characteristics of this syndrome are - hypertension, haemolysis, an elevation in liver enzymes and low platelets count (HELLP)[1]. The HELLP syndrome is seen in around 10% of preeclampsia and eclampsia cases, leading to a high maternal (3%) and infantile (24%) mortality rate [2,3].

Bilateral, serous or exudative retinal detachment is not a very common complication of toxaemia of pregnancy [4]. In majority of the cases, the detachment is seen along with hypertensive retinopathy. We report a case of primigravida, who was diagnosed with eclampsia and was detected to have defective vision. Ophthalmological examination revealed bilateral bullous exudative detachment, and the clinical features were suggestive of HELLP syndrome.

Her hypertension was managed medically, and hemodialysis was carried out to correct the renal parameters. The patient's response to the treatment was positive, along with the re-absorption of the sub-retinal fluid, followed by complete visual recovery in both eyes.

Conclusion: Not many cases of HELLP syndrome have been reported to be associated with bilateral exudative retinal detachment in literature. Gynaecologists and Ophthalmologists all around the world should be sensitized about this ocular condition, especially in patients of eclampsia and pre eclampsia, and a thorough ophthalmological examination, including fundoscopy should be done in all patients of eclampsia.

Key words: Eclampsia; Exudative Retinal Detachment; HELLP Syndrome

Introduction

Hypertension detected during pregnancy, if not controlled properly can Cause serious cardiovascular and central nervous system complications. When a pregnant lady complains of blurring of vision,

she is supposed to be subjected to a thorough systemic evaluation, including blood pressure monitoring along with ocular examination, as some patients having elevated blood pressure, do exhibit features of hypertensive retinopathy and exudative retinal detachment. Cases of HELLP syndrome with exudative retinal detachment is an atypical occurrence, with only a few reports of the same. Hence, we report a case of HELLP syndrome with bilateral exudative retinal detachment.

Case Report

A 24 year, female, primigravida with 32 weeks of gestation, presented with a history of recurrent seizures, in the emergency department. On admission, her blood pressure recorded was 210/100 mm of Hg and remained high (180-200 systole and 100-110 diastole) during the first 5 days of hospital admission, despite being administere d with anti-hypertensive therapy. Systemic evaluation revealed bilateral pleural effusion, mild ascites and acute kidney injury. Blood samples of the patient showed the following results: Haemoglobin-9.5 g/l (normal 12- 14g/dl), creatinine-53.04 µmol/l (normal 45-90 µmol/l), potassium-4.7 mmol/l (normal 3.5-5.0 mmol/l), platelets 162× 109/l (normal 150-450 × 103/l), leukocytes 12.8 × 109/l (normal 4-13.5 × 109/l), ALT 33 U/l (normal 15-22 U/l), AST 32 U/l (normal for women 4-19 U/l) and LDH 873 U/l (normal 120-240 U/l); and an ultrasound revealed fetal distress. Emergency LSCS was done under general anesthesia and a male infant was delivered. The patient developed three episodes of seizure in post op recovery room, because of which she was transferred to the Intensive Care Unit. As a result of the recurrent seizures and unstable systemic condition, she was not extubated, and had to be placed on mechanical ventilation. Blood samples on day 1 postpartum showed a marked increase in the creatinine 273 µmol/l and potassium 6.0 mmol/l, suggestive of acute renal injury. Liver function tests showed raised enzymes with, ALT 106 U/l (normal 15-22 U/l), AST 296 U/l (normal for women 4-19 U/l), leukocytes 32 × 109/l, LDH 2175 U/l (normal 120-240 U/l) and a drastic decrease in platelets 84 × 103/l . Her hemoglobin was 6.4 g/l and an INR recorded was 1.4, indicating a longer blood coagulation time. Urine output for 24hrs was reduced to 350ml, and she was diagnosed to have acute renal failure, following which hemofiltration was started immediately. Over the next 5 days, her systemic condition improved, she regained consciousness and was removed from ventilator, with no recurrence of seizures. Because of her persisting poor urine output and deranged renal function tests, hemodialysis had to be continued for another 3 weeks.

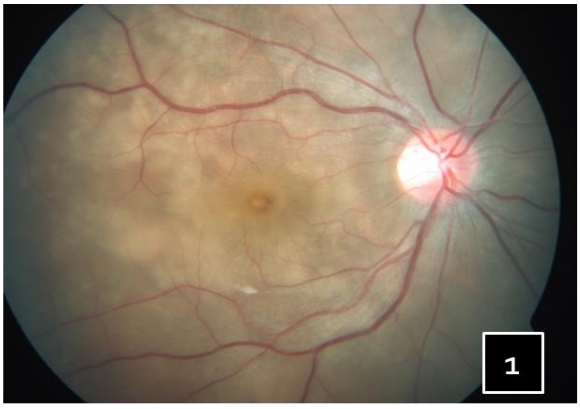
On the 7th postpartum day, the patient complained of gross diminution of vision, in both her eyes. Ocular examination revealed visual acuity in the right eye as hand motion close to face (HMCF), and in the left eye it was 5/200. Pupils were sluggishly reacting; however there was no afferent papillary defect. The anterior segment examination was unremarkable. Fundus evaluation revealed bilateral bullous exudative retinal detachment, with shifting fluid, involving the posterior pole, and inferior retina which was more on the right than the left side (Figure 1-4).

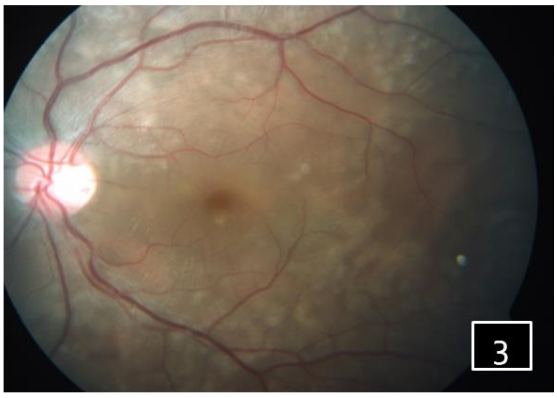
Before the occurrence of seizures, the patient was seen having good vision (20/20) in both her eyes, and there were no indications of hypertensive retinopathy as recorded in her anti -natal care follow up documents. The patient was managed conservatively with observation and head elevation, for her ocular condition and rest of the systemic conditions were managed by controlling blood pressure and hemodialysis. Visual acuity increased to 20/40 and 20/80 after 02weeks, and at the end of 03 months, fundus examination showed complete disappearance of sub retinal fluid, with yellowish white sub retinal opacities, and retinal pigment epithelial changes, which was confirmed by an optical coherence

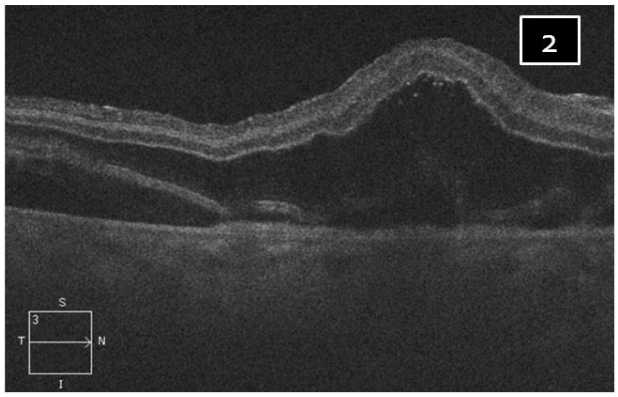
tomography . Bilateral visual acuity improved to 20/20 by 3 months, and the patient was doing good, and was followed up for a period of 12 months.

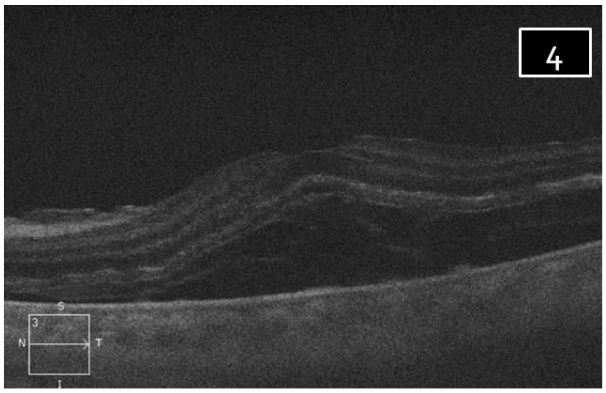
Figure (1 &3): Fundus picture showing exudative retinal detachment

Figure (2 &4): OCT macula showing the sub retinal fluid in the right and left eye respectively









Discussion

The HELLP syndrome is a life threatening condition which is seen during pregnancy characterized by hypertension, hemolysis, elevated liver enzymes and low platelets [1] . This affects the cardiovascular system, and can cause hematologic abnormalities, hepatic and renal impairment, CNS and ophthalmic manifestations.

The exact pathogenesis of serous retinal detachments in pre-eclampsia/ eclampsia is not well understood. One of the inferences is ischemiafF due to high blood pressure, leading to thrombosis and damage to the choriocapillaries. Exudative retinal detachment occurred secondary to choroidal ischemia in these patients. In hypertensive choroidopathy, there is leakage of endogenous vasoconstrictor agents from the choriocapillaries. These agents act on the walls of the choroidal vessels leading to vasoconstriction and later, ischemia. Retinal pigmented epithelium (RPE) which derives blood from choroid circulation is affected because of the ischemia. This causes the breakdown of the outer blood-retinal barrier and formation of a serous proteinaceous exudate from the choroid, which gets accumulated into the sub retinal space, producing serous retinal detachment. Fluorescein angiography and indocyanine green angiography support this hypothesis [5-7]

The other mechanism, which has been been postulated is that circulating placental

thromboplastic in the maternal circulation might initiate the extrinsic coagulation cascade which results in the activation of disseminated intravascular coagulation. This may be the reason for choroidal ischemia and subsequent serous retinal detachment [8]. There are four options available for the management of women with severe preeclampsia and HELLP syndrome [9-12] which include: 1) Immediate delivery (which is the primary choice) if the lady has completed more than 34 weeks of gestation. 2) Delivery within 48 hours, if the lady presents between 27 to 34 weeks of gestation, after stabilizing of the maternal clinical condition, along with supplemental corticosteroid therapy. 3) A conservative management for more than 48–72 hours in pregnant women before 27 weeks' gestation. 4) Termination of pregnancy should be strongly considered, if it is before 24 weeks of gestation [13].

Sometimes, HELLP syndrome can develop after child birth; usually within the first 48 hours [14, 15]. These women with post-partum HELLP syndrome, are at a higher risk of developing renal failure and pulmonary oedema as compared to those patients with an antenatal onset [16-19]. Postpartum HELLP syndrome requires management by a multi-specialty care. The standard care includes, twice a day administration of systemic corticosteroids (10 mg of dexamethasone), [20-23] plasma exchange with fresh frozen plasma [24-27] post-partum blood transfusion, as well as albumin supplementation.

Prophylaxis for DIC is done with ant thrombin or low-dose heparin administration [28].

Serous retinal detachment in toxemia of pregnancy can occur both ante partum and postpartum period leading to marked reduction in visual acuity[29-31]. Some people have tried giving systemic steroids to these patients, but without much effect.

In most cases, the detachment resolves spontaneously with a return to normal visual acuity within the period of a few days or weeks, in the postpartum period. However, some patients may develop residual retinal pigment called epithelial change, which can cause vision impairment.

Conclusion

Not many cases of HELLP syndrome presenting bilateral exudative retinal detach ment have been reported [29-31]. Hence, there is a pressing need to increase awareness regarding this disorder amongst Gynaecologists and Ophthalmologists. Whenever a patient of PIH or HELLP syndrome comes across a Gynecologist, an ophthalmological evaluation is advised. The ophthalmologist should know that exudative retinal detachment can develop in the cases which had previously recorded a normal fundus finding. Usually a favorable prognosis is seen in most of these patients.

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