

Posterior reversible encephalopathy associated with HELLP syndrome: A case report and review of the literature

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Abstract

We report a case of posterior reversible encephalopathy syndrome in association with hemolysis-elevated liver enzymes-low platelet counts (HELLP) syndrome. A 29-year-old primigravida presented at 35 weeks 4 days pregnancy with headache and blurred vision and the laboratory data revealed hemolysis, elevated liver enzymes, and low platelets. Cranial magnetic resonance imaging (MRI) showed hyperintense signal changes in the right occipital, left cerebellar regions and the right thalamus. Follow up brain MRI 10 days after the caesarean section showed resolution of the previous abnormalities. Posterior reversible encephalopathy syndrome (PRES) is a neuroradiologic condition associated with headache, seizures, altered sensorium, and visual disturbances. Although their association is rare, the diagnosis of PRES should be considered in all HELLP patients presenting with headache and neuro-ophthalmic signs. It is for the reason that the patients can recover well with complete reversal of clinical symptoms within several days, when adequate treatment is immediately initiated.

INTRODUCTION

HELLP (Hemolysis-elevated liver enzymes-low platelet counts) syndrome is a severe condition that occurs in the last trimester of pregnancy with severe preeclampsia, or immediately after delivery.¹ Hypertension or an immunopathological mechanism may lead to diffuse endothelial damage which in turn causes multi-organ dysfunctions, with possible brain involvement.² Posterior reversible encephalopathy (PRES) is characterized by symptoms such as headache, seizures, altered consciousness, hypertension and visual disturbances. It has also been described in cases of hypertensive encephalopathy, eclampsia, hypertension associated with renal insufficiency and also as a complication of therapy with immunosuppressant or antineoplastic medications.³ Although this condition was reported to be reversible, the cerebral injury may be permanent if the treatment is delayed.^{4,6} Thus, it is significant to recognize PRES in patients with HELLP syndrome to prevent persistent neurological complications. Here we describe a case of PRES in association with HELLP syndrome in a 35 week pregnant woman, and review of the literature.

CASE REPORT

A 29-year-old primigravida at 35 weeks 4 days by last menstrual period was admitted to our obstetrics department complaining of headache and blurred vision in the right eye for five days and in both eyes since that morning. She was followed by another maternity assessment unit until this week. The antenatal course was without any complications. Her sitting arterial blood pressure was 160/120 mmHg. Her abdominal ultrasonographic examination revealed a pregnancy of 33 weeks 1 day by appearance.

On her pelvic examination, cervix was found to be closed. She had pitting pedal edema up to the knee level. Urinalysis showed proteinuria 2+, her serum creatinine level was 1.4 mg/dl (Normal: 0.5-1.1), blood urine nitrogen level was 28 mg/dl (normal range, 9-23), uric acid was 9 mg/dl (Normal: 3,1-7,8), alanine transaminase (ALT) was 18 IU/L (Normal: 7-35) aspartate transaminase (AST) was 43 IU/L (Normal: 0-34), and lactate dehydrogenase (LDH) level was 800 IU/L (Normal: 120-240). Her complete blood count investigations showed a platelet count of $22 \times 10^3/\mu\text{L}$ (Normal: $150-400 \times 10^3$). Her hemoglobin and hematocrit levels were unremarkable.

On peripheral smear, schistocytes were seen indicating hemolytic anemia and platelet counts were approximately $80 \times 10^3 /\mu\text{L}$. Prothrombin time, partial thromboplastin time, and bleeding time were all normal. Lupus anticoagulant and anticardiolipin antibodies were not tested since the patient had no history of spontaneous recurrent abortions.

She was diagnosed with Class 1 HELLP syndrome and admitted to the eclampsia unit. On her follow-up, arterial blood pressure was measured as 220/110, and nifedipine was given. Magnesium sulphate treatment was started to prevent eclampsia. She was transfused with 4 units of platelet concentrates collected by platelet apheresis, and examined by neurology and ophthalmology specialists. On fundoscopic examination, bilateral macular edema was detected without papilledema. Pupils were equal including bilateral normal pupillary light reflexes. Her visual acuity was 0.6 and 0.8 in the right and left eye respectively.

On neurological examination, her Glasgow coma score was 15/15 with no evidence of neurological deficit. There was no meningism. Deep tendon reflexes were normal. The cranial magnetic resonance imaging (MRI) showed hyperintense signal changes in the right occipital, left cerebellar subcortical regions and right thalamus on T2-weighted and FLAIR images (Figure 1a and Figure 2a). Diffusion weighted MR images showed no restriction of diffusion.

An immediate cesarean section was performed. She was delivered of a male infant weighing 1930 gr. with APGAR scores of 6 and 7 in the first and fifth minutes, respectively. The baby was transferred to neonatal intensive care unit. The patient received dexamethasone (8mg, bid) treatment postpartum for 3 days. Postpartum arterial blood pressure was 160/110 mmHg. She received alpha-methyl dopa orally (250mg, qid) to stabilize her arterial blood pressure. Her serum creatinine, blood urine nitrogen and uric acid levels returned to normal, while ALT, AST and LDH levels increased to 84 IU / L, 87 IU / L and 1371 IU / L respectively on the second day postoperatively. Ecchymosis was observed around the incision area on the third postpartum day. She was discharged 5 days after the delivery with normal blood pressure and mildly blurred vision. On the tenth follow-up day, vision impairment was completely resolved and the biochemical profile was normal. MRI scan of the brain showed complete resolution of the previous lesions 20 days after the operation (Figure 1b and Figure

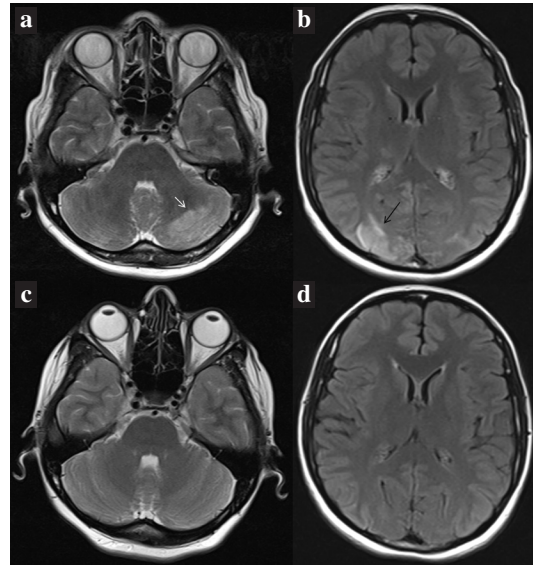


Figure 1. Axial T₂-weighted MRI showing hyperintensity on the left cerebellar hemisphere before the delivery (a, white arrow). The lesion disappeared 20 days after the cesarean section (c). On axial flair MR images, hyperintense lesions are seen on the right occipital region and right thalamus (b, black arrow). They resolved 20 days after the operation (d).

2b), consistent with the diagnosis of posterior reversible encephalopathy syndrome.

DISCUSSION

HELLP syndrome represents a complication of pre-eclampsia. It occurs in 0.5%–0.9% of all pregnancies.⁷⁻⁹ Delayed diagnosis or inappropriate treatment contributes to its poor prognosis increasing the maternal and perinatal death rates.¹⁰ HELLP is divided into three groups according to platelet counts: Class 1 (<50 000 platelets/ μl), Class 2 (more than 50 000 but less than 100 000 platelets/ μl), and Class 3 (> 100 000 platelets/ μl).² PRES is rarely associated with HELLP syndrome.³ A search in the English literature revealed only six cases reporting this association.^{2,11-15} Five cases were defined as Class 2 HELLP and one case was Class 1 HELLP as it is shown on Table 1.

MRI is the primary modality in diagnosis of PRES. Typical MRI findings of PRES are symmetrical hyperintense lesions in both T2-weighted and FLAIR images on the posterior regions of cerebral hemispheres, basal ganglia and brain stem.⁴ Diffusion-weighted imaging (DWI) is helpful in differentiating PRES from other disorders. Lesions of PRES display no restriction

Table 1: MRI findings and outcomes of patients reported in the literature

	Age	Gestational age	Symptoms and signs	Laboratory findings	Cranial MRI T2 and Flair	Diffusion	Outcomes
Marano et al.²	30	32 week	GTC seizure, anuria, drowsiness	Class 2 HELLP, thrombocytopenia, hypertransaminasemia,	Bilateral asymmetric hyperintensities in parieto-occipital white matter	Not mentioned	Complete recovery
Negro et al.¹³	37	39 week	Seizure, stupor, hypertension, visual loss	Class 2 HELLP, thrombocytopenia, Hypertransaminasemia, increased LDH, proteinuria (3+)	Diffuse cortical-subcortical hyperintense lesions in both hemispheres and thalamus, cerebellum, mesencephalon	Not mentioned	Complete recovery
Morton et al.¹²	26	28 week	GTC seizure, clonus, visual disturbance, proteinuria, epigastric pain, hypertension	Class 2 HELLP, thrombocytopenia, hypertransaminasemia, proteinuria (1+)	No MRI (CT scan showed hypodense areas on right frontal and both occipital		Complete recovery
Peng et al.¹⁴	36	38 week	Seizure, LOC, general edema,	Class 2 HELLP, thrombocytopenia, hypertransaminasemia, increased LDH	Hyperintense lesions in the right frontal lobe and bilateral basal ganglia	Not mentioned	Complete recovery
Grzesiuk et al.¹⁵	34	32 week	GTC seizure, hypertension	Class 2 HELLP, thrombocytopenia, hypertransaminasemia, increased LDH, hyperbilirubinemia	Hyperintensities in occipital, parietal and frontal lobes	No restriction	Full neurologic recovery, small areas of edema on MRI
Vijaya-lakshmi et al.¹⁶	28	16 week triploid pregnancy	Headache, visual disturbance, morning sickness, hematuria, epistaxis	Class 1 HELLP, thrombocytopenia, proteinuria (4+), hyperbilirubinemia, increased uric acid, increased creatinine, hypertransaminasemia	Hyperintensities in the head of caudate nucleus, bilateral external capsules and basal ganglia	Minor diffusion restriction in right paraventricular area	Complete recovery
Ates et al.	29	35 week	Headache, blurred vision, hypertension, proteinuria	Class 1 HELLP, thrombocytopenia, proteinuria (2+), increased LDH, increased uric acid, increased creatinine,	Hyperintense signals in right occipital lobe, right thalamus and left cerebellum	No diffusion restriction	Complete recovery

GTC, generalized tonic clonic; HELLP, hemolysis-elevated liver enzymes-low platelet counts; LDH, lactate dehydrogenase; MRI, magnetic resonance imaging; CT, computed tomography; LOC, loss of consciousness.

of diffusion on DWI proposing the nature of the edema as vasogenic.¹⁵ This vasogenic edema is explained in reports by intense endothelial damage associated with hemodynamic change in HELLP syndrome.¹⁴ Complete resolution of these radiological findings was observed within 14 days by many authors in the literature.^{16,17} We observed a similar time course for their resolution and noticed that clinical recovery precedes normalization of MRI findings. The standard treatment of neurological findings in PRES is aggressive blood pressure control and management of underlying pathology.¹⁵ It was recently shown that patients with preeclampsia have a less severe course and better response to treatment than other contributing diseases of PRES.¹⁸

In conclusion, we report a case of HELLP syndrome accompanied by PRES which was resolved completely after prompt management of hypertension and pre-eclampsia. Since this clinical entity is almost always reversible, obstetricians should be alert for patients with HELLP accompanied by neurological and visual symptoms.

DISCLOSURE

Conflict of interest: None

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