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Case Report

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The Unusual Presentation of HELLP Syndrome

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Abstract

HELLP syndrome is a serious pregnancy-related syndrome characterised by hemolysis, elevated liver enzymes and low platelet count occurring in 0.5-0.9% of all pregnancies and in 10-20% of cases with severe preeclampsia. Typical presenting symptoms are right upper quadrant or epigastric pain, nausea and vomiting. Seventy percent of the cases develop antepartum, majority between the 27th and 37th gestational weeks. Thirty percent of the cases are diagnosed postpartum, often within 48 h post-delivery. The occurrence of preeclampsia post-delivery is well established. However, in most reported cases, HELLP syndrome persisted since late pregnancy. We report a case of HELLP syndrome in combination with preeclampsia that developed 2 days post-delivery in an uncomplicated pregnancy.

Keywords: HELLP syndrome; Preeclampsia; Postpartum

Introduction

The HELLP syndrome is a serious pregnancy-related syndrome characterised by the presence of hemolysis, elevated liver enzymes and low platelet count in combination with preeclampsia/eclampsia [1]. It is currently regarded as a variant of severe preeclampsia or a complication. It occurs in about 0.5-0.9% of all pregnancies and in 10-20% of cases with severe preeclampsia [2]. Seventy percent of the cases develop antepartum, majority between the 27th and 37th gestational weeks. In 30% of the cases, it is diagnosed postpartum, often within 48 h after delivery [3].

The pathogenesis of the HELLP syndrome remains unknown. Some theories proposed include complement activation with anaphylatoxin release, nitric oxide, genetic and hydralazine-induced hepatocellular damage [4].

Typical presenting symptoms are right upper quadrant abdominal pain or epigastric pain, nausea and vomiting. The characteristic of the upper abdominal pain may be fluctuating and colic-like [3]. Diagnosis of HELLP syndromes have often been based on different criteria and this condition can be diagnosed based on biochemical evidence. Two commonly used classifications for HELLP syndrome are the Tennessee System classification and Mississippi classification [5].

The HELLP syndrome is associated with both maternal and neonatal complications. It is associated with serious maternal morbidity, especially when it arises in the postpartum period. Severe maternal complications are cerebral hemorrhage, disseminated intravascular coagulation and subsequent severe postpartum bleeding [5]. Women with postpartum HELLP syndrome, have a significantly increased risk of renal failure and pulmonary edema compared to those with antenatal onset [6].

We report a woman with HELLP syndrome, who developed 2 days post-delivery in an uncomplicated pregnancy.

Case Report

A 25-year-old woman, gravida 3 parity 3, had a spontaneous vaginal delivery and postpartum hemorrhage at home. The patient's medical history was completely uneventful and regular antenatal visits had shown no complications such as preeclampsia, eclampsia or hypertension. Her previous two pregnancies were uncomplicated, and modes of delivery were all by spontaneous vaginal delivery. The newborn's perinatal period was uneventful. Intramuscular vitamin K was given after delivery.

She was admitted for primary postpartum hemorrhage with estimated blood loss of 1.5 L. Examination under anesthesia and manual removal of placenta was performed with patient's consent, and she received two units of packed red cells. She was stable postoperatively and was remained as an inpatient for observations.

Approximately 30 h post-delivery, she experienced a sudden onset of headache, nausea and was feeling unwell. This was associated with hypertension (blood pressure (BP) 163/109 mm Hg). Other vital signs such as heart rate, respiratory rate and oxygen saturation were within normal limits. She had backache and epigastric tenderness. She denied of any visual disturbances.

On examination, her abdomen was soft and non-tender. Uterus was at umbilicus. She had a normal upper and lower limb neurological examination and cranial nerve examination with no clonus.

Antihypertensive therapy with oral nifedipine 10 mg was initiated, and her BP decreased to 139/93 mm Hg. Oral paracetamol 1 g was prescribed. She was on hourly monitoring

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for BP. Urine dip stick showed protein 3+ hence urine protein/creatinine ratio was taken. Bloods were taken and laboratory results revealed that her platelet count dropped sharply from 152 to 72 × 10^9/L, and total bilirubin increased from 9 to 41 µmol/L. She had raised serum alanine aminotransferase of 534 U/L, raised serum alkaline phosphatase of 319 U/L and raised serum lipase 31 U/L. She did not have any pre-pregnancy liver function tests to compare the results with.

The obstetric team diagnosed her as having postnatal severe preeclampsia with impending eclampsia. Taking a multidisciplinary team approach, she had a computed tomography head scan which ruled out any incidental intracranial tumors, cerebral hemorrhage, cavernous sinus thrombosis and post-dural puncture headache amidst the preeclampsia. She also had abdominal ultrasound scan of liver with portal vein Dopplers, which ruled out acute fatty liver of pregnancy and pancreatitis.

The patient was started on an intravenous magnesium sulphate regime. Her BP remained severely high hence requiring intravenous hydralazine 5 mg to further bring her BP down. Her pain worsened and she subsequently vomited, thus intravenous paracetamol 1 g and intravenous ranitidine 50 mg were administered.

Her BP normalized under continued antihypertensive therapy with oral nifedipine 10 mg twice daily and subsequently oral amlodipine 10 mg once daily. Laboratory results returned to their normal range during the following days. On discharge, patient was completely recovered. She will have regular BP monitoring by the midwife and repeat blood test during a medical review by the general practitioner in a week before attending a postnatal review by a specialist in 6 weeks.

Discussion

This case report suggests the postpartum development of a HELLP syndrome in combination with preeclampsia in a normal pregnancy. The occurrence of preeclampsia with hemolysis, elevated liver enzymes and low platelet count after delivery is well established. However, in most reported cases, HELLP syndrome persisted since late pregnancy [1].

Our case emphasises on the importance of active diagnostic investigations if preeclampsia should develop in the postpartum periods. Liver enzymes and platelet count which are conclusive for HELLP syndrome should be considered.

The clinical presentations may vary between patients, and the leading symptom which is pain in the upper right abdominal quadrant may even be absent as illustrated by a case report [1]. This may cause a delay in the diagnosis leading to severe complications. Thus, it is also crucial to diligently monitor the patient's BP regularly in the postpartum period.

Magnesium sulphate therapy should be rapidly initiated as it halves the risk of eclampsia, and probably reduces the risk of maternal death [7]. A review comparing the use of magnesium sulphate and other anticonvulsants such as phenytoin or nimodipine for women with preeclampsia reported that magnesium sulphate remains the drug of choice for prevention of eclampsia. It has also been shown that there is no clear evidence of this benefit having an effect on long-term outcome for either women or children [8]. From an economic perspective, magnesium sulphate is an inexpensive drug and is also appropriate for low-income countries.

Conclusions

Our case highlights the importance of active diagnostic investigations with consideration of liver enzymes and platelet count which are conclusive for HELLP syndrome if postpartum preeclampsia should develop. The HELLP syndrome is a serious condition with a wide range of complications associated with serious maternal morbidity, especially when it arises in the postpartum period. It is an uncommon complication of pregnancy and majority are diagnosed in the antepartum period.

Clinical presentations may vary between patients, hence it is crucial to diligently monitor a patient's BP regularly in the antepartum period and carry out active investigations and rapid initiation of therapy when postpartum HELLP syndrome is suspected.

Magnesium sulphate is the drug of choice for prevention and treatment of eclampsia, and is also highly recommended by the Royal College of Obstetricians and Gynaecologists [9].

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Conflict of Interest

None to declare.

Informed Consent

Not applicable.

Author Contributions

All authors contributed to the study conception and design. Material preparation, data collection and analysis were performed by RL. First draft of the manuscript was written by RL and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

References

1. Yilmazturk A, Schluter W. Postpartum HELLP syndrome. Eur J Obstet Gynecol Reprod Biol. 1992;43(3):243-244.

2. Kirkpatrick CA. The HELLP syndrome. Acta Clin Belg. 2010;65(2):91-97.
3. Haram K, Svendsen E, Abildgaard U. The HELLP syndrome: clinical issues and management. A Review. BMC Pregnancy Childbirth. 2009;9:8.
4. Geary M. The HELLP syndrome. Br J Obstet Gynaecol. 1997;104(8):887-891.
5. Mihu D, Costin N, Mihu CM, Seicean A, Ciortea R. HELLP syndrome - a multisystemic disorder. J Gastrointestin Liver Dis. 2007;16(4):419-424.
6. Sibai BM, Ramadan MK. Acute renal failure in pregnancies complicated by hemolysis, elevated liver enzymes, and low platelets. Am J Obstet Gynecol. 1993;168(6 Pt 1):1682-1687; discussion 1687-1690.
7. Altman D, Carroli G, Duley L, Farrell B, Moodley J, Neilson J, Smith D, et al. Do women with pre-eclampsia, and their babies, benefit from magnesium sulphate? The Magpie Trial: a randomised placebo-controlled trial. Lancet. 2002;359(9321):1877-1890.
8. Duley L, Gulmezoglu AM, Henderson-Smart DJ, Chou D. Magnesium sulphate and other anticonvulsants for women with pre-eclampsia. Cochrane Database Syst Rev. 2010;11:CD000025.
9. Hypertension in pregnancy: diagnosis and management: Guidance [Internet]. NICE. [cited 2019Aug18]. Available from: https://www.nice.org.uk/guidance/cg107/chapter/1-Guidance#management-of-pregnancy-with-pre-eclampsia.