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HELLP Syndrome: A Rare but Critical Obstetric Conundrum

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Authors' contributions

This work was carried out in collaboration among all authors. Authors MA and SSMNV gathered the case from emergency ward and author MA arranged the theoretical and the presentation review of the case report. Author SSMNV aided in reviewing the literature part and authors PKY and RLG chipped away at the case show alongside the remaining writers. Authors TSLT and NP aided in drafting the presentation, discussion, and summarized the conclusion part of the case report. Author BAC alongside author SSMNV dealt with the writing searches and other authors in the arrangement of the manuscript. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Background: HELLP Syndrome is one of the significant difficulties of pregnancy and the acronym represents H=Hemolysis, EL=Elevated Liver Enzymes, LP=Low Platelets. It is a significant and hazardous type of toxemia, which is a condition where a pregnant lady has hypertension that harms the Liver and Kidney. It typically develops between the 26th to 40th long stretches of Fetal

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Development, and at times in the week after the child is conceived. Eclampsia is the most extreme type of toxemia joined by seizures. The rate is 0.5-0.9% in most pregnant ladies, however, in extreme Toxemia, 10-20% of cases have been accounted for.

Case Subtleties: A 29-year-old Multigravida lady who was pregnant for the third time, at 35 weeks of pregnancy was presented with the objections of serious stomach torment for one day, and fetal development was not evaluated. She had a known instance of hypertension during her past pregnancy and was dealt with apparently with Labetalol, furthermore gone through a lower Cesarean segment two years earlier. She was given Magnesium Sulfate 4g IV over 5 Minutes.

Discussion: Blood products like platelets, red blood cells, and fresh frozen plasma ought to be transferred. On account of early pregnancy, corticosteroids can be utilized to invigorate the development of fetal lungs. Basic follow-up is expected for patients with HELLP Syndrome.

Conclusion: Early location and treatment of HELLP syndrome, either by inception or by typical work or by cesarean area is valuable for both mother and embryo and forestalls further confusions.

Keywords: HELLP; hypertension; serum glutamic-oxaloacetic transaminase; serum glutamic pyruvic transaminase; alkaline phosphatase; hemoglobin.

1. INTRODUCTION

In pregnant women, HELLP syndrome is one of the major complications and is characterized by a triad of red blood cell breakdown (hemolysis), elevated liver enzymes, and thrombocytopenia (low platelet count). In HELLP syndrome, the incidence is 0.5–0.9 % in most pregnant women, but in severe preeclampsia, only 10–20% of cases have been reported [1,2]. HELLP was previously named Edema-proteinuria-gestational hypertension type B and it was later changed to HELLP (H=Haemolysis, EL=Elevated Liver enzymes, LP=Low Platelets) syndrome. This is an acronym used to describe the condition of patients with preeclampsia or eclampsia occurring in 5-10% of pregnancies. It is best described as a pregnancy-specific syndrome that can affect any organ system. 15-20% of women with HELLP syndrome do not have preeclampsia but 10-20% of women with severe preeclampsia develop HELLP syndrome.

It can lead to various maternal complications such as eclampsia, stroke, placental abruption, pulmonary edema, acute renal failure, microangiopathic hemolytic Anemia, and HELLP syndrome [3]. Weinstein considered this as a unique variant of preeclampsia and coined the term HELLP syndrome for this entity.

In HELLP syndrome, cell death, and periportal bleeding are the most common histopathologic findings, and several studies have also shown that levels of laboratory findings such as liver enzymes and platelet counts are not related to liver histopathology [4]. Various genetic variants that play an important role in the increased risk of HELLP syndrome have been reported but found to have no role in clinical management. It is hypothesized that the complement cascade is a key mediator in systemic inflammatory disorders such as severe preeclampsia/HELLP and it has been observed that women with mutations in complement regulatory protein appear to be at increased high risk of severe preeclampsia and HELLP syndrome. Long-chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD) deficiency in the fetus appears to be associated with an underlying cause. However, in contrast to preeclampsia, nulliparity is not a risk factor for HELLP syndrome, as most of the affected patients are multiparous [5,6].

The main clinical symptoms of HELLP syndrome are right upper quadrant pain or epigastric pain due to certain pathologies such as gallstones, cholecystitis and liver disease, pancreatitis, hernia, and peptic ulcer. In rare cases, pregnancy symptoms such as fatty liver, and hepatic venous occlusion, with or without severe hypoglycemia, may also be considered. In half of the women, viral flu-like symptoms are most common within a few days, and nausea and vomiting are common symptoms in all pregnant women. The symptoms usually continuously progress and their intensity often changes spontaneously. Physicians and surgeons should be aware of the diagnosis of HELLP syndrome because the primary pattern of vague symptoms can be seen during the first 3 to 22 days of pregnancy. In most cases, almost 15% of pregnant women are likely to develop HELLP syndrome in the second trimester [7].

Two major guidelines, the Tennessee Classification, and the Mississippi Triple Class System, are currently used for the diagnosis of

HELLP syndrome based on Hemolysis, elevated liver enzymes, and low platelet count. According to the Tennessee Classification System, elevated liver enzymes, such as AST, can be identified by laboratory findings ≥70 IU/L and platelets ≤ 100,000 cells/mm3. If these parameters were not observed or absent in the patient, this condition was diagnosed as partial HELLP syndrome.

Martin proposed the Mississippi HELLP classification system in which he classified it into three classes based on the lowest perinatal platelet count. Class 1 and class 2 are associated with Hemolysis (LDH≥ 600 IU/L) and elevated AST or ALT (≥ 70 IU/L) levels. Platelet counts are ≤ 50,000 cells/mm3, for class 1, and ≤ 100,000 cells/mm3 for class 2, while class 3 requires LDH ≥ 600 IU/L, AST or ALT ≥ 40 IU/L, and platelet count >100,000 or ≤ 150,000/ µL. This class 3 of HELLP syndrome is considered a clinically significant transition stage or a phase that can progress [8,9]. Diagnosis of the complete form of HELLP syndrome requires the presence of all 3 major components, while partial or incomplete HELLP syndrome consists of only 1 or 2 elements of the triad (H EL or LP) [10].

`After diagnosis, a decision must be made about childbirth. Due to the progressive nature of the disease, these patients require constant hospitalization and careful management of labor and delivery. It is of the utmost importance to assess the maternal condition through medical history, laboratory tests, and blood test parameters [11]. High blood pressure should be managed and the need for magnesium sulfate injections and blood and blood product transfusions should be determined. Patients with HELLP syndrome have increased morbidity and mortality [12]. Attention must be paid to the development of multiple organ system disorders. It gradually affects various systems and can cause Eclampsia, acute renal failure, DIC, severe Anemia, liver rupture, and intracerebral hemorrhage. Intravascular volume, fluid, and electrolyte balance must be maintained. High-dose corticosteroids have been shown to accelerate improvements in laboratory parameters, thereby reducing maternal hospitalization and adverse complications.

Fetal monitoring is performed by ultrasound, non-stress testing, and Doppler studies. Corticosteroids are used for fetal lung maturation in cases of premature birth. Maternal and fetal status, gestational age, Bishop's index, and cervical sensitivity are some of the factors that help determine the mode of delivery. Due to continued pregnancy in HELLP syndrome, there is a risk to the mother as well as to the fetus at the time of delivery, resulting in preterm delivery [13]. Preventing disease recurrence, screening, and identifying high-risk pregnant women is considered a major issue for management.

Early diagnosis and identification of complications of HELLP syndrome and prompt intervention are the main treatment strategies. Patients with HELLP syndrome are at risk for anemia and thrombocytopenia. Therefore, blood components such as packed red blood cells, fresh frozen plasma, and platelets are administered to prevent further complications during pregnancy. For thrombocytopenia, platelet transfusion therapy is given when the platelet count is well below the normal value of 50,000/mL. In the case of surgery, platelet transfusions are given for prophylaxis.

The main complication of HELLP syndrome, occurring in 1% of pregnancies, is subcapsular hematoma, which can lead to maternal death. Diagnostics can be done with computed tomography, magnetic resonance imaging, or ultrasound to identify and monitor this complication. Hepatic hematoma is one of the emergencies requiring surgery. Evacuation and drainage of the hematoma, stitching of the laceration, and bandaging are most often required [14-16].

2. PRESENTATION OF THE CASE

Here, we present the instance of a 29-year-old multigravida at 35 weeks gestation with a main grumbling of unexpected windedness. The patient additionally grumbled an extraordinary, sharp, wounding mid-sternal chest torment for 1 day, and on checking, the power is 7/10 and emanating to the back. The patient showed that the aggravation increased with profound breathing or while lying level on her back/stomach. Non-stress test (NST) was finished for the patient and it was observed that there was an extremely low fetal pulse. The subject gave a circulatory pressure of 121/74 mmHg, a heartbeat of 86 bpm, and a respiratory pace of 23 cycles per minute. Actual assessment was relevant with reproducible mid-sternal chest wall delicacy. Different Laboratory findings included Hemoglobin of 8.2 g/dL, Hematocrit of 32%, platelet count of 82, 000/mm3, Serum Creatinine of 1 mg/dL, Aspartate aminotransferase of 125 IU/L, Alanine

transaminase of 187 IU/L, and Total bilirubin of 1.7 mg/dL. Hence, Urinalysis was requested and noted to be negative for protein and Glucose. A Chest CT check showed diminished constriction in the distal parts of both right and left lower curve pneumonic veins, dubious of two-sided pneumonic emboli. The patient likewise had two-sided pedal edema which was treated with diuretics and Inj. ceftriaxone 1gm (1-0-1) was given for the prophylaxis. Inj. Ranitidine 50mg (1-0-1) was given to forestall acidity, Inj. Dexamethasone 2cc was given to expand the platelet count in the patient in light of the fact that, upon the arrival of affirmation, the platelet count was exceptionally low (0.82 lakhs). 4 pints of Platelets were transfused for the patient, and 4 pints of Fresh frozen plasma bonding was additionally done in light of the fact that the patient's HB level was additionally extremely low like 8.2 g/dl. A Fetal ultrasound uncovered an expected fetal load of tenth percentile and a biophysical profile of 8. The perinatologist suggested corticosteroids for fetal lung development and two times week-by-week follow-up. The conclusion as of then was pneumonic embolism. The Subject was at first anticoagulated with intravenous heparin (Usual Therapeutic Dose) and was thusly kept up with on low sub-atomic weight heparin at 10,000 units like clockwork. The next day, the patient's platelet count in this way dropped to 1, 00,000/mm3. This was ascribed to heparin use. Platelet count hence expanded to 1, 39, 000 mm3. The patient was discharged home 3 days after the fact on low sub-atomic weight heparin at 10,000 units like clockwork. The patient got back to the trauma center 7 days after the fact with a central grievance of intense epigastric agony emanating to the back. The patient showed that the aggravation started 4 hours earlier and with moderate to serious force of 8/10. The patient had a circulatory strain of 141/78 mmHg, a heartbeat of 71 bpm, and a respiratory pace of 26 cycles per minute, with delicacy at the right upper quadrant without rebound. The patient's lab discoveries were as per the following: hematocrit 33%, platelet count 82, 000/mm3, AST in the degrees of 174 IU/L, and ALT 198 IU/L. Low sub-atomic weight heparin administration was ended after 3 days. Following 8 hours, the patient's platelet count dropped to 39, 000/mm3 and hematocrit was 29%. Because of deteriorating maternal status, a developing position of a mediocre vena cava channel and a cesarean segment were performed. A reasonable male newborn child was conveyed with an Apgar score of 6 and 7 at 1 and 5 minutes and a birth weight of 2150 g. Intraoperative discoveries uncovered an enormous hemoperitoneum and a diffusely blocked liver with 2 capsular hematomas covering areas of unconstrained break [17-19]. Hemostasis was accomplished by coagulating the exposed liver surfaces utilizing an argon beam. The postoperative determination was HELLP Syndrome. On the following day of the conveyance, 1 16 ounces of blood bonding was finished, CBC was requested after blood transfusion and her examination uncovered hemoglobin values were 9.9 g/dL, platelet count was 11, 450/mm3, SGOT 149 U/L, SGPT 191 U/L, and Alkaline Phosphatase 124 U/L. On the second day of conveyance, the patient was griped of migraine and retching, and an Infusion of MgSO4 to forestall seizures was given. Tablet Meftal spas were given for relief from discomfort. B protein powder was managed as a dietary enhancement and Inj. Vitcofol 2cc was regulated each and every day to treat the sickly condition. Magnesium Sulfate was given for seizure prophylaxis at 4g IV for 5 Minutes. The patient was bonded with multiple donor red platelets and other blood items. The patient was alluded to conceivable liver transplantation. A subsequent stomach CT examination showed a subcapsular hematoma without a perceptible expansion in size. The patient was discharged following 9 days in stable condition. Treatment was continued for 6 days. CBC was performed before the day of discharge, and tests uncovered that her hemoglobin level was 11.3 g/dL. The patient was discharged on the ninth day after conveyance. For lack of calcium, she was given medications like Tablet Calcilace (0-1-0) and for sickliness, she was given medications like Cap Autrine (1-0-1). Upon the arrival of the discharge day, her pulse was typical. Her side symptoms were eased and the patient's condition was gotten to the next level. After release, 30 days follow-up was given. On follow-up visits with her obstetrician, the patient was accounted for to show ordinary actual discoveries.

3. DISCUSSION

Until this point in time, there is significant variety in the definition and wording of the HELLP condition. The indicative measures utilized are variable as well as conflicting. Hemolysis, characterized as the presence of microangiopathic hemolytic Frailty is the sign of HELLP condition. The normal lab discoveries incorporate microangiopathic Hemolysis, unusual peripheral smear, raised serum bilirubin

(roundabout), and a huge drop in Hemoglobin levels. The second part of the HELLP condition set of three is raised liver enzymes. Raised liver proteins imply unusual serum levels of AST, ALT, and bilirubin values. Nonetheless, the obvious cut-off levels were not explicitly characterized. In ensuing clinical examinations, unusual AST and ALT levels were accounted for to be 17 to 72 IU/L, while strange platelet counts went from 75,000 to 150,000 mm3 [20,21].

Our case report endeavors to show the demonstrative problem that a clinician faces in diagnosing an abnormal show of HELLP condition. As represented by this case, HELLP disorder can happen in the setting of a normotensive patient without proteinuria, yet with different side effects present (right upper quadrant torment, epigastric torment, windedness, and retrosternal chest torment). HELLP condition isn't generally clear in cases wherein the normal hemolysis, thrombocytopenia, or liver catalyst rise happens uniquely. It is moreover really challenging to analyze hepatic burst in its beginning phases due to its changed show. The way to determination is, in this way, a high file of doubt. A pivotal highlight making the conclusion of HELLP disorder is the thought of hepatic burst optional to anticoagulant treatment. Various case reports imply hepato-splenic crack auxiliary to anti-thrombolytic specialists, warfarin, intravenous heparin, and more current unfractionated heparin arrangements, for example, Dalteparin and Enoxaparin conversely, intravenous heparin treatment in our patient endured exclusively for 3 days. It is to be sure a really uncommon occasion when an organ immediately breaks with practically no perceptible sore. Ostensibly, this supports a hypothetical earlier pathology in this understanding's liver (periportal rot in HELLP condition) that at last prompted hepatic break optional to HELLP disorder. Moreover, the subcutaneous heparin portion that our patient got isn't accounted for anyplace in that frame of mind as a portion that might actually cause the crack of an intra-stomach viscera. It would be ideal to approve the analysis of HELLP condition through placental pathology, which tragically was not acted for this situation.

There were a few traps of the conclusion of HELLP condition for this situation. The essential trap was the way that windedness and retrosternal torment were ascribed to aspiratory embolism (PE). This finding depended on an imaging concentration on that demonstrated a doubt of reciprocal PE notwithstanding typical Pulse oximetry and venous Doppler studies. Second, slight rises in pulse, gentle thrombocytopenia, and gentle heights in liver chemicals are potential appearances of early HELLP disorder. In this specific case, the patient was unpretentiously presented with windedness, chest torment, and raised liver compounds unaccompanied by thrombocytopenia or hemolysis. While the succeeding research facility results showed HELLP disorder, the underlying finding of aspiratory embolism was as yet kept up with. Third, the bounce-back expansion in platelets was credited to steroid organization as it were. Thus, the finding of the HELLP condition was killed naturally from the differential determination. But on readmission, the way that the patient displayed extreme right upper quadrant torment optional to subcapsular hematoma ought to have educated the clinician to a finding of HELLP condition. Luckily, the patient endures such an overwhelming and possibly lethal sequela of liver damage.

4. CONCLUSION

HELLP condition is extremely uncommon and is chiefly brought about by toxemia. Care ought to be taken during pregnancy to advance fetal turn of events. Transfusions of blood items like platelets, red platelets, and new frozen plasma are required. In early pregnancy, corticosteroids might be utilized to animate the development of the fetal lungs. Antihypertensive specialists are likewise used to control hypertension. Patients with HELLP disorder require essential observation. Early identification and treatment of HELLP disorder, either by inception or typical work or by cesarean area, is useful for both mother and embryo and assists with forestalling further inconveniences.

In outline, we depict an instance of HELLP disorder with an abnormal show at first analyzed and treated as pneumonic embolism. We, in this manner, suggest an objective stepwise methodology toward the finding of HELLP disorder and its abnormal show to forestall the expensive result of a missed determination and it's going with maternal and perinatal bleakness and mortality. In a patient giving any unexplainable research facility irregularity of one or the other Hemolysis or raised liver chemicals or low platelets, the file of doubt for HELLP disorder ought to stay high within the sight of side effects, for this situation, windedness and retrosternal torment. When a patient gets back

with demolishing side effects, we suggest utilizing subordinate tests, for example, uterine Doppler review or stomach ultrasound image, which could prompt the right conclusion of HELLP disorder.

CONSENT

Every author proclaims that informed consent was acquired from the patient and her husband, and the signed consent form is submitted alongside the manuscript and submission form.

ETHICAL APPROVAL

It is not applicable.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Chawla Sushil, Marwaha Ashish, Agarwal Raju. HELLP or help: A Real Challenge Journal of Obstetrics and Gynecology of India (May-June 2015) 65(3):172-175 DOI 10.1007/s13224-014-0482-8.
2. Aesha A. Patel, Tushar M. Shah. A Case report of HELLP syndrome, IAIM. 2015;2(1):108-111.
3. Nidhi O. Bang, Meena N. Satia, Supriya Poonia. Obstetric and neonatal outcome in pregnancies complicated by hemolysis elevated liver enzymes low platelet count syndrome at a tertiary care center in India, Int J Reprod Contracept Obstet Gynecol. 2016;5(7):2407-2412.
4. Ashwini Mallesara, Srushti R. Kanta, Prashanth Shivappa, A clinical study of HELLP syndrome and its outcome in a tertiary health care system, Int J Reprod Contracept Obstet Gynecol. 2016;5(12): 4196-4199.
5. Lachmeijer AM, Arngrı´msson R, Bastiaans EJ. A genome-wide scan for preeclampsia in the Netherlands. Eur J Hum Genet. 2001;9:758.
6. Abildgaard U, Heimdal K. Pathogenesis of the syndrome of hemolysis, elevated liver enzymes, and low platelet count(HELLP): a review. Eur J Obstet Gynecol Reprod Biol. 2013;166:17.
7. Michael Geary. The HELLP syndrome, British Journal of Obstetrics and Gynaecology. 1997; 104:887 – 891.
8. Kestutis Rimaitis, Lina Grauslyte, Asta Zavackiene, Vilda Baliuliene, Ruta Nadisauskiene, Andrius Macas. Diagnosis of HELLP syndrome: A 10-year survey in a perinatology centre, Int. J. Environ. Res. Public Health. 2019;16:109.
9. Douglas M Woudstra, Sue Chandra, G Justus Hofmeyr, and Therese Dowswell. Corticosteroids for HELLP (hemolysis, elevated liver enzymes, low platelets) syndrome in pregnancy, CDSR. 2010;9.
10. Audibert F, Friedman SA, Frangieh AY, Sibai BM. Clinical utility of strict diagnostic criteria for the HELLP (hemolysis, elevated liver enzymes, and low platelets) syndrome. Am J Obstet Gynecol. 1996; 175:460–464? DOI: 10.1016/S0002-9378(96)70162-X.
11. Martin JN, Jr, Rose CH, Briery CM. Understanding and managing HELLP syndrome: The integral role of aggressive glucocorticoids for mother and child. Am J Obstet Gynecol. 2006;195:914–934? DOI: 10.1016/j.ajog.2005.08.044.
12. Sibai BM. The HELLP syndrome (hemolysis, elevated liver enzymes, and low platelets): much ado about nothing? Am J Obstet Gynecol. 1990; 162:311–316?
13. Barton JR, Sibai BM. Diagnosis and management of hemolysis, elevated liver enzymes, and low platelets syndrome. Clin Perinatal. 2004;31:807–33. DOI: 10.1016/j.clp.2004.06.008.
14. William M. Curtin, Louis Weinstein. A review of HELLP syndrome. Journal of Perinatology, 1999;19(2):138–143.
15. Eklof B, Gjores JE, Lohi A, Staszkiewicz W, Norgren L. Spontaneous rupture of liver and spleen with severe intra-abdominal bleeding during streptokinase treatment of deep venous thrombosis. Vasa. 1977;6:369-71.
16. Taccone FS, Starc JM, Sculier JP. Splenic spontaneous rupture (SSR) and hemoperitoneum associated with low molecular weight heparin: A case report. Support Care Cancer 2003;11:336-8.
17. Arnulf G, Chacomac R, Morel JP. Hémorragie gravissime par rupture d'hématome souscapsulaire rate saine au cours d'héparinisation pour désoblitération artérielle. Lyon Chirurgical. 1968;64: 961-4.

18. Weiss SJ, Smith T, Laurin E, Wisner DH. Spontaneous splenic rupture due to subcutaneous heparin therapy. J Emerg Med. 2000;18:421-6.
19. Stella CL, Sibai BM. Preeclampsia: Diagnosis and management of the atypical presentation. J Matern Fetal Neonatal Med 2006;19:381-6.
20. Petca A, Miron BC, Pacu I, Dumitrașcu MC, Mehedințu C, Șandru F, Petca RC, Rotar IC. HELLP syndrome—holistic insight into pathophysiology. Medicina. 2022;58(2):326.
21. Bazzan M, Todros T, Tedeschi S, Ardissino G, Cardaropoli S, Stella S, Montaruli B, Marchese C, Roccatello D, Cugno M. Genetic and molecular evidence for complement dysregulation in patients with HELLP syndrome. Thrombosis Research. 2020;196:167-74.

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