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A Case Report of a Patient with Postpartum HELLP Syndrome

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ABSTRACT

Background: HELLP syndrome is a multisystemic disorder characterized by elevated liver enzymes, hemolysis and low platelet count. If left untreated, it is associated with high risk of maternal and fetal mortality. It usually occurs in the third trimester of pregnancy but may sometimes occur after pregnancy. Herein, we report a patient with postpartum HELLP syndrome.

Case description: A 32-year-old woman (G2Ab1) with gestational age of 36 weeks and a history of hypothyroidism, multiple sclerosis, favism, gestational diabetes and pregnancy-induced hypertension was admitted to hospital due to labor pain. The patient underwent cesarean section and showed triad of postpartum HELLP syndrome. Fortunately, with timely diagnosis and appropriate intervention, the patient was discharged with good general condition after four days of hospitalization in intensive care unit.

Conclusion: Pregnancy-induced hypertension is a life-threatening condition for mothers. HELLP syndrome is often related to preeclampsia but can also occur as a stand-alone disorder. Absence of symptoms should not rule out this syndrome, and it is recommended to consider risk of postpartum HELLP syndrome during follow ups.

Keywords: HELLP syndrome; Preeclampsia

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INTRODUCTION

Triad of hemolysis, elevated liver enzymes and low platelet count known as the HELLP syndrome, is a serious and life-threatening form of preeclampsia. The incidence of the disease is estimated to be 0.17-0.85% per live birth. HELLP syndrome usually occurs in at gestation weeks 32-34 and in 30% of postpartum cases. The most important clinical symptoms include right upper quadrant or epigastric pain accompanied with nausea and vomiting (1). Common complications of the disease are disseminated intravascular coagulation (DIC), placental abruption, pulmonary edema, subcapsular liver hematoma, retinal detachment, laparotomy due to severe intra-abdominal bleeding and maternal death. Other complications include acute respiratory distress syndrome, shock, sepsis, kidney failure and incision site bleeding in patients with thrombocytopenia. Fetal outcomes are strongly dependent on the disease severity and include prematurity, intrauterine growth restriction and placental abruption. However, newborns are not at increased risk of liver disease or thrombocytopenia compared to healthy counterparts (2). Herein, we report a patient with postpartum HELLP syndrome.

CASE PRESENTATION

The patient was a 32-year-old pregnant woman (BMI of 23.7 kg/m^2) with gestational age of 36 weeks and 3 days, a second pregnancy and a history of abortion (G2Ab1). On January 5, 2018, the patient was admitted to hospital for cesarean section. The patient had contraction and in the vaginal examination: cervix was 3 cm dilated, 40% effaced and station was -3.

Blood pressure (BP) and fetal heart rate were in the normal range. The patient had a history of pre-pregnancy hypothyroidism, hypertension and gestational diabetes and was on daily treatment with oral levothyroxine, methyldopa and insulin, respectively. In addition, the patient had a history of favism and multiple sclerosis that were controlled during the pregnancy without medication.

For cesarean section, the patient was transferred to the operating room at 5 pm and underwent general anesthesia. A baby with intrauterine growth restriction, meconium aspiration and Apgar scores of 8 and 10 was born. Results of preliminary laboratory tests were obtained during the surgery which indicated severe liver dysfunction and coagulation abnormalities. Table 1 shows the results of laboratory tests from admission to discharge.

Table 1. Results of the laboratory tests from admission to discharge

Test parameter	At admission	Post-op day 2	Post-op day 3	Post-op day 4	At discharge
Hemoglobin (g/dl)	13.3	8	7	9	9.4
Hematocrit (%)	41.3	NR	NR	NR	NR
White blood cell (cell/mm^3)	13,000	NR	NR	NR	NR
Bile salt (mg/dl)	61	NR	NR	NR	NR
Alkaline phosphatase (U/l)	1,279	441	263	253	NR

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SGOT (U/l)	114	50	44	50	66
SGPT (U/l)	134	45	27	25	31
LDH (U/l)	739	770	617	NR	NR
Urea (mg/dl)	38	NR	NR	NR	NR
Creatinine (mg/dl)	1.4	NR	NR	NR	NR
Total bilirubin (g/dl)	17.9	9.5	7.2	8.3	9
Direct bilirubin (mg/dl)	11.8	6.9	5.1	5.6	6.4
Platelet (per mm^3)	109,000	73,000	72,000	79,000	68,000
Prothrombin time (Sec)	NR	21.4	16	14.9	14.3
Partial thromboplastin time (Sec)	NR	75.8	31	30	30
INR ratio	NR	2.05	1.8	1.6	1.5
Fibrinogen (mg/dl)	NR	<80	283	310	300

SGOT: Serum glutamic oxaloacetic transaminase, SGPT: Serum glutamic pyruvic transaminase, LDH: Lactate dehydrogenase, NR: not reported.

During the surgery, the uterus was completely yellow and yellow ascitic fluid was observed. Postoperative BP was 155/90 mmHg and pulse rate was 90. The patient was transferred to the intensive care unit (ICU) at 6:30 pm because of severe icterus and impaired hepatic and clotting enzymes. Magnesium sulfate injection (2 g/hour) was initiated. Postoperative liver dysfunction was decreased but still high. At 11:30 pm, the patient developed tachycardia (pulse rate= 120) and experienced a drop in BP (90/50 mmHg), hemoglobin (8.5 g/dl) and platelet count (86,000 per mm^3) as well as leukocytosis (WBC=25500) and oliguria. The patient also had diplopia and blurred vision, coagulation factors were severely impaired (PT = 33.9, PTT> 120, INR = 5.7) and the patient experienced anuria at midnight. Metabolic acidosis was reported in the arterial blood gas test. At 3 am, abdominal and pelvic ultrasound was performed and intra-abdominal free fluid and blood accumulation in the subhepatic space and grade I fatty liver were reported. The patient was transferred to the operating room after receiving 2 units of packed cells and 4 units of fresh frozen plasma (FFP). At first, a vascular surgeon placed a central venous line for the patient due to hypotension, the urgent need for rapid infusion of fluid and blood products as well as an increase in INR. Then, diagnostic laparotomy was performed at 4:45 am which indicated ecchymosis and subperitoneal

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diffuse hematoma with brief intra-abdominal hemorrhage. Two intra-abdominal and sub-fascicular drains were placed due to coagulopathies and the possibility of subsequent bleeding, and the patient was transferred to the ICU after recovery.

Based on the clinical and laboratory findings, the patient was diagnosed with HELLP syndrome and a team of doctors composed of a gynecologist, neurologist, infectiologist, internist, cardiologist, oncologists, nephrologist and gastroenterologist was assigned to the patient. Peripheral blood smear was sent to the Comprehensive Cancer Center, which did not raise the possibility of DIC. Due to the possibility of acute liver failure, necessary arrangements were made for liver transplantation at Imam Khomeini Hospital in Tehran, Iran. The patient received two FFP units every 8 hours. On the second day, she received 2 units of blood and 5 units of platelets. The injection of magnesium sulfate was discontinued 24 hours after delivery. On the third day, she received 2 units of blood, and administration of 2 units of FFP every 8 hours was continued. On the fourth day, she received a unit of blood and FFP injection continued. Abdominal and pelvic ultrasound was normal and intra-abdominal free fluid was absent. The patient's coagulation factors were normal and the patient was transferred from the ICU to a ward upon her husband's request due to financial problems. The patient was alert during the hospitalization period and did not have seizures. After giving a personal consent due to financial problems, she was discharged on the fifth day after receiving necessary trainings and medication instructions.

.DISCUSSION

HELLP syndrome is characterized by hemolysis, elevated liver enzymes and low platelet count, and is associated with high maternal and fetal mortality if left untreated. It is usually related to preeclampsia, but due to lack of proteinuria and elevated BP (seen in 15-20% of patients), the syndrome is sometimes considered different from preeclampsia (3-5). The diagnosis is made based on laboratory findings, including microangiopathic hemolytic anemia characterized by presence of schistocytes in the peripheral blood smear. Other symptoms of hemolysis include increased serum LDH (≥600 IU/l) and indirect bilirubin, decreased serum haptoglobin (≤25 mg/dl), platelet count of ≤100000 cell/µl, total bilirubin of <1.2 mg/dl and serum AST of ≤ 70 U/l. Some researchers consider ALT alone or in addition to ALP as a criterion for HELLP syndrome, but AST indicates hepatocellular necrosis as well as RBC hemolysis. The severe maternal complications occur rapidly and include DIC, placental abruption, acute liver failure, pulmonary edema, subcapsular liver hematoma and retinal detachment. Jaundice and ascites may also occur. Thrombocytopenia-associated bleeding is uncommon and some patients are asymptomatic. Proteinuria and hypertension are seen in 85% of patients (2). Hypertension is absent in 20% of patients with HELLP syndrome, and 5-15% of pregnant patients have mild or no proteinuria. Early detection of hemolysis is more sensitive than determination of serum hepatoglobulin. Increase in AST and ALT often occurs before platelet count falls (1). There are currently two classification system diagnostic criteria for HELLP syndrome: The Tennessee classification system (platelets < 100×10^{9}, AST > 70 IU, LDH > 600 IU/l) (5) and the Mississippi classification system (Class I: platelets < 50×10^{9}, AST > 70 IU, LDH > 700 IU/l; Class II: 50×10^{9} < platelets < 100×10^{9}, AST or ALT > 70 IU, LDH > 600 IU/l; Class III: 100×10^{9} < platelets < 150×10^{9}, AST or ALT > 40 IU, LDH > 600 IU/l) (6).

Depending on the timing of its occurrence, the syndrome could be classified into postpartum or prepartum. In case of prepartum HELLP syndrome, treatment depends on the gestational age. In cases with gestational age of more than 34 weeks, immediate termination of pregnancy is recommended after monitoring BP. In addition, in cases with gestational age of 27-34 weeks, delivery can proceed normally 48

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hours after receiving corticosteroids and monitoring BP (7).

A rapid decline in platelet count, particularly on the third postpartum day is observed in patients with postpartum HELLP syndrome. Subjects with this condition are at high risk of renal failure and pulmonary edema (8).

It has been shown that administration of corticosteroids in patients with HELLP syndrome can increase platelet count and decrease serum LDH and ALT levels, thus shortening hospital stay and reducing the need for blood transfusion. However, it does not reduce the risk of maternal or fetal mortality and overall complications (9). Plasmapheresis has been recently suggested as an alternative treatment for HELLP syndrome (10).

Differential diagnoses include acute fatty liver of pregnancy, gastroenteritis, hepatitis, appendicitis, gallbladder diseases, antiphospholipid syndrome, thrombotic thrombocytopenic purpura, hemolytic uremic syndrome, immune thrombocytopenic purpura and lupus flare. Recurrence of hypertensive disorders in pregnancy occurs in 27% to 48% of cases (1). Patients with a history of HELLP syndrome are at increased risk of preeclampsia in subsequent pregnancies (11-13). There is currently no preventive measure for HELLP syndrome.

CONCLUSION

Given the history of pregnancy induced hypertension and treatment with methyldopa and the clinical and laboratory findings, the patient was diagnosed with postpartum HELLP syndrome class II according to the Mississippi criteria. Interestingly, symptoms of severe preeclampsia appeared unexpectedly in the patient. Moreover, the patient presented with labor pain and did not have common symptoms of the syndrome, such as hypertension. If left untreated, postpartum HELLP syndrome and its complications could be life threatening. Fortunately, our case was eventually discharged with a good general condition due to timely diagnosis and appropriate treatment interventions. Early diagnosis of HELLP syndrome for prevention of complications in susceptible individuals is crucial and lifesaving.

DECLARATIONS

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Ethics approvals and consent to participate

Written consent was obtained from the patient and she was assured that her personal information would remain confidential.

Conflict of interest

The authors declare that there is no conflict of interest regarding publication of this article.

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