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Case Report

Atypical HELLP Syndrome in a Hydatidiform Molar Pregnancy: A Case Report

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Abstract

HELLP syndrome is a serious complication in pregnancy characterized by hemolysis, elevated liver enzymes and low platelet count. HELLP syndrome is often thought to be an advancement of preeclampsia with associated elevated blood pressure and proteinuria. HELLP syndrome may develop during the antepartum or postpartum period. HELLP syndrome occurs in 0.1%-0.6% of all pregnancies and in 4%-12% of patients with preeclampsia. The true etiology of HELLP syndrome remains unclear; however it is thought to be a manifestation of pre-eclampsia. Early diagnosis is imperative due to the morbidity and mortality associated with this syndrome. Treatment of HELLP syndrome includes supportive therapy with antihypertensive medications and anticonvulsants. Definitive management includes delivery irrespective of gestational age. This case report involves a 29 year old G8P4034 with Chronic hypertension at 17 weeks 5 days who presented with superimposed pre-eclampsia/HELLP syndrome based upon new onset proteinuria, increased and uncontrolled blood pressure, increase in liver enzymes, and the 1999 guidelines of the Mississippi Classification system. Patient elected for termination and subsequent karyotype was significant for 69, XXY. There was no evidence of a molar pregnancy prior to this examination.

ABBREVIATIONS

HELLP: Hemolysis, Elevated Liver Enzymes, and Low Platelets; ALT: Alanine Aminotransferase, AST: Aspartate Aminotransferase

INTRODUCTION

HELLP syndrome is a serious complication in pregnancy characterized by hemolysis, elevated liver enzymes and low platelet count [1]. First named by Weinstein in 1982, HELLP syndrome is a very serious manifestation of elevated blood pressure and proteinuria in pregnancy. Among pregnant women in the United States, approximately 5-8% will develop preeclampsia. Of those women, 15% will develop HELLP syndrome [2]. As many as 48,000 women per year will develop HELLP in the United States. The pathophysiology of HELLP syndrome is still not well defined. Several theories suggest that HELLP is a progression of preeclampsia. Others believe that HELLP syndrome is an entity of its own. Although the cause of HELLP syndrome is unknown, certain risk factors, including a maternal age of older than 34 years, multiparity, and European descent, have been described [3]. HELLP syndrome typically occurs between week 27 of gestation and delivery, or immediately postpartum in 15%-30% of cases [3]. The typical patient with HELLP syndrome will present with signs or symptoms similar to preeclampsia such as elevated blood pressure, uncontrolled headache, visual disturbances, and/or hyperreflexia. However, HELLP syndrome may also result in more serious complaints such as right upper quadrant or epigastric pain suggestive of liver involvement. The initial evaluation of HELLP syndrome often includes laboratory work up with a complete blood count, comprehensive metabolic panel, lactate dehydrogenase level, uric acid level, and urinalysis. The serum transaminase levels may be elevated to as high as 4,000 U per L, but milder elevations are typical. Platelet counts can drop to as low as 6,000 per mm^3, but any platelet count less than 150 per mm^3 warrants attention [4]. Once the diagnosis of HELLP syndrome has been established, the best markers to follow are the maternal lactate dehydrogenase level and the maternal platelet count [5]. Laboratory abnormalities typically worsen after delivery and peak at 24 to 48 hours postpartum [5].

At present, there are two major definitions for diagnosing HELLP syndrome. In the Tennessee Classification System, the hemolysis component of HELLP syndrome is diagnosed by abnormal peripheral blood smear, increased serum bilirubin (≥ 20.5 µmol/L or ≥ 1.2 mg/100 mL) and elevated LDH levels (> 600 units/L (U/L). The Mississippi Triple Class System (1999) further classifies the disorder based on the nadir platelet count at any

time during the course of the disease. Class I has platelets less than 50,000 per mm^3, Class II has platelets between 50,000 to 100,000 per mm^3 and Class III has platelets between 100,000 and 150,000 per mm^3 or AST >40 IU/L.

Patients with HELLP syndrome should be treated prophylactically with magnesium sulfate to prevent seizures, whether hypertension is present or not. A bolus of 4 to 6 g of magnesium sulfate as a 20 percent solution is given initially. This dose is followed by a maintenance infusion of 2 g per hour. The infusion should be titrated to urine output and magnesium level. Patients should be observed for signs and symptoms of magnesium toxicity [4]. Additionally, antihypertensive medications should be used to control blood pressure. Target blood pressure includes systolic pressure less than 160mmHg and diastolic pressure less than 100mmHg. The most common antihypertensive medications used are intravenous hydralazine and labetalol. The definitive management of HELLP syndrome is delivery of the fetus/infant. For women with HELLP syndrome and gestational age before fetal viability, it is recommended that delivery be undertaken shortly after maternal stabilization [6]. For women with HELLP syndrome and gestational age between 24 0/7 weeks and 34 0/7 weeks, delivery is recommended after a short course of antenatal corticosteroids if maternal and fetal condition is stable [6]. For women with HELLP syndrome and gestational age after 34 0/7 weeks, delivery is recommended after initial maternal stabilization [6]. Mode of delivery should be based on patient condition, gestational age, and Bishop score. The laboratory abnormalities in HELLP syndrome typically worsen after delivery and then begin to resolve by three to four days postpartum [5].

CASE PRESENTATION

A 29 year old Gravida 8 Para 4034 with recent diagnosis of chronic hypertension at an estimated gestational age of 17 weeks and 5 days based on an 11 week ultrasound presented to the Howard University Hospital Emergency Department with a chief complaint of chest pain, epigastric pain, and headache. Initial evaluation in the Emergency Department revealed an initial blood pressure of 173/115, brisk deep tendon reflexes and 2+ pitting edema. Baseline investigations revealed Hemoglobin 12.2 gm/dl, platelets 157,000mm^3, alanine aminotransferase (ALT) 42 IU/L, aspartate aminotransferase (AST) 92 IU/L, lactate dehydrogenase 323 IU/L, uric acid 6.1 mg/dl , and urine protein 600mg/dl. Electrocardiogram showed a normal sinus rhythm. Cardiac enzymes were negative. The patient was admitted for uncontrolled chronic hypertension and 24-hour urine collection was started for total protein and creatinine clearance. Despite management with intravenous antihypertensive medications, blood pressures were persistently elevated with a range of systolic BP 170-200 mmHg and diastolic BP 90-110s. Internal Medicine and Maternal Fetal Medicine were consulted for further investigation of other causes for uncontrolled hypertension and additional recommendations for blood pressure management. Sonogram performed by Maternal Fetal Medicine revealed intrauterine growth restriction with the fetus measuring only 14 weeks 4 days. Platelet count fell to 112,000mm^3. 24-hour urine protein resulted in 11.3 grams (compared to baseline of 236 mg). Diagnosis of Chronic Hypertension with superimposed Pre-Eclampsia /HELLP syndrome was confirmed. The patient was counseled on risks/benefits/and alternatives associated with HELLP syndrome and current recommendations on management. The patient decided to terminate the pregnancy and informed consent was obtained. The patient was started on magnesium sulfate for seizure prophylaxis. Induction of Labor was begun with misoprostol per vagina. After two doses of misoprostol, the patient spontaneously delivered a non-viable fetus. Magnesium sulfate was continued for a total of 24 hours post- delivery. Oral Labetalol and Hydrochlorothiazide for blood pressure management. Repeat investigations post delivery revealed Hemoglobin 11.2mg/dl, platelets 134,000, alanine aminotransferase (ALT) 33 IU/L, aspartate aminotransferase (AST) 58 IU/L, lactate dehydrogenase 287 IU/L, and Uric Acid 6.3 mg/dl. The patient continued to improve clinically and was discharged home on hospital day number 4 in stable condition. Pathology report revealed a male fetus weighing 49 grams with no congenital anomalies identified and a karyotype of 69, XXY associated with a partial hydatidiform mole. Upon review of the prenatal records, there were no previous findings suspicious for a molar pregnancy. The gross pathologic specimen consisted of a placenta with attached cord and membranes. The umbilical cord measures 28 cm and is attached to the placenta and fetus. It has a diameter of 0.3 cm and is inserted 4.0 cm from the margin of the disk. Vascular lumens cannot be identified grossly. The membranes are thin and translucent and show normal insertion onto the disk. The rupture point cannot be appreciated. The disk is ovoid in shape and measures 16 x 9.0 x 1.0 cm and weighs 119 grams. The fetal surface is pink in color. The maternal surface shows the usual cotyledons and cut surfaces are pale brown in color and spongy. The fetus is phenotypically male and weighs 49 grams with the following circumference: Head 7.8 cm, chest 7.0 cm, abdomen 8.2 cm, crown-rump length 9.5 cm, foot length 1.5 cm. No gross external or internal abnormalities are appreciated. There were increased nucleated red blood cells in fetal circulation. The placental weight of 119gm differs from 18 weeks mean weight in the literature. The placenta was immature with areas of focal intervillous hemorrhage. The three vessel umbilical cord had no pathologic diagnosis. Prior to discharge, patient received Depo Provera 150mg intramuscular. At follow-up visit with primary obstetrician, beta HCG was negative.

DISCUSSION

HELLP is a multi-system disease, resulting in generalized vasospasm, microthrombi formation and coagulation defect. The clinical course of a woman with HELLP syndrome is often characterized by progressive and rapid deterioration of the maternal and/or fetal condition [6]. Significant signs and symptoms in any patient with preeclampsia include headache, blurred vision, altered consciousness, clonus, increasing serum creatinine level, consumptive coagulopathy with thrombocytopenia, and abnormal liver function tests. These signs and symptoms were evident in our case presentation. Based on the gestational age in this case, immediate delivery was recommended. After delivery, immediate resolution of abnormal laboratory values was observed. Most patients with HELLP syndrome stabilize within 24-48 hours. The mortality rate for women with HELLP syndrome is approximately 1.1

percent [7]. From 1-25 % of affected women develop serious complications such as disseminated coagulopathy, placental abruption, adult respiratory distress syndrome, hepatorenal failure, pulmonary edema, subcapsular hematoma, and hepatic rupture. A significant percentage of patients receive blood products [7]. Patients who have had HELLP syndrome should be counseled that they have a 19 to 27 percent risk of developing the syndrome in subsequent pregnancies [8]. They also have up to a 43 percent risk of developing preeclampsia in another pregnancy [8]. Patients with class I HELLP syndrome have the highest risk of recurrence. An interesting finding in our case was the presence of a partial hydatidiform mole on pathology. A hydatidiform mole is characterized as abnormal proliferation of the syncytiotrophoblast and replacement of normal placental trophoblastic tissue by hydropic placental villi. Complete moles do not have identifiable embryonic or fetal structures. Partial moles are characterized by focal trophoblastic proliferation, degeneration of the placenta, and identifiable fetal or embryonic structures [9]. Abnormal proliferation of syncytiotrophoblasts lead to hemoconcentration and alteration in vascular hemodynamics. This abnormal placentation may often lead to elevated blood pressure or even progress to preeclampsia or HELLP syndrome as seen in our case presentation. In the management of a hydatifidorm mole, it is very important to evacuate all fetal tissue and close follow-up is recommended to ensure gestational trophoblastic neoplasia does not persist. Our patient was given adequate contraception after termination of the pregnancy. Postpartum follow-up displayed a negative beta HCG suggestive of no evidence of persistent trophoblastic tissue.

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