A Case of Fatal Maternal-Fetal Conflict: HELLP Syndrome Diagnosed in a Second Trimester Pregnancy

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ABSTRACT

HELLP syndrome is part of the preeclampsia-eclampsia spectrum, and it typically presents after 20 weeks of gestation. In this case report, we review the hypotheses for the pathophysiology of HELLP and report one of the few cases of the syndrome presenting prior to 20 weeks of gestation.

Keywords

Pregnancy, HELLP, Second-Trimester.

Introduction

HELLP syndrome is characterized by hemolysis, elevated liver enzymes, and thrombocytopenia. This syndrome is considered to be a severe variant of preeclampsia, and like preeclampsia, it most often presents in the third trimester of pregnancy or in the early postpartum period. Here we report a case of HELLP syndrome presenting at 18 weeks of gestation. HELLP syndrome, especially in its early stage is often associated with antiphospholipid antibody syndrome.

Case Presentation

A 36 year-old Hispanic female G3P2002 with confirmed intrauterine pregnancy of 18 weeks 0 days was admitted to our institution for management of hypertension. She reported a recent onset of headache, scotoma, epigastric pain, an episode of non-bloody, non-bilious vomiting and bilateral lower extremity edema for 2 weeks. She denied any other associated symptoms. Her past medical history was significant for obesity, preeclampsia, antepartum transient hypertension, and two cesarean deliveries.

Upon admission, her vital signs were significant for blood pressures of 150/90 to 160/100, lowering to 140-150/90 after one dose of

labetalol 20 mg intravenous (IV push). Her laboratory data were significant for a hemoglobin of 10.2 g/dL, hematocrit of 31.1%, platelets of 92,000/UL (down from 182,000 /UL in a visit 3 months prior), aspartate transaminase of 103 U/L, alanine transaminase of 110 U/L, and lactate dehydrogenase of 284 U/L. Her urine protein/creatinine ratio was 2.30 and urine creatinine was 352 mg/dL. She tested negative for Covid-19. Ultrasonography confirmed a single intrauterine gestation, with the fetal biometric measurements appropriate for gestational age and a fetal heart rate of 153 beats per minute.

With a diagnosis of maternal HELLP syndrome and a previable fetus, a magnesium sulfate infusion was started for seizure prevention and the patient was counseled about delivery of the fetus, a potentially life saving decision for obvious maternal indication. The patient opted for dilation and evacuation (D&E). She was then transferred to an institution where the D&E procedure was safely performed.

Discussion

HELLP (hemolysis, elevated liver enzymes, and low platelets) syndrome is considered a more severe version of preeclampsia and complicates 0.01–0.6% of pregnancies [1]. About 70% of HELLP syndrome cases occur in the third trimester of pregnancy, and the remainder occur within 48 hours of delivery [2]. The mortality rate

of women with HELLP syndrome is 0 to 24%, with a perinatal death rate of up to 37% [2]. Approximately, 5–10% of women with preeclampsia go on to develop HELLP syndrome [3]. However, it has been postulated that as many of 15–20% of HELLP syndrome patients do not have antecedent preeclampsia [1]. The average gestational age of HELLP syndrome is 34 weeks [2], with multiparous mothers over the age of 35 years old [4]. Common clinical presentation includes; hypertension, proteinuria, nausea and vomiting. Thirty to 60% of patients present with headache and 20% have a visual disturbance [4]. Differential diagnoses include acute fatty liver of pregnancy, thrombotic thrombocytopenic purpura, antiphospholipid syndrome, and hemolytic uremic syndrome [5].

This patient presented at 18 weeks gestation with evidence of hemolysis, elevated liver transaminases, and thrombocytopenia. While this patient had a history of preeclampsia in a previous pregnancy, this pregnancy was uncomplicated prior to presentation. HELLP syndrome is rare prior to 20 weeks of gestation, with only a limited number of cases reported in the literature. The earliest described case of HELLP syndrome was during the 8th week of gestation, one of the very few cases of HELLP syndrome in the first trimester of pregnancy [3]. The diagnosis of HELLP syndrome before the 20th week of gestation has been most commonly reported in association with antiphospholipid antibody syndrome (APS) or triploid chromosomal anomalies [6]. Risk factors associated with HELLP syndrome in patients without antiphospholipid syndrome or fetal aneuploidy include African American ethnicity, family history of hypertension, and undiagnosed chronic hypertension [7].

Several hypotheses about the initial insult leading to the pathophysiology of HELLP syndrome have been suggested. One hypothesis suggests the alteration in maternal-fetal immune balance creates a maternal immune rejection of a genetically foreign fetus [1]. Another hypothesis proposed that HELLP is an acute inflammatory condition instigated by the placenta [1]. It has also been postulated that HELLP syndrome may be due to poor placentation during early pregnancy, with involvement of the coagulation cascade and complement system [2]. Of note, studies done on women diagnosed with early HELLP syndrome showed clinical and laboratory value improvement when taking complement C5 protein inhibitors [2]. Ultimately, the inflammatory response seen in HELLP syndrome leads to an ischemic-reperfusion injury. This injury activates the coagulation cascade, consuming platelets and shearing red blood cells as they traverse platelet-fibrin rich capillaries, causing the formation of schistocytes and subsequently microangiopathic hemolytic anemia. This microvascular injury impacts multiple organ systems, including the liver. The combination of fibrin deposits and intrahepatic vascular congestion leads to hepatic necrosis and ultimately the development of HELLP syndrome. In regards to the diagnosis of HELLP syndrome, classification systems used include the Tennessee Classification system and the Mississippi Classification

system. The Tennessee classification system for HELLP includes hemolysis, increased LDH (≥ 600 IU/L), increased AST (≥ 70 IU/L), and low platelets ($<100 \times 10^9/L$) [8]. The Mississippi classification system categorizes the severity of HELLP syndrome into Class I, II, and III based on platelet count, LDH, and AST (8). Both of these classification systems utilize a clinical diagnosis of a LDH of ≥ 600 IU/L for classes 1, 2, and 3. However, according to Smulian et al. LDH values below 600 IU/L may be expected due to different laboratory methods utilized to calculate the value, which may be the case for this patient [8].

Conclusion

In conclusion, this case provided some diagnostic and therapeutic challenges due to gestational age at presentation. Clinicians should consider possible early presentations for common pathologies that may complicate pregnancy such as HELLP syndrome when faced with the undeniable clinical evidence for the condition. This case illustrates an example of maternal-fetal conflict resulting in the discontinuation of the pregnancy before fetal viability with the goal of preventing serious maternal morbidity and lifelong disability or death. Utilizing the diagnostic criteria, clinical features, and laboratory data will allow a clinician to appropriately diagnose and manage similar patients, even if they present prior to 20 weeks of gestation.

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