Two Cases of Postpartum HELLP Syndrome: A Rare Presentation of Preeclampsia-Associated Liver Disease and Hepatocellular Dysfunction

Van Anh Do¹, Arvind R. Murali¹, Rutva Vora¹, Christine Greves³, and Steve J. Carlan⁴,*

ABSTRACT

Background: HELLP syndrome (hemolysis, elevated liver enzymes, and low platelets) is a severe and sometimes fatal pregnancy condition characterized by hemolysis, increased liver enzymes, and low platelet count. While most cases occur before delivery, approximately 25% of cases manifest within 48 hours after delivery. One rare but life-threatening complication of HELLP syndrome is intraparenchymal liver hematoma.

Case: In this report, we present two postpartum HELLP syndrome patients with diverse clinical manifestations. Case 1 involves a 30-year-old woman who presented with a significant subcapsular liver hematoma as a complication of postpartum preeclampsia-associated liver illness. Following conservative treatment, she improved significantly, and no surgical intervention was required. This type of subcapsular hematoma occurs in less than 2% of pregnancies complicated by HELLP. Case 2 describes an unusual HELLP presentation in a 38-year-old woman who was diagnosed with postpartum HELLP syndrome with acute liver injury and hepatic hemorrhage.

Conclusion: These unique presentations underscore the significance of early detection and prompt treatment of postpartum HELLP syndrome, as it can significantly reduce risks to the mother’s health and improve overall outcomes.

Keywords: HELLP syndrome, Preeclampsia.

1. INTRODUCTION

HELLP syndrome (hemolysis, elevated liver enzymes, and low platelets) is a serious and potentially life-threatening condition that can affect postpartum women. It is a variant of preeclampsia, a hypertensive disorder that occurs during pregnancy. HELLP syndrome affects a small percentage of pregnancies, occurring in around 0.2% to 0.6% of all cases. Superimposed HELLP, which is a combination of preeclampsia or eclampsia with HELLP, develops in 4% to 12% of women with these conditions.

HELLP typically presents during the antepartum period in about 70% of cases, with the bulk of instances taking place between weeks 27 and 37 of pregnancy [1]. However, it can also present during the postpartum period in 25%–30% of cases, especially within the first 48 hours. Complications such as disseminated intravascular coagulation (DIC), subcapsular liver hematoma, renal failure, pulmonary oedema, and renal failure are among the consequences that are more common in women with postpartum HELLP [2].

Postpartum HELLP syndrome presents significant complications, leading to considerable maternal morbidity, particularly during the immediate postpartum period. Therefore, timely detection and proper management are crucial to minimize potential difficulties and ensure optimal outcomes for affected women. In this report, we present two cases of acute postpartum HELLP syndrome with unique presentations that occurred after delivery following severe preeclampsia.

2. CASE PRESENTATIONS

2.1. Case 1

A 38-year-old G2P0010 woman at 39 weeks of gestation presented with complaints of no fetal movements starting the previous evening. Her prenatal course to that point had...
been uneventful. She was noted to be in spontaneous labor with an overall reassuring fetal monitoring strip with occasional fetal deceleration. During labor she was treated with magnesium sulfate (MgSo4) seizure prophylaxis because of a diagnosis of preeclampsia. The patient underwent a spontaneous vaginal delivery on the next day to a vigorous male infant with Apgar scores of 8 and 9. However, several hours after delivery, she reported worsening thoracic back pain.

The patient was transferred to the intensive care unit (ICU) due to severely elevated liver enzymes, thrombocytopenia, and a positive hemolysis panel concerned for HELLP syndrome with impending organ failure (Fig. 1). She was started on empiric acyclovir for possible herpes simplex virus infection. A computed tomographic (CT) scan of the abdomen showed a heterogenous nodular liver, raising suspicion of hepatocellular disease. Infectious disease workup including blood cultures, cytomegalovirus (CMV) culture, and herpes simplex (HSV) polymerase chain reaction (PCR) were negative and antiviral medication was discontinued.

The patient was managed conservatively as she was stable with no signs of bleeding or hemodynamic instability. The patient’s condition continued to improve, with liver enzymes showing significant improvement on day 5. A repeat ultrasound at this time showed stable hemorrhage compared to the previous CT but with small changes around the liver due to hemorrhage or microabscesses. Given the patient’s stable condition, hemodynamic instability and no signs of infection, there was a low suspicion of infectious causes. Repeat abdominal CT 3 days later only showed the stable known hemorrhage. Her condition continued to improve, and she was discharged after 10 days. Outpatient abdominal ultrasound outpatient confirmed general hepatocellular disease likely hepatosteatosis but revealed no other abnormalities.

### 2.2. Case 2

A 30-year-old woman, G2P1001, with a history of pre-eclampsia without severe features during pregnancy, presented to the labor and delivery unit and had a successful spontaneous vaginal delivery soon after. However, six hours post-delivery, the patient developed severe hypertension with arterial blood pressure of 195/91 mmHg, without tachycardia, tachypnea, or fever. She reported persistent right upper quadrant (RUQ) pain. On physical exam, tenderness on the RUQ was appreciated without signs of rebound tenderness or guarding. Vital signs were seen in Table III and corresponding laboratory values in Table IV.

The patient was admitted to the medical unit for a diagnosis of elevated liver enzymes and low platelets suspicious of HELLP syndrome. Transabdominal ultrasound revealed a complex multisepated lesion in the right hepatic lobe and a hypoechoic lesion in the left hepatic lobe, along with the presence of a subcapsular hematoma. Other liver-related disorders, additional testing including acute hepatitis panel, antinuclear antibody (ANA), anti-smooth muscle antibody, antimitochondrial antibody, ceruloplasmin, and alpha-1 antitrypsin deficiency (A1AT), were performed, all of which yielded negative results.

The patient was hemodynamically stable and no active intra-abdominal bleeding was observed. As the subcapsular hematomas were stable on imaging and the patient’s hemoglobin levels were steady, she was managed conservatively. On day 3 of admission, a repeat ultrasound showed stable subcapsular hematomas. Her transaminases improved on day 4 along with her platelets and
hemoglobin. The patient was discharged from the hospital after the 2-week postpartum period. Outpatient magnetic resonance imaging (MRI) abdomen with and without contrast performed one month after showed a decreasing size of chronic subcapsular hepatic hematomas. No other adenomas or lesions were appreciated.

3. Discussion

HELLP syndrome and hypertensive disorders are leading causes of maternal mortality. HELLP syndrome affects around 0.5%–0.9% of all pregnancies and accounts for 10%–20% of severe preeclampsia cases. The pathogenesis of liver involvement in HELLP syndrome is complex and unclear. An interesting theory proposes that intravascular fibrin deposition may lead to liver sinusoidal obstruction, which is associated with hypovolemia in patients with preeclampsia, and later develops into HELLP syndrome. Hepatic ischemia from this process may cause hepatic infarction, subcapsular hematomas, and intraparenchymal hemorrhage, with potential hepatic rupture in severe cases [3]. Spontaneous rupture of subcapsular liver hematoma during pregnancy occurs about 1 in 40,000 to 1 in 50,000 births and affects 1% to 2% of individuals with HELLP.

<table>
<thead>
<tr>
<th>Table IV: Patient 2: Laboratory Values on Admission and 6 Hours Post-Delivery</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Lab values</strong></td>
</tr>
<tr>
<td>Hematology:</td>
</tr>
<tr>
<td>White blood cell (per mm$^3$)</td>
</tr>
<tr>
<td>Hemoglobin (mg/dL)</td>
</tr>
<tr>
<td>Platelet (per mm$^3$)</td>
</tr>
<tr>
<td>Lactate dehydrogenase (IU/L)</td>
</tr>
<tr>
<td>Hepatology:</td>
</tr>
<tr>
<td>Aspartate aminotransaminase (IU/L)</td>
</tr>
<tr>
<td>Alanine aminotransaminase (IU/L)</td>
</tr>
<tr>
<td>Alkaline phosphatase (IU/L)</td>
</tr>
<tr>
<td>Bilirubin (mg/dL)</td>
</tr>
<tr>
<td>Nephrology:</td>
</tr>
<tr>
<td>BUN (mg/dL)</td>
</tr>
<tr>
<td>Creatinine (mg/dL)</td>
</tr>
<tr>
<td>Coagulopathy:</td>
</tr>
<tr>
<td>INR</td>
</tr>
<tr>
<td>aPTT (seconds)</td>
</tr>
<tr>
<td>PT (seconds)</td>
</tr>
<tr>
<td>Fibrinogen (mg/dL)</td>
</tr>
</tbody>
</table>

Fig. 1. Patient 1: CT scans (1–3) and ultrasound (4) of the abdomen-pelvis showing heterogeneous infiltrative appearance throughout the right hepatic lobe, possibly reflecting hepatic necrosis and/or hemorrhage in the setting of HELLP syndrome, with hepatomegaly and serpiginous hypodensity extending throughout the majority of the right hepatic lobe. *L–Liver.
syndrome [4], presenting a significant risk with over 50% of maternal and fetal deaths associated with this complication [4].

Treatment approaches for subcapsular liver hematoma (SLH) can be either conservative or invasive, guided by the American Association for the Surgery of Trauma (AAST) staging. In hemodynamically stable patients, conservative management with intravenous fluids and blood products corrects coagulopathy effectively. Serial imaging investigations are necessary to monitor subcapsular liver hematoma (SLH) size [5]. For patients with poor resolution and uncontrolled bleeding, surgical management may be necessary, involving perihepatic packing, surgical site drainage, ligation of portal vein or hepatic artery branches, omentum patching, and partial liver resection, and even liver transplantation has been considered [5].

Case 1 is intriguing as the patient developed SLHs in the setting of elevated liver enzymes and thrombocytopenia; however, there was no definitive evidence of hemolysis to confirm a diagnosis of HELLP syndrome. Subcapsular liver hematomas as a complication of preeclampsia alone are rare, making this case even more uncommon. To our knowledge, there have been two prior cases reported similar to ours. Anyfantakis et al. report a case of a 32-year-old primiparous female developing preeclampsia and fetal distress requiring an emergent caesarean delivery [6]. Following delivery, the patient experienced acute hemolysis and mildly elevated liver enzymes without thrombocytopenia, with an associated subcapsular hematoma of the right hepatic lobe observed on abdominal CT. Luhning reports a more complicated case of a 40-year-old female who was also diagnosed with preeclampsia at 39 weeks’ gestation requiring a cesarean delivery [7]. She developed a 16 cm subcapsular hepatic hematoma postpartum. Her hospital course was also complicated by an infected pleural effusion, requiring video-assisted thoracoscopic surgery. All patients eventually made a complete recovery.

The literature review on stable subcapsular hematomas reveals several relevant studies. Sibai conducted a 13-year retrospective review of three patients with subcapsular liver hematoma (SLH) [7]. Two of them were treated conservatively and discharged from the hospital, while the third patient underwent hepatic resection but unfortunately succumbed to multiple organ failure. In another study, Wicke et al. [8] conducted a 10-year retrospective review of 5 patients with subcapsular liver hematoma. Out of these, three patients were treated conservatively, while two required urgent surgical intervention, with one of them undergoing liver transplantation. Carlson et al. [9] reported a case of a hemodynamically stable patient with ruptured subcapsular liver hematoma during pregnancy. In their report, the patient received non-surgical conservative management.

In severe cases, subcapsular liver hematoma may rupture, a feared complication carrying a mortality that ranges from 16% to 59%. Shames et al. [10] report a total of 8 liver transplants in the United States performed for complications related to HELLP syndrome between 1987 and 2003. As of the most recent follow-up, 6 of the 8 patients are alive, with both deaths occurring within 1 month of transplantation, and 2 patients have required retransplantation.

HELLP syndrome sometimes presents as diffuse generalized liver hemorrhage and necrosis as in case 2. Our patient had controlled hemorrhage with minimal necrosis of the liver and made a complete recovery. In contrast, Mikolajczyk et al. report a case of hepatic infarction in a 30-year-old woman diagnosed with HS [11]. The patient
subsequently developed multi-organ failure and underwent liver transplantation with a complete recovery. Simic *et al.* report another case of HELLP syndrome complicated by ruptured subcapsular liver hematoma causing shock and multi-organ failure [12]. The patient was taken to emergent surgery, and the liver bleeding was stopped, but despite resuscitation attempts and permanent transfusion, the outcome of surgery was lethal.

Invasive radiology techniques may be considered in unstable patients. In cases of severe bleeding or hemorrhage, transarterial embolization may be used to block blood vessels supplying the bleeding site [13]. This technique can help control bleeding and prevent further complications. Furthermore, interventional radiology-guided drainage can be used for hematoma or liver infarction. If there are thrombotic complications leading to blood vessel blockages, angioplasty (with or without stenting) may be considered to restore blood flow.

4. CONCLUSION

Postpartum HELLP syndrome is a relatively rare yet clinically significant condition that can manifest in diverse ways. Timely identification and appropriate management are crucial to reducing severe maternal morbidity and mortality. The cases presented in this study underscore the importance of considering postpartum HELLP syndrome as a potential differential diagnosis in postpartum patients presenting with elevated liver function tests and thrombocytopenia. To ensure accurate diagnosis and optimal treatment for affected individuals, healthcare practitioners should remain vigilant for atypical presentations and conduct thorough evaluations.

**Data Availability**

All data underlying the results are available as part of the article and no additional source data are required.

**Funding**

Funding was not involved in the manuscript writing, editing, approval or decision to publish.

**Ethics Approval**

The project did not meet the definition of human subject research under the purview of the IRB according to federal regulations and therefore was exempt.

**Conflict of Interest**

Authors declare that they do not have any conflict of interest.

**Abbreviations**

HELLP syndrome: hemolysis, elevated liver enzymes, and low platelets

MgSO4: magnesium sulfate

ICU: intensive care unit

CT: computed tomographic

CMV: cytomegalovirus

HSV: herpes simplex

PCR: polymerase chain reaction

ANA: antinuclear antibody

A1AT: alpha-1 antitrypsin deficiency

MRI: magnetic resonance imaging

SLH: subcapsular liver hematoma

AAST: American Association for the Surgery of Trauma

**References**


