ACS Case Reviews in Surgery
Vol. 3, No. 2

A Life-Threatening Second Occurrence of Hemorrhagic Shock in Recurrent HELLP Syndrome and Liver Capsule Rupture

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Background	Hemolysis, elevated liver enzymes, and low platelet count make up the rare entity known as HELLP Syndrome (HELLP). This entity complicates an exceedingly scant amount of pregnancies, and due to its infrequency, diagnosis can be delayed, leading to potentially fatal complications. One of these serious complications is the development of subcapsular liver hematoma, which can rupture and result in significant hemorrhage, hemodynamic instability, hemorrhagic shock, or even exsanguination.
Summary	The case herein describes a 38-year-old female in her 29th week of gestation, with history of prior HELLP diagnosis, liver hematoma, and capsule rupture in a previous pregnancy, who developed recurrence of all three entities in her now subsequent pregnancy. This recurrence resulted in immediate intervention after the patient developed hemodynamic instability progressing to hemorrhagic shock after substantial blood loss. Her prolonged hospital course required emergent surgery by both obstetrics and acute care surgery, transfusion of multiple blood products, admission to the intensive care unit, evaluation by psychology, and several subsequent returns to the operating room.
Conclusion	Our case illustrates the life-threatening morbidities that can occur in HELLP patients complicated by hepatic hematoma with capsule rupture, and emphasizes the necessity of a high index of suspicion for the diagnosis. This case is exceptionally unique in that the patient developed recurrence of HELLP syndrome, liver hematoma, and rupture, which on literature review has been previously described only once before.
Keywords	HELLP syndrome; HELLP syndrome recurrence; subcapsular liver hematoma; Liver hematoma rupture; liver hematoma rupture recurrence

DISCLOSURE STATEMENT:
The authors have no conflicts of interest to disclose.

To Cite: Curfman KR, Dodson BD, Antony M, Morrissey SL. A Life-Threatening Second Occurrence of Hemorrhagic Shock in Recurrent HELLP Syndrome and Liver Capsule Rupture. *ACS Case Reviews in Surgery*. 2020;3(2):57–60.

Case Description

An acronym, HELLP, standing for hemolysis, elevated liver enzymes, and low platelet count, was first used to describe this syndrome by Weinstein in 1982.[1] Symptoms include abdominal pain, nausea, and vomiting, although can often be nonspecific.[2] Pathogenesis of this entity is unclear, which frequently makes the diagnosis difficult. Histopathologic findings include schistocytes, burr cells, or echinocytes on peripheral blood smears (PBS), which are diagnostic of hemolysis.[3] Also, histology often reveals intravascular fibrin deposits that lead to sinusoidal obstruction, causing: intrahepatic vascular congestion and necrosis, intraparenchymal and subcapsular hematoma, and with progressive hematoma formation, capsule rupture.[2] HELLP is esti-mated to affect only 0.5 to 0.9 percent of pregnancies, and the diagnosis is often complicated and missed, leading to a dangerous treatment delay with significant consequences.[4] We present a 38-year-old female requiring emergent oper-ative intervention after developing a recurrence of HELLP, subcapsular liver hematoma formation, and capsule rup-ture. HELLP itself is a rare entity; however, this case is exceedingly unique, as evidenced by the development of recurrent HELLP, hepatic hematoma, and capsular rupture. Prior documentation of recurrent HELLP Syndrome alone is limited, and recurrence of hepatic hematoma for-mation with capsule rupture in the setting of pregnancy has been reported only once previously. [5]

We present a 38-year-old female at 29 weeks gestation, with a history of HELLP, subcapsular hematoma formation, and hemorrhage secondary to rupture during a prior pregnancy. She presented with three days of epigastric pain, and one day of vomiting, in the setting of known HELLP recurrence. She abruptly became hypertensive with systol-ic blood pressure (SBP) 170 mmHg, followed by sudden hypotension (SBP 80 mmHg) and reported fetal distress. Given her recurrent HELLP diagnosis and concern for fetal viability, she was taken for emergent Cesarean sec-tion. Upon entry into the peritoneal cavity by obstetrics, blood was noted to have filled the entire cavity. The acute care surgery team was emergently consulted intraoperatively due to the substantial, uncontrolled intraperitoneal bleeding. The blood was evacuated, and the source of bleeding was identified at the ruptured liver capsule. Two large hepatic capsule tears were seen and located on the anterior and posterolateral aspects of the liver. Hemostasis was achieved at the anterior tear with use of argon beam and the posterolateral tear with packing.

A temporary abdominal closure (TAC) was placed, and the patient was transferred to the intensive care unit (ICU). Peri-operatively, she was transfused eight units packed red blood cells (PRBC), five units fresh frozen plasma (FFP), three packs of platelets, two units cryoprecipitate, and one gram tranexamic acid. Upon returning to the operating room (OR) the following day, slow oozing was noted at the anterior site. A cellulose polymer hemostatic agent was applied, the liver was repacked, and the TAC replaced. The cellulose polymer agent was applied via electrocautery to bond with the liver, creating a pseudo-capsule.

On hospital day three, she returned to the OR for abdominal washout and packing, where a segment of dusky bowel with questionable viability was identified and observed. In the OR the next day, the bleeding had ceased, and the bowel was run in its entirety with a resolution of the previously questionable viability. Due to severe fascial retraction, only peritoneum and skin were able to be closed. The patient returned to the ICU for extubation, pain control, and diet advancement. She experienced brief post-partum psychosis with derealization symptoms for which she was evaluated by psychology, and symptom resolution was achieved without intervention. The remainder of her stay was uncomplicated, with transfer to the maternity unit on day eight and discharged on day 11.

Given that the fascial layer was unable to be closed, the patient developed a large incisional hernia. Approximately ten months after the initial procedure, the patient returned to the operating room and underwent a coordinated, multidisciplinary procedure that included gynecology, general surgery, and plastic surgery. The procedure was exploratory laparotomy, lysis of adhesions, bilateral tubal ligation, right oophorectomy, and abdominal wall reconstruction with bilateral component separation and placement of a retrorectus mesh. The procedure was tolerated well without significant postoperative complications. The patient was safely discharged home on postoperative day four.

Discussion

HELLP is a unique pathology which includes hemolysis, elevated liver enzymes, and low platelets, typically in the setting of pre-eclampsia or eclampsia.[1] Diagnosis of HELLP can be difficult and often elusive due to its mysterious presentation and variability, for which it can be known as "The Great Imitator".[4] Several aspects can aid in establishing the diagnosis, the cornerstone of which is hemolysis. Hemolysis is diagnosed via elevated serum bilirubin, low haptoglobin, decreased hemoglobin, or abnormal PBS showing schistocytes, burr cells, or echinocytes.[3]

Elevated livers enzymes are the second component of the HELLP triad, though no clear criteria exist for which enzymes or elevation level is diagnostic.[3] Low platelet count is the final aspect of diagnosis. Unfortunately, once again, no clear definition of "low" in this entity has been established. The Mississippi Criteria was developed as a grading system in HELLP to predict outcomes, prognosis, recovery time, and recurrence. Per this criteria, Class 1 is defined by a platelet count <50,000/mm^3, Class 2 by a range of 51,000/mm^3–100,000/mm^3, and finally Class 3 with a count up to 150,000/mm^3.[3] Fortunately in our patient, diagnosis for this occurrence was readily apparent on this presentation due to her low platelet count (130,000/mm^3), decrease in hemoglobin and hematocrit (13.8 gm/dL to 10.0 gm/dL and 41 percent to 35 percent, respectively), elevated alanine aminotransferase (331 U/L), and elevated aspartate aminotransferase (248 U/L) and her prior recorded history of the syndrome.

HELLP predisposes both mother and fetus to a heightened risk of morbidity and mortality. It has been associated with malaise, pain, nausea, vomiting, pulmonary edema, renal failure, premature birth, fetal growth restriction, disseminated intravascular coagulation, and liver hematoma development.[2] Due to the lack of data on recurrent HELLP, there has been a wide suggested recurrence range from 3 percent to 27 percent.[6,7] Subcapsular hematoma formation and rupture occurs in approximately 1/40,000 to 1/250,000 pregnancies with a suspected higher incidence reported in HELLP.[2]

Only one other report was identified. A recurrence of subcapsular hepatic hematoma development with rupture in the setting of HELLP had occurred despite the increased hematoma formation rate noted in HELLP.[5] Due to the few reported initial cases of hematoma rupture, there is no specific, designated optimal management modality, and thus a variety of treatments have been proposed. First, it is essential to provide supportive therapy and ensure that the patient is adequately resuscitated, monitored, and symptoms controlled.[2] Then, the mandated operative intervention has been proposed for worsening pain, increasing hematoma size, infection, blood loss, or hemodynamic instability.[2]

Multiple techniques were recommended for hemostasis and repair, including embolization, packing of bleeding surfaces, collagen fleece, resection, and transplantation.[2] In our particular case, resuscitation with transfusion of blood products began intraoperatively as our patient's sudden onset hemodynamic instability was readily explained upon entry into a blood-filled peritoneal cavity. This resuscitation was continued postoperatively until her rotational thromboelastometry studies normalized.

As mentioned, the patient proceeded to the operating room, given her abrupt hypotension and concern for fetal demise. Once the surgery team was able to visualize both liver lacerations, one laceration was packed coinciding with the prior recommendations for hemostasis. The second laceration obtained hemostasis via the use of the argon beam, which though not previously described as a hemostatic recommendation in this entity, showed an appropriate response and via our report appears to be a suitable option.

Conclusion

This case is unique and necessitates presentation for multiple reasons. The patient had an initial diagnosis of HELLP followed by recurrence in her following pregnancy. More uniquely, she experienced the development of a subcapsular hepatic hematoma and capsule rupture leading to hemorrhagic shock in the previous pregnancy, and recurrence of all of these entities in this subsequent pregnancy. HELLP syndrome and recurrence are rare, though the most significant finding was recurrent hepatic hematoma with rupture resulting in shock where only one prior similar case is reported.[4–7] With the described substantial associated risks, emergent surgical intervention was undoubtedly warranted and believed to be the only appropriate intervention that allowed for the survival of both mother and fetus.

Lessons Learned

HELLP Syndrome is a serious entity that affects a limited number of pregnancies. A serious known consequence of HELLP is the formation of a subcapsular hepatic hematoma, which in its own right, harbors the potential for rupture that can lead to devastating hemorrhagic shock and possible exsanguination. The occurrence of all three pathologies in one setting is unique, with the recurrence of all three entities being so exceedingly rare, that it has only been presented one other time in the literature.[5] With the constellation of these three phenomenons recurring, we now understand and implore physicians to recognize this pathology's emergent nature and the essential need for aggressive resuscitation and achievement of hemostasis. Otherwise, there is a strong likelihood of mortality in either mother, fetus, or both.

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