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Case Report

Severe HELLP Syndrome Complicated by Multiorgan Impairment and DIC: The Role of Drain Placement: A Case Report

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Abstract

Background: Disseminated intravascular coagulation (DIC) can present postpartumly in women with hemolysis, elevated liver enzymes, low platelets (HELLP) syndrome. This report highlights the importance of drain insertion following emergency cesarean section in anticipation of DIC. This would avoid the need of an open exploratory laparotomy procedure. ***Case:*** At 34 weeks of gestation, a P1G4 woman was diagnosed with severe pre-eclampsia and HELLP syndrome and her baby with intrauterine growth restriction (IUGR) and fetal distress. The woman received hydralazine, labetalol, methyl-dopa, magnesium sulphate and an emergency cesarean section was performed. A few hours later, the woman developed DIC. Red blood cells (RBC), fresh frozen plasma (FFP) and platelets (PLTs) were administered and an abdominal CT was performed, revealing intra-abdominal and anterior rectus sheath haematomas. An open exploratory laparotomy was performed to identify areas of bleeding and a drain was inserted to monitor the bleeding. The woman was closely monitored and regained haemodynamic stability. ***Conclusion:*** DIC is a common postpartum complication in women diagnosed with HELLP syndrome. Preemptive drain insertion following cesarean section in these women could avoid the need for additional revision surgery should DIC occur. Clear guidelines on the topic are lacking. ***Teaching Points:*** Postpartum surgical drain placement in women diagnosed with HELLP syndrome who later develop DIC could avoid the need of additional surgery.

Precis

Preemptively placing a surgical drain following cesarean section in patients with HELLP syndrome may avoid additional surgery in patients that go on to develop DIC.

Keywords: ***HELLP Syndrome, Drainage Placement, Multiorgan Impairment, DIC, Caesarean Section.***

Introduction

HELLP syndrome was first defined in a 1982 publication by Louis Weinstein, where he described findings of Haemolysis, Elevated Liver enzymes, and Low Platelets (HELLP) in pregnant women [1]. Currently, the most widely accepted definition of HELLP syndrome is via the Tennessee criteria, and defined as, lactate dehydrogenase (LDH) >600 IU/L, platelet count <100,000 cells/μL, and aspartate aminotransferase (AST) >70 IU/L [3,4]. On the blood smear, HELLP reveals a type of microangiopathic haemolytic anaemia (MAHA), with schistocytes and reticulocytes. Other important findings include: low haptoglobin, low fibrinogen, very high D-dimer, high bilirubin, and increased blood urea nitrogen/creatinine ratio [2]. Clinically, HELLP presents with non-specific findings of nausea, vomiting, epigastric and right upper quadrant pain, headache, blurry vision, and jaundice [3].

HELLP syndrome develops in 10-20% of women who have severe pre-eclampsia during pregnancy [5]. Pre-eclampsia is defined as new onset hypertension and proteinuria after 20 weeks of gestation in a previously normotensive woman and in its severe form can lead to MAHA and thrombocytopenia. Both pre-eclampsia and HELLP syndrome are conditions of pregnancy and should resolve following prompt delivery of the foetus and further treatment to stabilize the patient. Complications of HELLP for the mother can be severe, and include: placental abruption, subcapsular haematoma of the liver, hepatic rupture, disseminated intravascular coagulopathy (DIC), pulmonary oedema, acute respiratory distress syndrome (ARDS), and acute kidney injury (AKI). Complications for the foetus are termination of pregnancy and foetal growth restriction (FGR) [6,7].

Approximately 15-38% of HELLP patients will develop DIC as a complication. Due to the blood profile of HELLP patients, there is evidence to suggest that following a caesarean section, a drain should be placed in anticipation of DIC. Patients with thrombocytopenia are at an increased risk of subfascial and wound haematoma, following caesarean. It is left to the surgeon's discretion

to place a drain following laparotomy, as there is no standardized risk score to use as assessment.

Here we present a case of a woman who was diagnosed with severe pre-eclampsia complicated by HELLP syndrome. Whilst the foetus was successfully delivered and the mother hemodynamically stable at the time of delivery, a few hours later she deteriorated due to the development of intra-abdominal bleeding because of DIC. A subsequent open exploratory laparotomy was performed, which led to the removal of intrabdominal haematomas and restoration of haemeostasis. This surgery could have been avoided if a surgical drain had been inserted at the time of the caesarean section. We would like to emphasize the importance of having a high index of suspicion of DIC developing as a complication of HELLP syndrome, and to consider drain insertion following delivery to avoid further surgery.

Case Study

A 41-year-old, Para 1 Gravida 4, woman presented to Limassol General Hospital at 34/40 weeks gestation with severe right upper quadrant and epigastric pain. She had no prior medical history apart from one previous caesarean section. Her antenatal history was unremarkable. Upon being admitted to the Obstetrics and Gynaecology department, she underwent a full clinical examination, ultrasonography and blood analysis. She had a blood pressure (BP) of 190/90 mmHg in both arms, a pulse of 90, and normal oxygen saturations. Apart from the severe abdominal pain, she also had a headache and visual blurring which began four weeks prior, and increased in severity up until admission. Speculum exam revealed an undilated cervix with no vaginal bleeding. Ultrasonography estimated a foetal weight below the 5th centile, roughly 1,500 grams, indicating intrauterine growth restriction (IUGR). There was oligohydramnios and abnormal blood flow on the Doppler studies. The Cardiotocography (CTG) showed no baseline variability, with episodes of bradycardia lasting 2 minutes each, indicating foetal distress. Schistocytes were present on the blood smear, indicating haemolysis. There was borderline haemoglobin (12,3 g/dL), low platelets (65,000/µl), elevated uric acid (6.9 mg/dL), elevated potassium (5,4mmol/l), elevated LDH (5896 U/L), elevated AST (1934 U/L), and elevated ALT (1911 U/L). Creatinine level was 0.66 mg/dL and urea 28 mg/dL. Total bilirubin (TBil) was 0,53mg/dL, total protein in urine (14560 mg/L). INR (0.83), while APTT and fibrinogen were in the normal range. A diagnosis of severe preeclampsia with haemolysis, elevated liver enzymes, and low platelets (HELLP) syndrome was diagnosed. Antihypertensive medication (Hydralazine 5mg every 10 minutes titrated intravenously, labetalol 200mg per os (po) and methyl-dopa 500mg po), magnesium sulphate (6g in 250 mL of normal saline for a duration of 20 minutes), spinal anaesthesia and fluids were administered and an emergency Caesarean Section was performed. During the operation, the patient's BP was 145/92 mmHg, the urine output was adequately maintained and the patient had a normal consciousness level. The Caesarean section was a success, with haemostasis occurring and an IUGR male weighing 1540 grams was delivered.

The patient was transferred to the Obstetric ward after the operation, where her vital signs, and fluid balance were closely monitored. IV hydralazine, PO labetalol and magnesium sulfate were continued. Despite the good regulation of blood pressure and adequate fluid resuscitation, the patient started deteriorating after two hours. A reduction in her consciousness level was noted, her urine output was less than 15ml/h, her platelet level decreased rapidly and coagulation times were affected. The patient was admitted immediately to the ICU for closer monitoring and possible intubation due to reduced consciousness level. Intubation took place as the patient developed respiratory distress. Red blood cells (RBC), fresh frozen plasma (FFP) and platelets (PLTs) were administered as the levels had dropped. There was no urine output despite adequate fluid resuscitation. Due to the very rapid reduction in haemoglobin, and haemodynamic instability, a CT scan was performed, which revealed intra-abdominal and anterior rectus sheath haematomas. A decision was made to perform an exploratory open laparotomy and the patient was transferred to the operating theatre again. Blood clots were removed and oozing from the surgical surface of where the Caesarean section had previously been performed was identified. The oozing was corrected with FFP and PLT transfusion. Abdominal and post subfascial drains were put in place and the patient was transferred to the ICU for further hospitalization.

During her 9-day hospitalization in the ICU, the patient developed renal failure. The peak creatinine level recorded was 7.11 mg/dL and the potassium level was 6.5 mmol/L, necessitating haemodialysis. INR, APTT and fibrinogen levels were also affected, with the highest INR recorded at 1.40, indicating disseminated intravascular coagulation (DIC). Due to a decline in her haemoglobin levels (lowest levels recorded 6 g/dL) and platelet levels (lowest level recorded was 16000/µl) as a result of further abdominal oozing into the drain, the patient was transfused with 10 units of red blood cells (RBC), 3 units of fresh frozen plasma (FFP) and 6 units of platelets (PLTs). The patient gradually regained haemodynamic stability, and no longer needed mechanical ventilation, drain placement or haemodialysis. Creatinine, bilirubin, haemoglobin, platelet levels and coagulation times were all within their normal ranges.

After the 9th day, the patient was transferred to the Gynaecology ward. Antihypertensive treatment was continued (amlodipine po, labetalol po and hydralazine po), and the patient was closely monitored with daily recordings of creatinine and blood pressure. During her stay, the patient's BP was well maintained, however the patient started experiencing multiple episodes of diarrhoea, as well as a daily tension type headaches. Faecal cultures were taken and were negative for Clostridium toxins A and B. It is possible that the headaches and diarrhoea were a result of administration of hydralazine, as both symptoms disappeared upon cessation of this medication. The patient was discharged after 9 additional days in the Gynaecology ward in a hemodynamically stable condition, with a normal FBC and chemical panel. Two of the three anti-hypertensives were to be continued following discharge (labetalol 200mgX3 and amlodipine 10mgX1 daily), the patient was asked to measure her BP on a daily basis and was referred to a cardiologist for further changes to her medication.

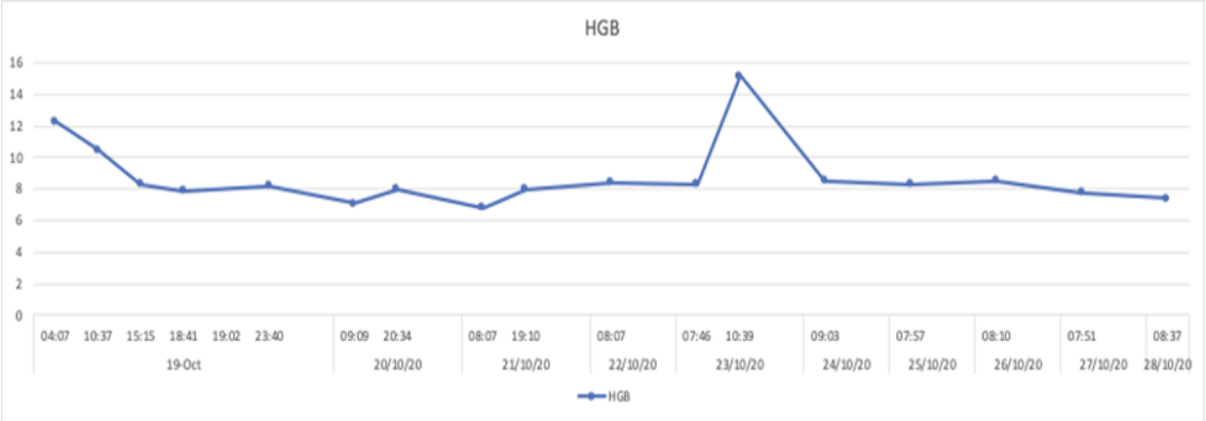

Diagram 1: Haemoglubin Levels

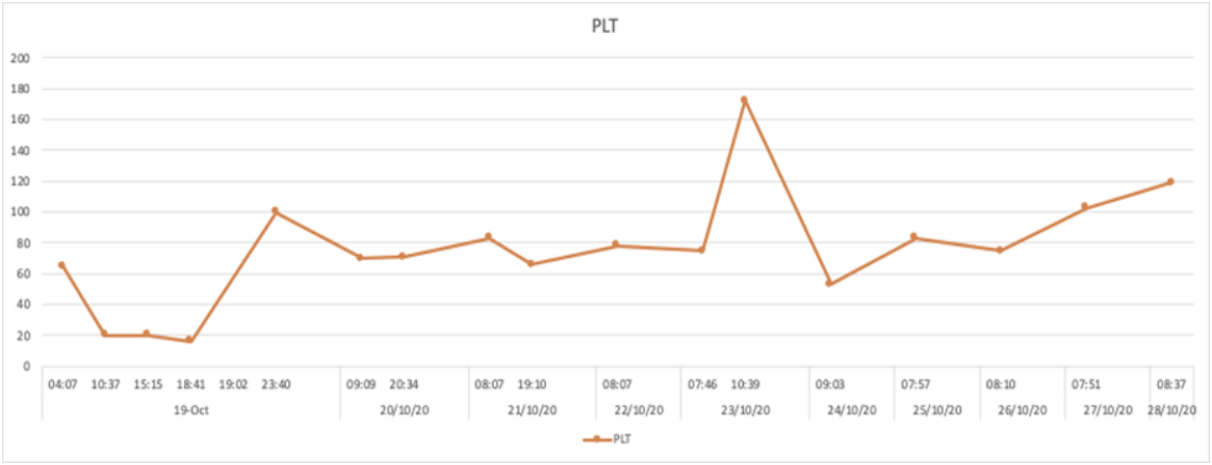

Diagram 2: Platelets Levels

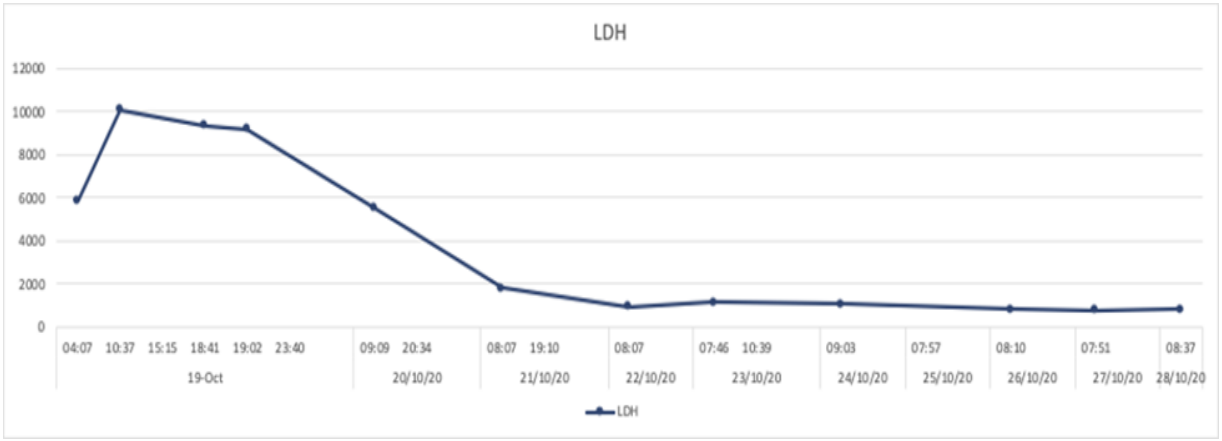

Diagram 3: LDH Levels

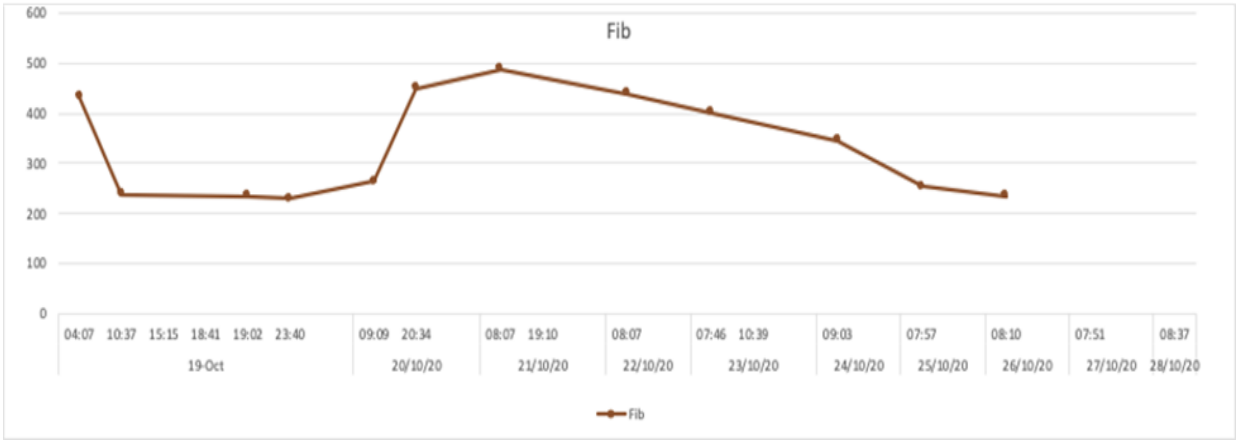

Diagram 4: Fibrinogen Levels

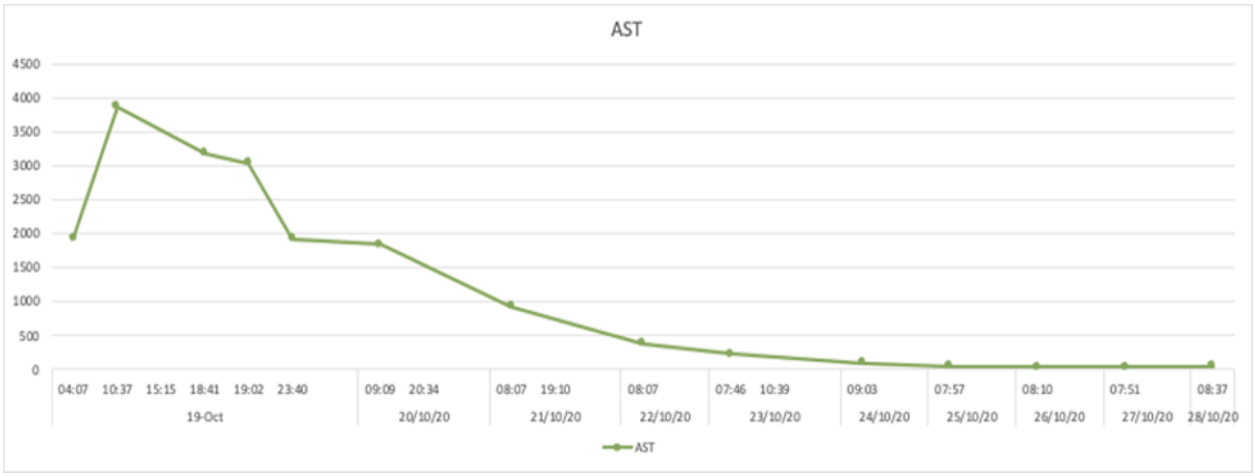

Diagram 5: AST Levels

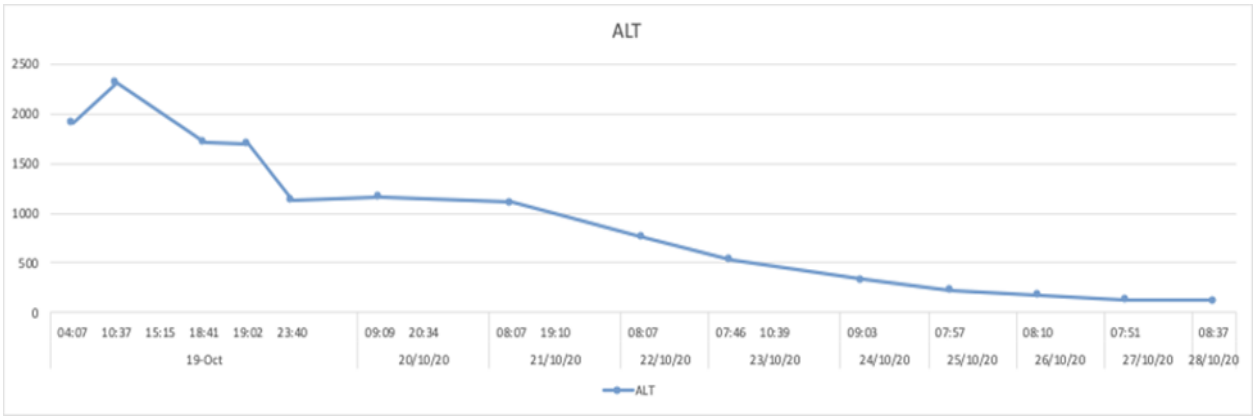

Diagram 6: ALT Levels

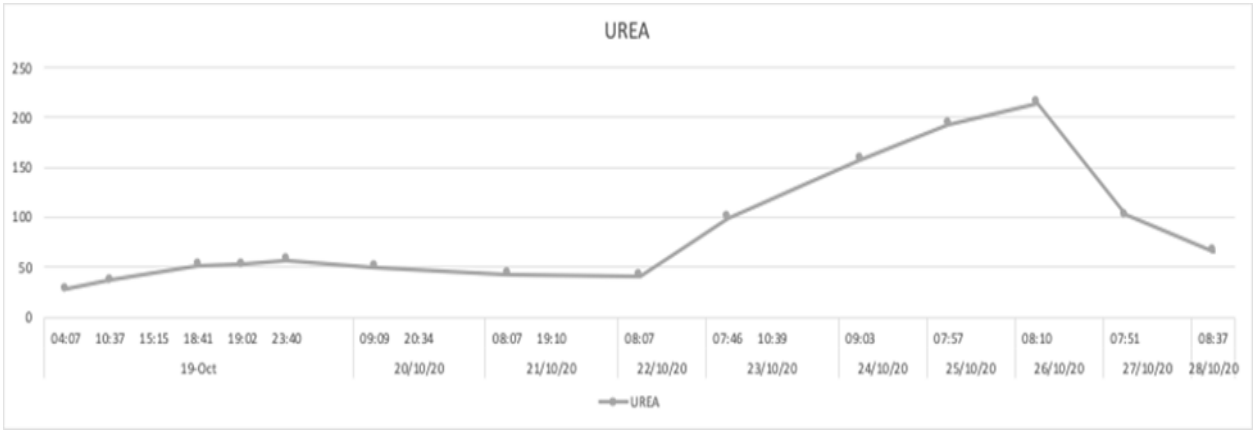

Diagram 7: Urea Levels

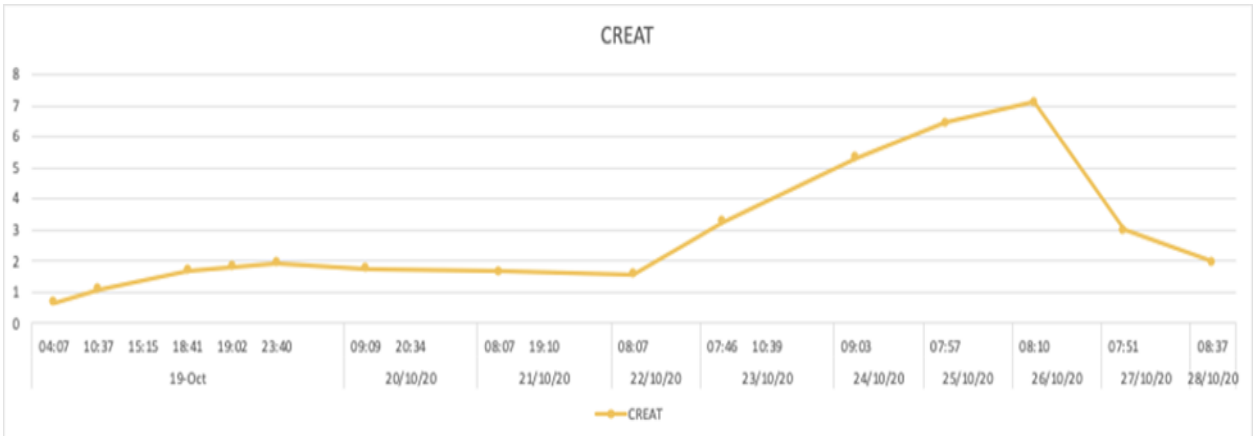

Diagram 8: Creatinine Levels

Discussion

Following a diagnosis of HELLP, it is important for the clinician to assess the severity of the condition in the patient. The percentage increase of liver enzymes coupled with the percentage decrease of platelets and haemoglobin is a useful tool to predict the patient's future haemodynamic instability. It is expected for laboratory values to worsen in the first 48 hours following delivery. In severe cases, platelets may fall to below 20 x 10^3 /μL, with an associated rise in LDH levels, as was seen in our patient. These levels are expected to normalize by the fourth postpartum day. Increased haemolysis is a major risk factor for the development of acute kidney injury. As a result, urea and creatinine levels should be monitored and referral to a nephrologist may be needed for further treatment with haemodialysis. Thankfully, the literature suggests that there is no long-term effect on renal function in patients who develop AKI following HELLP. For patients who progress to DIC, the recovery may take longer, and ICU admission is advised.

The number of HELLP patients that will develop DIC is significantly large enough to justify the need for revised guidelines regarding drain placement following caesarean in this patient population. Some studies propose for surgeons to close the incision with staples, as a means of providing quick access to the site if a haematoma forms. Previously a method has been described to apply a subfascial drain following caesarean and leaving the skin incision open for 48 hours. However, both of these methods necessitate a revision surgery, should the patient start to deteriorate. Roughly 2% of HELLP patients will proceed to laparotomy as a result of intra-abdominal bleeding. Several studies have indicated the benefits of prompt drain insertion as a way of avoiding an exploratory laparotomy when there is indication of intra-abdominal bleeding or ascites.

Crucially, to the author's knowledge, there is no report of drain placement following caesarean, where haemostasis is achieved during the operation. By placing a drain in anticipation of DIC in HELLP patients, surgeons can potentially avoid the need for revision surgery and an exploratory laparotomy. Drain placement following caesarean will also provide important information on fluid requirements. Clinicians will be able to better calculate the amount of blood to transfuse and potentially avoid transfusion-related acute lung injury. In other regards, patient management would remain the same with emphasis on obtaining haemodynamic stability.

In conclusion, HELLP syndrome is a serious obstetrics presentation which can lead to several life-threatening complications. The percentage of HELLP patients that proceed to DIC should be taken into account when forming a treatment plan. Drain placement can be considered in HELLP patients following caesarean section to try and avoid the need for revision surgery should DIC occur.

Abbreviations

HELLP syndrome: Haemolysis, Elevated Liver enzymes and Low Platelets syndrome
DIC: Disseminated Intravascular Coagulation
P1G4: Para1 Gravida4
IUGR: Intrauterine Growth Restriction
RBC: Red Blood Cells
FFP: Fresh Frozen Plasma
PLTs: Platelets
CT: Computed Tomography
CS: Caesarean Section
LDH: Lactate Dehydrogenase
AST: Aspartate Amino Transferase
ALT: Alanine Amino Transferase
MAHA: Microangiopathic Haemolytic Anaemia
ARDS: Acute Respiratory Distress Syndrome
AKI: Acute Kidney Injury
FGR: foetal growth restriction
BP: Blood Pressure
CTG: Cardiotocography
TBil: Total bilirubin
INR: International Normalized Ratio
APTT: Activated Partial Thromboplastin Time
IV: Intravascularly
PO: Per Os

Ethical Approval and Patient Consent for Publication

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Running title

HELLP complicated by DIC, role of drain (40 characters)

Availability of supporting data

Data are available from the corresponding author upon reasonable request.

Competing Interest

None

Author contributions

The authors confirm contribution to the paper as follows:

Study, conception and design: Xenophon Bazoukis, Ioannis Kazakos
Data collection: Xenophon Bazoukis, Alkistis Victoros Khristianov, Giannis Pavlides, Athanasios Chasiotis
Analysis and interpretation of results: Xenophon Bazoukis, Ioannis Kazakos
Draft manuscript preparation: Xenophon Bazoukis

All authors reviewed the results and approved the final version of the manuscript.

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Not Applicable

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