Review Article

Hemolysis, Elevated Liver Enzyme, and Low Platelet Syndrome in Pregnancy

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Abstract

Hemolysis Elevated Liver Enzymes and Low Platelets (HELLP) syndrome is a life threatening disorder, associated with substantial maternal and perinatal morbidity and mortality. HELLP syndrome occurs in 0.5 to 0.9% of pregnancies and 70-80% of cases coexist with pre-eclampsia. It occurs in the later stage of pregnancy as a complication of severe preeclampsia or eclampsia. The syndrome itself is manifested by hemolytic anemia, increased liver enzymes, and decreasing platelet counts with neurological manifestations such as hemorrhagic stroke or subarachnoid hemorrhage. Almost one half of women with HELLP syndrome have activation of coagulation factors and platelets precipitates disseminated intravascular coagulation, which may cause multiorgan failure. Rapid clinical assessment of maternal and fetal condition, early diagnosis & management of the problems is needed to minimize adverse maternal and fetal outcomes.

Key words: DIC, HELLP Syndrome, Pregnancy

Introduction

Hemolysis Elevated Liver Enzymes and Low Platelets (HELLP) syndrome is a life threatening complication in pregnancy. It was first described by Weinstein in 1982, as a multisystem disorder usually seen in the third trimester and in the postpartum period within 48-72 hours following delivery (Mallesara et al., 2016 ; Güven et al., 2012). Hemolysis occurs as a result of the fragmentation and distortion of erythrocytes during passage through small damaged blood vessels. This may result in low red blood cell level or anemia, a condition in which the blood doesn't carry enough oxygen to the body (Missmolls, 2016; Moore, 2017; McKinney et al., 2013). Liver enzyme levels increase when hepatic blood flow is obstructed by fibrin deposits. Inflamed or injured liver cells leak high amounts of certain chemicals, including enzymes, into the blood. Hyperbilirubinemia and jaundice may occur as a result of liver impairment. Low platelets levels are caused by vascular damage resulting from vasospasm, platelets aggregation at sites of damage resulting systematic thrombocytopenia (McKinney et. al., 2013: Moore, 2017).

HELLP Syndrome develops in approximately 0.5 to 0.9% of all pregnancies and in 10–20% of women with severe preeclampsia. Among them, around 70% of the cases develop HELLP syndrome before delivery and the majority developed the syndrome between 27^{th} to 37^{th} weeks of gestation and remaining (30%) develops after delivery within 48 hours (Haram, Svendsen, & Abildgaard, 2009) elevated liver enzymes and low platelet count occurring in 0.5 to 0.9% of all pregnancies and in 10–20% of cases with severe preeclampsia. The present review highlights occurrence, diagnosis, complications, surveillance, corticosteroid treatment, mode of delivery and risk of recurrence.\n\nMethods\nClinical reports and reviews published between 2000 and 2008 were screened using Pub Med and Cochrane databases.\n\nResults and conclusion\nAbout 70% of the cases develop before delivery, the majority between the 27th and 37th gestational weeks; the remainder within 48 hours after delivery. The HELLP syndrome

may be complete or incomplete. In the Tennessee Classification System diagnostic criteria for HELLP are haemolysis with increased LDH (> 600 U/L. The prominent symptoms are pain in the right upper quadrant of abdomen, the lower chest or epigastric region due to liver distention. The other features are nausea, vomiting and severe edema (McKinney et. al., 2013).

The exact cause of HELLP syndrome is not yet identified. However both preeclampsia and HELLP syndrome have their origin in the placenta (Haram, Mortensen, & Nagy, 2014). Immunological maladaptation is the most probable cause while trophoblastic invasion during fetal development (Abildgaard & Heimdal, 2013). A previous HELLP syndrome in pregnancy is associated with an increased risk as well as pre-eclampsia in subsequent pregnancies (Rezai et al., 2017). Advanced maternal age (above 30), obesity, poor diet, history of pre-eclampsia and diabetes are considered as the risk factors of HELLP syndrome (Cunningham, 2014; Moore, 2017). The prognoses of HELLP syndrome in Infants and fetus have higher mortality (6-36%) than mothers (1-3%) (Moore, 2017)

Diagnostic Criteria & Classification of HELLP Syndrome
According to Mississippi & Tennessee Classification HELLP Syndrome

Mississippi classification	Tennessee classification
Class 1	**True or Complete**
• Platelets < 50,000/mm^3	• Platelets < 100,000/mm^3
• AST or ALT > 70 IU/L	• AST > 70 IU/L
• LDH >600 IU/L	• LDH >600 IU/L
Class 2	**Partial or Incomplete**
• Platelets =50,000-100,000 /mm^3	Severe preeclampsia with any one or two of above
• AST or ALT > 70 IU/L	
• LDH >600 IU/L	
Class 3	
• Platelets=100,000-150,000/mm^3	
• AST or ALT >40 IU/L	
• LDH >600 IU/L	

(AST: Aspartate Transaminase; ALT: Alanine Transaminase; LDH: Lactate Dehydrogenase)

(Haram et al., 2009; Rahaman, 2017; Satpathy et al., 2009)

Clinical Manifestations

Malaise, epigastric or right upper quadrant pain, nausea and vomiting and nonspecific viral flu like symptoms.

Hypertension and proteinuria may be absent or slightly increase.

Excessive weight gain and generalized edema precede the syndrome in more than 50% of the cases (Haram et al., 2009) elevated liver enzymes and low platelet count occurring in 0.5 to 0.9% of all pregnancies and in 10–20% of cases with severe preeclampsia. The present review highlights occurrence, diagnosis, complications, surveillance, corticosteroid treatment, mode of delivery and risk of recurrence.\n\nMethods\nClinical reports and reviews published between 2000 and 2008 were screened using Pub Med and Cochrane databases.\n\nResults and conclusion\nAbout 70% of the cases develop before delivery, the majority between the 27th and 37th gestational weeks; the remainder within 48 hours after delivery. The HELLP syndrome may be complete or incomplete. In the Tennessee Classification System diagnostic criteria for HELLP

are haemolysis with increased LDH (> 600 U/L.

Headache, visual changes, bleeding, ascitis, jaundice, shoulder and neck pain and pulmonary edema (Satpathy et al., 2009).

In the postpartum period the HELLP syndrome usually develops within the first 48 hours in women who have had proteinuria and hypertension (Satpathy et al., 2009).

Management

Because of progressive nature of disease, patient should be hospitalized with bed rest and care should be provided under close supervision to prevent deterioration of maternal and fetal condition (Haram, Mortensen, & Nagy, 2014). After assessment and stabilization of maternal status, the fetus is evaluated by fetal heart rate tracing, biophysical profile and doppler studies, which helps to determine when delivery is appropriate (Dusse et al., 2015). If mother and fetus both are stable and gestational age is less than 34 weeks it is better to delay delivery for 24-48 hours for corticosteroid administration. Immediate delivery is indicated if gestational age is above 34 weeks or earlier in nonreassuring fetal status or complication of HELLP syndrome (multiorgan dysfunction, DIC, abruptio placenta, renal failure, pulmonary edema, liver infarction or hemorrhage etc) are already present (Rahaman,2017).

If the fetus is < 24 weeks, expectant management is to extend period of gestation as much as possible. However, such prolongation of management may not improve perinatal outcome. In this condition following care should be provided to the women: bed rest, control of blood pressure, administration of magnesium sulphate, use of antithrombotic and steroid and plasma volume expanders (fresh frozen plasma, crystalloid, colloids, etc) (Mallesara, Kanta, & Shivappa, 2016).

Vaginal delivery is preferred if women is after 32 weeks of gestation and in active labour. Induction or augmentation of labour with oxytocin or prostaglandins is acceptable if needed. In the women of gestation less than 30weeks with unfavorable cervix and in the absence of active labour cesarean section (CS) is preferred for delivery. If there is fetal growth retardation or oligohydraminous elective CS is recommended (Haram et al., 2009).

Magnesium sulphate should be administered intrapartum and early postpartum for seizure prophylaxis regardless of blood pressure. Continuous monitoring of serum creatinine to identify compromised renal function is required. As in patients with severe preeclampsia, antihypertensive are used for systolic blood pressures above 160 mm Hg and or diastolic pressures of more than 105 mmHg to avoid intra cerebral bleeding (Rahaman, 2017).

Nursing Management

Careful assessment of mother should be done and arrangement of intensive care facilities should be made.

Antiseizure prophylaxis (magnesium sulfate) to the patient.

Control of the blood pressure of the patient (antihypertensive as needed).

Avoidance injuring of the liver by abdominal palpation. Sudden increase in the intrabdominal pressure could lead to the rupture, leading to maternal and fetal mortality.

Management of prescribed fluid replacement accurately to avoid worsening the woman's reduced intravascular tone. However, excessive fluid administration could lead to pulmonary edema or ascites.

Continuous fetal monitoring for early identification of fetal compromise

Continuous monitoring of maternal heart rhythm by ECG

Continuous monitor the oxygen level in blood by pulse oximetry

Transfusion of fresh-frozen plasma or platelets as ordered to improve the platelet count.

Provision of needed care when transporting the woman.

Psychosocial support.

Complications

Maternal: hepatic rupture, acute renal failure, pulmonary edema, ascites, pleural effusion, abruptio placenta, postpartum hemorrhage, disseminated intravascular coagulation, stroke and death.

Fetal and neonatal: perinatal death, IUGR, preterm delivery. neonatal thrombocytopenia & respiratory distress syndrome (RDS) (Abildgaard & Heimdal, 2013)elevated liver enzymes, and low platelet count.

Conclusion

Hemolysis Elevated Liver Enzymes and Low Platelets (HELLP) syndrome is a life-threatening obstetric complication considered as a severe form of preeclampsia involving haemolysis, thrombocytopenia and liver dysfunction. Both HELLP and pre-eclampsia occur during the later stages of pregnancy, and sometimes after childbirth. Perinatal mortality is associated with early gestational age and its complication. Early diagnosis by assessing symptoms and laboratory test and treatment by multidisciplinary team is crucial to prevent maternal and perinatal complications.

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