

HELLP Syndrome: Early Diagnosis Alleviates Complications in Primigravida

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Abstract

HELLP syndrome is a multisystem disorder, characterized by thrombocytopenia, hemolytic anemia and liver dysfunction considered as a consequence of microvascular endothelial activation and cell injury. The chief presenting features include malaise, nausea, vomiting, epigastric and right upper abdomen pain and edema etc. It is frequently associated with severe preeclampsia or eclampsia. The aim of reporting this case is to emphasize the importance of early diagnosis and rapid treatment in these patients which in turn save lives and alleviate fatal complications. Since HELLP syndrome is associated with 0.5-0.9% of all pregnancies, early diagnosis and treatment warrants high index of suspicion. We herein report a case of HELLP syndrome in 23-years-old primigravida with pregnancy induced hypertension (PIH), who was managed successfully with caesarean section.

Keywords: Pregnancy induced hypertension; Pih; HELLP syndrome; Hypertension; Steroids; Lscs; Caesarean section; Primigravida; Maternal complication; Thrombocytopenia; Hemolytic anemia; Liver dysfunction

Introduction

HELLP syndrome was first described by Weinstein in 1982 [1]. The HELLP syndrome is a serious complication in pregnancy, defined as hemolysis (H), elevated liver enzymes (EL) and low platelet count (LP) occurring in 0.5 to 0.9% of all pregnancies and in 10-20% of cases with severe preeclampsia. HELLP syndrome mostly occurs in the third trimester, although a 5 to 8% of cases occur within 72 hours after delivery [2]. The Etiopathogenesis of HELLP syndrome is not completely understood but it should not be considered as a variant of disseminated intravascular coagulation (DIC), even if microangiopathic hemolytic anemia is characterized for both disorders because the prothrombin time (PT), the partial thromboplastin time (PTT), and serum fibrinogen levels are normal in HELLP syndrome whereas are usually altered in DIC [3]. Presenting features include nausea, vomiting, and epigastric and right upper quadrant pain 30%-90% of patients, headache in 33%-68%, visual changes in 10%-20%, and jaundice in 5% [4-7].

Case Presentation

A 23-years-old non-smoker, non-alcoholic, non-vegetarian primigravida presented with sudden onset of lower abdomen pain with decreased fetal movements. On examination, per abdomen uterus was 35-36 weeks, relaxed with cephalic presentation. Fetal movements were present but decreased. Fetal heart sound (FHS) was 140/minute and was present on right side. On cardiotocography (CTG) baseline fetal heart rate (FHR) was 130-140 with presence of beat to beat variation. Two accelerations on 155 and 160 suggested reactive CTG. Per vaginal findings were normal. There was no bleeding or leaking. Pulse rate was 86/min, Blood pressure in lying posture was 140/92 mmHg,

respiratory rate was 18/min, oral temperature was 98.60F, SPO₂ was 99% in room air. Other systemic examinations were normal. Ultrasonography with Color Doppler examination of abdomen revealed single live intrauterine pregnancy with cephalic presentation with relatively less amniotic fluid index (AFI) with normal fetoplacental and uteroplacental circulation. The placenta was posterior with grade-II maturity. There was single loop of cord around neck present. ECG was normal. Hemoglobin was 12.3 gm/dL. Total leukocyte count was 10900/cumm with normal differential counts. Platelet count was low i.e. 1,20,000/cumm. ALT, AST, Alkaline Phosphatase and serum uric acid were 70.0 U/L, 63.0 U/L, 300.6 U/L and 7.8 mg/dL respectively. Urine examination revealed passage of albumin 2+(75 mg/dL). Serum LDH was highly elevated (779 U/L) indicating hemolysis.

Depending on patient's general condition especially primigravida with PIH, elevated liver enzymes, low platelet count and signs of hemolysis, the diagnosis of HELLP syndrome was made and alpha methyl dopa 500 mg thrice daily per oral was started. The patient was further referred to the tertiary center for standard care. Follow-up of the patient revealed that she had again developed lower abdomen pain with severely decreased fetal movements after five days. Her total leukocyte counts were severely raised to 14800/cumm with neutrophilia (83%). She had then undergone emergency caesarean section with a delivery of healthy baby weighing 2165 gms. Her postoperative period was uneventful and she was discharged after 8 days of hospitalization with tablet Labetalol 100mg thrice daily per oral with other supporting medicines.

Discussion

HELLP syndrome is a life threatening condition, commonly associated with the severe preeclampsia or eclampsia. The pathogenesis of this syndrome is still obscure. In preeclampsia, defective placental vascular remodeling during weeks 16-22 of pregnancy with the second wave of trophoblastic invasion into the decidua results in inadequate

placental perfusion. Agatasa et al. suggested that there is an abnormal expression of cell adhesion molecules, as well as of the endothelial cell growth factor and its receptor in the trophoblast, causing an uteroplacental vascular insufficiency that will result in an abnormal release and metabolization of nitric oxide, prostaglandins and endothelin in the placental tissue. These changes induce platelet aggregation, endothelial dysfunction and arterial hypertension [8]. The activation of the coagulation cascade causes consumption of platelets due to adhesion onto a damaged and activated endothelium, in addition to microangiopathic hemolysis caused by shearing of erythrocytes as they traverse through capillaries laden with platelet-fibrin deposits. Multiorgan microvascular injury and hepatic necrosis causing liver dysfunction contribute to the development of HELLP syndrome [9-13]. Complications include cerebral hemorrhage, tubular necrosis and cortical ischemia in kidney, diabetes insipidus, hepatic hemorrhage and hematoma. Perinatal infantile mortality varies between 6.7 and 70%. It is caused by the premature detachment of the normally inserted placenta, intrauterine asphyxia, and prematurity. About 60% of fetuses die intrauterinely, 30% show intrauterine growth retardation, and 25% thrombocytopenia [3]. Women with a history of HELLP syndrome carry an increased risk of at least 20% (range 5-52%) that some form of gestational hypertension will recur in a subsequent gestation. Whereas thrombophilia screening is accepted for patients with prior-severe, acute onset pre-eclampsia, one should always consider screening of a number of particular mutations in certain subunits of the complement system, and its regulatory proteome [2].

Conclusion

High index of suspicion is of paramount importance to deal with the patients of HELLP syndrome because early diagnosis and proper management of these cases may save complications and lives of both mother and fetus as was shown in our case.

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