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ORIGINAL ARTICLE

HELLP or Help: A Real Challenge

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Abstract

Objectives To ascertain the prevalence, presentation, diagnosis, severity, and complications of HELLP syndrome.

Materials and Methods This is a prospective observational study analyzing the conditions and the data of 24 cases of HELLP syndrome in a tertiary care hospital. The analysis was done for the demographic characteristics, presentation of these patients, complications associated, and the perinatal outcome.

Results 0.45 % of the patients admitted for delivery developed HELLP syndrome. Majority of the patients developed the condition in 30–36 weeks period of gestation, while five patients developed it in the postpartum period. The condition led to 12.5 % of maternal and 45.8 % of perinatal mortality.

Conclusion HELLP syndrome is an important cause for maternal and perinatal morbidity and mortality.

Keywords HELLP Syndrome · Severe pre-eclampsia · Maternal mortality

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Introduction

The HELLP syndrome is a serious complication in pregnancy characterized by a triad of hemolysis, 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 pre-eclampsia. The syndrome was originally described by Pritchard et al. [1] in 1954, and the acronym was coined in 1982 by Weinstein [2].

The diagnosis of HELLP syndrome may become very challenging because the patients may present with vague symptoms like nausea, vomiting, headache, malaise, or flu-like symptoms. This leads to misdiagnosis of HELLP

syndrome with various mild conditions like viral hepatitis to serious life-threatening conditions like acute fatty liver of pregnancy [3, 4]. The condition is regarded as a variant or a complication of severe pre-eclampsia, and the diagnosis of the complete form requires the presence of all the 3 major components, while partial or incomplete HELLP syndrome consists of only 1 or 2 elements of the triad.

This study was undertaken with the objective to ascertain the prevalence, presentation, diagnosis, severity, and complications of HELLP syndrome and to evaluate the maternal and fetal outcome in pregnancies complicated with HELLP syndrome.

Materials and Methods

A prospective observational study was carried out from 01 Jan 2008 to 30 Jun 2010, and the analysis of the data was done on all the patients diagnosed with HELLP syndrome at a tertiary care hospital. The gestational age of the pregnancy was determined from the last menstrual period or early sonography, if available. When nothing was available, the uterine height at presentation was taken to ascertain period of gestation (POG). The diagnosis and classification of HELLP syndrome were made using the criteria established by Sibai et al. [3]—complete or partial HELLP depending on the components involved and Mississippi classification [2]—i.e., abnormal peripheral blood smear, raised lactic dehydrogenase (LDH) (>600 U/L), elevated total bilirubin (>1.2 mg %), elevated liver enzymes [increased plasma aspartate amino transferase (AST) >70 U/L], and low platelets (platelet count <100,000/cmm). Other associated symptoms in the mother and the perinatal outcome in terms of the IUFD, preterm delivery, stillbirths, and NICU admissions were also noted.

The clinical data included were value of blood pressure, pulse, respiratory rate, and urinary output, while the laboratory evaluations recorded were serial measurement of complete blood cell count, coagulation profile (PT, APTT), liver function tests (S Bilirubin, ALT, AST, and LDH), and renal function tests (B Urea, S Creatinine, and S Uric Acid). Abdominal ultrasonography was done when a subcapsular liver hematoma was suspected. The presence of acute renal failure and pleural effusion were utilized as determinants of clinical status. Disseminated intravascular coagulation (DIC) was defined as the presence of low platelets, low fibrinogen, prolonged PT and APTT, and raised fibrinogen degradation products or D-dimer.

A central venous catheter was inserted to aid in hemodynamic management, when required, and the patient was shifted to ICU if she required mechanical ventilation or was hemodynamically unstable despite the treatment. Inj Magnesium sulfate as per Pritchard's regimen was used for seizure prevention and control. The diastolic BP was maintained between 90 and 100 mmHg using Inj and oral labetalol, amlodipine, α-methyldopa, or a combination of these drugs. Blood and blood products were used to correct coagulation abnormalities or anemia as needed. Oliguria was initially managed using colloid plasma substitute fluid challenges, and if there was no response, infusion of dopamine and frusemide was given.

Results

During the study period, there were a total of 5,307 deliveries including 642 women with hypertension in pregnancy. A total of 24 cases of HELLP syndrome were diagnosed and managed at our center.

0.45 % of the patients who were admitted to the hospital for the delivery and 3.7 % of the hypertensive pregnancies developed this life-threatening complication. The average age of the patients was 19–31 years (mean age 24.25 years). 16 (67 %) patients were admitted with HELLP syndrome at less than 36 weeks POG and of this one patient was at <28 weeks and four were between 28–30 weeks POG. Cases that were diagnosed with HELLP syndrome in the postpartum period formed a significant group with 5 cases (20 %). Two of these cases had complete HELLP and three were partial, while only three of the balance nineteen developed complete HELLP. Thus, complete HELLP was seen more often in the postpartum period. The condition was seen equally distributed in primigravidae and multigravidae, thus indicating that the condition should also be suspected in multigravidae. The mean systolic and diastolic BP in these patients were 166 ± 18.65 and 110.5 ± 12.7, respectively.

These patients had varied ways of presentation to the hospital (Table 1). Majority of them presented to the hospital with features of severe pre-eclampsia and eclampsia. Two patients developed severe PPH and were later diagnosed as HELLP syndrome. One case developed respiratory distress and was noted to have ascites and pleural effusion, while two had visual symptoms with headache 6–8 h after the delivery, in the postpartum patients. One patient developed subcapsular hepatic hematoma and two developed DIC during their course in the hospital. Three patients presented with acute renal failures. 13 patients were found to have abruption of the placenta. 13 patients had symptoms in the form of scotomas and episodes of temporary blindness, and on follow-up, none had a permanent damage to the vision.

Investigations of these patients showed that the platelet count was less than 50,000 per cmm in two cases, 50,000–100,000 per cmm in three cases, and the rest had a platelet count more than 100,000 per cmm. The LDH was >900 IU/L in all the cases at the diagnosis with a

Table 1 Demographic and clinical characteristics

Characteristic	Mean (±2SD)	Range
Maternal age (years)	24.25 ± 3.05	19–31
Gestational age (weeks)	32.89 ± 2.66	28–38
Parity	2.08 ± 0.89	1–4
Systolic BP (mmHg)	166 ± 18.65	138–210
Diastolic BP (mmHg)	110.5 ± 12.7	98–140
Proteinuria (1+ to 4+)	2.29 ± 1.04	1–4

Table 2 Clinical presentation

Characteristics	N (%)
Epigastric pain	16 (67)
Nausea and vomiting	17 (71)
Headache	18 (75)
Visual symptoms	13 (54)
Eclampsia	7 (29)
IUFD	9 (38)
Placental abruption	13 (54)
DIC	2 (8)
ARF	3 (12.5)
Ascites	6 (25)
Pleural effusion/pulmonary edema	3 (12.5)
Wound hematoma	3 (12.5)

Table 3 Lab characteristics

Characteristics	Mean ± SD	Range
Platelets (per cmm)	104,966.7 ± 22,595.8	44,000–142,000
AST (IU/L)	111 ± 51.44	69–315
LDH (IU/L)	1,157.75 ± 287.53	700–1,976
Total bilirubin (mg %)	4.6 ± 2.54	1.4–11.9
Urea (mg %)	30.75 ± 18.3	18–87
Creatinine (mg %)	0.98 ± 0.53	0.6–3.3

maximum of 1,976 IU/L in one case. Transaminases were also raised in all the cases, thus showing complete HELLP syndrome in five cases and partial HELLP syndrome in rest of the patients. These 24 patients required a total of 52 bags of packed RBCs, 11 units of single-donor platelet concentrates, 114 bags of fresh frozen plasma, and 47 bags of cryoprecipitate (Tables 2, 3).

Three patients succumbed to their conditions. The patient who had presented with IUFD in labor was suspected to have liver hematoma, which was confirmed on sonography. She delivered a fresh stillborn baby and developed hemodynamic instability. She was planned to be shifted from labor room to ICU when she succumbed to her condition. She had complete HELLP with LDH levels reaching 1,976 IU/l and platelet count of 88,000. The patient with eclampsia had partial HELLP as per the reports of the immediate prior to labor. She also developed respiratory complications in the form of pulmonary edema during management, and whether she died only of HELLP or HELLP plus two other complications of severe pre-eclampsia could not be ascertained. The patient with DIC had a cesarean delivery, and the intraoperative period showed oozy margins and abdomen was closed on drains. The blood parameters were of complete HELLP. USS showed intraperitoneal collection with hematomas in the abdominal wall. She was being resuscitated with blood components when she had a cardio-respiratory arrest.

There were a total of 11 perinatal deaths. Of these, 9 patients presented to our center with intrauterine fetal death and 2 died of the prematurity—one at 27 weeks and one at 31 weeks (developed RDS).

Discussion

HELLP syndrome is a poorly understood pregnancy-related condition with a rapid onset and is typically seen in patients with severe pre-eclampsia, although it can occur in the absence of pre-eclampsia in 10 % of the cases. Excessive weight gain and generalized edema precede the syndrome in more than 50 % of the cases [2, 5]. HELLP syndrome develops with a peak frequency between the 27th and 37th gestational weeks. The postpartum cases of HELLP syndrome usually develop within the first 48 h after delivery and these are at a higher risk of developing renal failure and pulmonary edema. The risk of recurrence of HELLP syndrome is 24 % after the index pregnancy [5, 6].

Various studies have reported an incidence of HELLP syndrome from 0.5 to 1 %, which is similar to our study [4–6]. But our study revealed that 20 % of the patients developed HELLP in the postpartum period. The study showed that two patients had class I and three patients had class II HELLP syndrome.

The HELLP syndrome is associated with both maternal and neonatal complications [3, 6, 7]. Nine patients were diagnosed with intrauterine fetal demise at presentation. Three patients developed wound hematoma after cesarean section, and the condition is said to be more common in patients with HELLP syndrome. There were no neurological complications associated with HELLP syndrome in this study, while Isler et al. [8] have reported serious intracerebral hemorrhage in these patients. Severe postpartum bleeding was seen in two cases and forms an important complication to anticipate in these patients. Maternal mortality, up to 25 %, has been reported with

HELLP [7–9]. Our study had maternal mortality of 12.5 %. Patients succumbed to these three complications—Pulmonary edema, liver hematoma, and DIC. The perinatal mortality rate related to the HELLP syndrome is between 7.4 and 34 %. Prematurity, placental insufficiency, with or without intrauterine growth restriction (IUGR), and abruptio placenta are the leading causes [7, 9]. This study had a perinatal mortality of 45.8 %.

To conclude, HELLP syndrome is an alarming diagnosis, which brings in soaring high maternal and perinatal morbidity and mortality. The early and timely detection of this complication in the obstetric set-up and a joint team work by the obstetrician, transfusion medicine specialist, and the anesthesiologist can work wonders for the patient.

Compliance with ethical requirements and Conflicts of interests Informed consent was obtained from all patients for being included in the study. Sushil Chawla, Ashish Marwaha and Raju Agarwal declare that they have no conflict of interest.

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