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RE-EMERGENCE OF A POST NATAL NORMOTENSIVE PATIENT FROM AN ATYPICAL HELLP SYNDROME

Gynecology

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ABSTRACT

HELLP Syndrome is a serious complication in pregnancy , characterised by hemolysis, elevated liver enzyme and low platelet count. HELLP syndrome may develop in the absence of maternal hypertension. HELLP Syndrome occurring in less than 1%(2) of all pregnancies and in 30% cases, it develop in post partum cases.

Hereby, in this study we present reports of 3 cases with HELLP syndrome in normotensive patients, complicated with mild ascites with paralytic ileus .

KEYWORDS

INTRODUCTION

HELLP Syndrome is a serious complication in pregnancy, characterised by Hemolysis, Elevated liver enzyme and Low platelet count . It probably represents a severe form of preeclampsia but relationship between two is controversial(4). The HELLP syndrome may develop in the absence of maternal hypertension(1). HELLP Syndrome occurring in less than 1%(2) of all pregnancies , and in all only 15-20%(3) of patients have HELLP Syndrome without antecedent hypertension and proteinuria(4). In 30%(5) cases, HELLP Syndrome develop in post partum stage .

Main risk factor for developing HELLP Syndrome is 'Pre eclampsia or HELLP Syndrome in previous pregnancies'.

Symptoms of HELLP syndrome include(3) –

- Nausea and vomiting(30%)
- Epigastric pain (65%)
- Malaise (90%)
- Headache(31%)

Complication of HELLP Syndrome

- DIC(20%)(6)
- Acute Renal failure(84%)(7)
- Pulmonary Edema(6%)(6)
- Liver rupture

Criteria for HELLP SYNDROME-

- UNIVERSITY OF TENNESSEE(8)(9)-

Hemolysis on peripheral smear
LDH >600 u/L
Serum aspartate transferase> 70 IU/L
Total platelet count <1 lakhs/ul

- UNIVERSITY OF MISSISSIPPI , 1999(10)

Class 1- Severe thrombocytopenia <50,000 cells/1
Class 2- Moderate thrombocytopenia (50,000- 1lakhs cells/l)
Class 3-Thrombocytopenia (1lakhs- 1.5 lakhs cells/ l)

Outcome in baby of HELLP Syndrome-

- Perinatal mortality(11)
- Preterm labour pain
- LCHAD [long chain acyl hydrogenase coenzyme A deficiency]
- Intra uterine growth retardation[12]
- Respiratory distress syndrome

Hereby, in this study we present reports of 3 cases with HELLP syndrome in normotensive patients, complicated with mild ascites with paralytic ileus .

CASE REPORT 1-23 year old patient G3P1L1A1, Hetalben Maheshbhai Solanki, with 8 MOA, with previous Caesarean section, residing at Dabhoi, was admitted to DGH for blood transfusion, as patient was severely anaemic, afebrile, pulse- 118/min and blood pressure - 104/60mm hg with HB 3.2 gm%, TLC- 3,400, Platelet- 20,000/mm3. All other investigations were normal. Tests for Dengue and malaria were negative. LDH was raised to 3689. D –Dimer index was >5000. Peripheral smear showed mild anisopokilocytosis with few elliptocytes, WBC and platelets were decreased on smear. She was a booked case. On admission, patient was injected 2 doses of steroid 24 hours, apart to accelerate fetal lung maturity.

After 2 days and 2 blood transfusions, patient started getting uterine contractions, which did not subside by tocolytics and an emergency caesarean section was performed for Foetal Distress. A preterm baby, weighing 1.6 kg., was delivered. 1 PCV was given post operatively. On 3rd POD, patient developed epigastric pain, silent abdominal distension, nausea and vomiting and a fever spike. There was no history of passage of stool. Opinion of Surgeon was sought,& an RT was inserted. Patient was kept NBM. Investigations were repeated. Haemoglobin- 6.7 gm%, TLC – 2000/mm3, Platelet -30,000/mm3, urine R/M showed presence of RBC and pus cells, S. Billirubin- 1.7mg%. X-Ray of Abdomen, was normal with no air fluid level. USG showed signs of paralytic ileus with moderate ascites. Clinically the patient was diagnosed to be in DIC, on basis of high LDH, & high bilirubin & low platelets, On successive 4 days, 4 PCV were given along with higher antibiotic (meropenem and ciprofloxacin), 2 doses of albumin, potassium and IV Fluids.

After 4 days, Haemoglobin again reduced to 6.7gm%, platelet count was 20,000, TLC- 4,000 and S. bilirubin 17.6. LDH was 1371, alkaline phosphatase was 68; both the values were significantly raised. Peripheral smear showed Schistocytes with burr cells. BP constantly remained normal. Patient also complained of breathlessness. After this, she was transferred to ICU, where she was diagnosed to a patient with HELLP syndrome, due to Pancytopenia, with progressive jaundice & paralytic ileus. 2 higher Antibiotic - Rifagut was added along with liver enzymes. 2 PCV were given. Patient was monitored and kept under observation.

After 2 days, patient started recovering. The same treatment was continued. As RT aspiration decreased, RT was removed. CT contrast pelvis showed mild collection in the peritoneal cavity. Pulse rate had settled. Investigations started returning to normal. In the entire treatment duration, patient received 9 PCV.

At the time of discharge, patients' investigations had improved results- -Haemoglobin 8 gm%, S.Bilirubin 3.6gm%, peripheral smear showed

mild anisopoikilocytosis. LDH level decreased. All other investigations returned to normal range. Patient was discharged after Stitch Removal. She was called for follow up visit after 8 days.

CASE REPORT 2- 26 yr old patient, Primigravida, Shardaben M. Pargi with 8 montths of amenorrhoea, residing at Simaliya, admitted in Dhiraj Hospital , with true labour pain with leaking per vaginum, with positive plasmodium vivax infestation as reported on a private lab.report, and found to be normal, with hospital lab. report. She had temperature 99.7 degree farenheit, pulse-116/min, blood pressure-124/88mmhg. Patient had history of fever since 7 days.

Investigations were done, Hb- 9.3gm%, TLC- 11,000/cumm, platelet count- 94,000/cumm , malaria antigen and dengue tests were negative. Rest all investigations were normal. Peripheral smear shows anisopoikilocytosis with thrombocytopenia, WBC were normal on smear, no malarial parasite was present. She was not a booked case.

Patient delivered female child , 2kg, vaginally. Delivery was uneventful. Baby was preterm according to Ballard's scoring. Baby kept in NICU under observation.

On 2nd post natal day, patient developed epigastric pain, abdominal distention, vomiting , nausea, breathlessness with decreased urine output (200ml in 12 hr). Investigations were repeated , Hb- 6.6gm%, TLC- 20,000 cumm , Platelets- 70,000/cumm, INR-1.2, Urea- 61, creatinine-1.8, serum bilirubin is 4.8(indirect -3.7 , direct- 1.1), SGOT-214, SGPT-369,Alkaline phosphatase- 468,LDH-2657, D dimer-3682. Ultrasonography showed ascitis with liver parenchymal disease, with paralytic ileus.

Medicine reference was done. They adviced to rule out hepatits A, B ,C ,E virus. All hepatitis virus tests were negative.

On basis of investigations, patient was diagnosed as HELLP syndrome.

On successive 4 days, ceftriaxone , metro , syrup LIV-52 were given. 1 dose of Vit K was given. 1 PCV was transfused. Patient was monitored and kept under observation.

After 5 days , patient started to recover. The treatment was continued. Investigations started returning to normal.

At the time of discharge, her Hb was 8.4gm%, serum bilirubin-2.3mg%, SGOT- 75, SGPT-82, LDH level decreased. All other investigations returned to normal range. Patient was discharged. She was called up for follow up visit after 15 days.

CASE REPORT 3- 22yr old patient G2P1L1, Laxmiben Amitbhai Rathodiya with 9 months of amenorrhoea residing at Pipaliya ,was admitted in Dhiraj hospital with true labour pain with severe anaemia , afebrile, pulse was 108/min and blood pressure was 124/82 mmHg, with Haemoglobin -4.9gm%, TLC-8200/cumm, Platelets-1,50,000/cumm. Rest all investigations were normal, test for malaria and dengue were negative, peripheral smear shows showed moderate aniso-poikilocytosis with few burr cells , WBC and Platelets were normal on smear. She was unbooked case.

After 1 unit of blood transfusion , emergency caesarean section was performed for fetal distress with previous caesarean section, male child of 2.1 kg was delivered who was kept on ventilator for severe birth asphyxia with meconium aspiration syndrome, unfortunately , baby died after 12hrs of extensive monitoring. 1 unit of blood transfusion was given intra-operatively to the patient.

On 2nd post op day, patient developed epigastric pain , silent abdominal distention, nausea , vomiting, breathlessness , with fever (99.9 degree farenhiet) with tachycardia (124/min) , fall in blood pressure(90/40mmHg), tachypnea(28/min), with decreased urine output (75ml in 12hr). Investigations were repeated , HB- 3.4gm%, TLC-20,000/cumm, platelets- 50,000/cumm, INR- 1.7, urine showed presence of RBC and pus cells, urea – 64, creatinine – 3.1, S. Bilirubin – 7.8, LDH-2268. Ultrasonography showed signs of moderate ascites with paralytic ileus with haemoperitoneum. Medicine and surgery reference was done for same. RT tube was inserted. For haemoperitoneum , laparotomy was done under general anaesthesia. There was haemoperitoneum, about 700ml.blood present in peritoneal cavity. No active bleeder was present and drain tube was inserted . During intraoperative period, 1 unit of PCV and 2 unit of FFP were transfused.

Post operatively, patient was shifted to ICU, she was on ventilator. On 3rd day , patient was diagnosed clinically to be in DIC, on basis of high LDH- 2849, serum bilirubin- 3.8gm%, low platelet- 30,000/cumm, urea – 68 and creatinine- 3.3, D-dimer index- more than 5000.

On successive 6 days, 6 PCV , 3 FFP were given and 8 cycles of dialysis were performed along with higher antibiotics like meropenem and vancomycin, iv fluids. Inspite of 8 cycles of dialysis , patient had urine output of around 300 cc/24 hrs .

On 10th day , tracheostomy was done , as patient was breathless.

On 11th day , investigation were repeated , Hb was reduced to 4.5gm%, TLC- 30,400/cumm, platelet count – 60,000/cumm, serum bilirubin-5.8, LDH- 1541, alkaline phosphtase- 254, urea – 107, creatinine- 3.9, INR-1.79. Patient was diagnosed as HELLP syndrome with multiorgan dysfunction syndrome due to progressive jaundice , thrombocytopenia, paralytic ileus with tachypnea with breathlessness with decreased urine output.

Her GCS (M2E1V1) was poor. Patient was monitored and kept under observation. Even with stringent monitoring, patient's condition remained same . on 16th postoperative day, patient expired.

DISCUSSION

HELLP is a multisystem disorder resulting in generalized vasospasm, micro thrombi formation, and coagulation defect. HELLP is generally associated with pre eclampsia. However, in rare cases there may be no evidence of antecedent pre eclampsia or during labour as in this pregnancy.[13]

In our study the incidence of HELLP syndrome in normotensive patients is 68/1,00,000 deliveries per year.

In all case, the first significant presenting symptom was epigastric pain and nausea with vomiting. This is similar to many studies which conclude that the first and often the most important symptom, of this syndrome is epigastric pain which is assumed to be caused by stretching of Glisson's capsule, due to sinusoidal obstruction of blood-flow.[14]

The HELLP syndrome usually occurs with preeclampsia. However, in 20% of cases, there may be no evidence of pre-eclampsia[15]. In these cases, the patients remained normotensive, during their antenatal period and post natal period. However, 2 patients out of 3 had history of mild pre eclampsia in previous pregnancy. The patients had no episode of hypertension, and all laboratory findings were normal till her second trimester of pregnancy. It was in the last trimester that patient's blood count started getting deranged.

The initial haematological report exhibited reduced haemoglobin, & Thrombocytopenia, with normal values of other L.F.T.s.

The first event of initial morbidity in all the cases, presented after 48hrs of delivery, was in the form of persistent abdominal distention, not resonding to usual line of conservative management, even though it was diagnosed as paralytic ileus in the 1st and 3rd case. The opinion of a surgeon also concurred.

The patients were first managed as a case of DIC, later they were diagnosed as Progressive jaundice with HELLP, thrombocytopenia, and DIC, due to high D-Dimer index, on accounts of laboratory investigations, and albuminuria. As per a study, patients with the HELLP syndrome, especially those with DIC, may demonstrate delayed resolution or even deterioration in the post-partum period. Therefore, the use of heparin has been proposed for patients with preeclampsia, HELLP syndrome and DIC.[15]

In another study, retrospectively analysed, revealed that in women with DIC in the post-partum period,6 of 9 patients developed post-partum bleeding including retroperitoneal haematoma. Treatment with heparin was put off to avert post-partum bleeding. Thus, most authors oppose the routine use of heparin[16]. However, in these cases, heparin was used only in the 1st case.

In most patients with a HELLP syndrome, the maternal Platelet counts continue to decrease immediately post-partum with an increasing trend on the third day [16]. About 30% of the HELLP syndromes develop after birth, the majority within the first 48 hours. However, the time of onset might range from a few hours to 7 days after delivery[17]

In all patients, there was presence of mild to moderate ascites, which was demonstrable clinically as well as on Ultrasonography . According to Woods et al. [18] in a retrospective study, the incidence of large-volume ascites in patients with HELLP syndrome who had caesarean was about 10%. They compared HELLP syndrome patients without ascites, to those with HELLP associated ascites and found six fold increase in the incidence of congestive heart failure and a nine fold increase in the incidence of adult respiratory distress syndrome. They observed a rapid deterioration in maternal condition with rise in blood pressure and increasing proteinuria[18].

The presence of HELLP syndrome is associated with an increased risk of maternal death (1 percent)[14] and increased rates of maternal morbidities such as pulmonary edema (8 percent), acute renal failure (3 percent), DIC (15 percent), abruptio placentae (9 percent), liver hemorrhage or failure (1 percent), adult respiratory distress syndrome (ARDS), sepsis, and stroke (<1 percent). [19]

In our study , maternal mortality is 1 out of 3 cases due to multi-organ dysfunction.

Fetal mortality rate is 10 to 35 percent that depends on the gestational age at delivery in HELLP syndrome . [20] This syndrome is also associated with intrauterine growth restriction and premature delivery. In our study,all 3 were preterm deliveries in which 2 babies were survived and 1 was died due to severe birth asphyxia. [20]

CONCLUSIONs

These case reports demonstrate atypical presentations of HELLP syndrome in a normotensive patient. These patient had severe anaemia who developed symptoms of this syndrome in post natal period following delivery. Their routine 2nd trimesteric antenatal work up was normal. The patient was treated under combined care (obstetrics, medicine and surgery) and 2 patients showed excellent and rapid recovery but one patient who developed multi-organ dysfunction, died.

At the conclusion, one can make a statement, that, vigilant and properly applicable therapy, with improvisations implemented as & when indicated, helped us save two lives, though we failed in one case. Well, one can't be a winner all the times, & all the way. One has to accept the limitations of any type of treatment modality, particularly in case of Multi-organ failure, as it happened with the third patient.

ETHICALAPPROVAL

A written and informed consent has been given by the patient for publication of this case.

CONFLICT OF INTEREST

There is no conflict of interest

FOOTNOTE

Based on three investigations reports, viz. Thrombocytopenia, moderately raised liver enzymes,

Raised LDH, abnormal peripheral smear, % high D-Dimer level, the condition was labelled as HELLP syndrome. But it did not clinically fit into a classical picture of HELLP syndrome. What should we label it as?

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