Successful management of severe HELLP syndrome: A case report

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Abstract

Haemolysis, elevated liver enzymes, and low platelet count (HELLP) syndrome is a severe pregnancy complication that is life-threatening for both mother and fetus. When HELLP syndrome associates with disseminated intravascular coagulation (DIC) or with other complications, it is necessary to terminate the pregnancy. We present a case of a 26-year-old primigravida with rare combination of HELLP syndrome with coagulopathy and renal complication at 34 weeks of gestation. She had emergency caesarean section followed by haemorrhagic complications, DIC and acute renal failure. During her stay in intensive care unit, she had massive blood transfusion and fresh frozen plasma. She had haemodialysis because of acute renal failure. A gradual improvement of her condition was observed after exploration and drainage of haemoperitoneum. After 13 days she was discharged from the intensive care unit.

HELLP syndrome complicated by DIC and Acute kidney injury is a condition that is associated with high maternal and perinatal morbidity and mortality. Prompt recognition, teamwork and treatment with timely administration of blood products along with other supportive care is crucial in the management of this life-threatening and challenging condition.

Key words: AKI, Preeclampsia, Disseminated intravascular coagulation, HELLP syndrome

Highland Med Res J 2020;20(1):56-60

Introduction

Hypertensive disorders in pregnancy represent a large spectrum of disorders. HELLP syndrome is a complication of the condition. HELLP is an acronym which refers to the triad of **H**aemolysis, **E**levated **L**iver enzymes and a **L**ow **P**latelet count.[1-3] HELLP syndrome is estimated to complicate 0.1% to 0.8% of pregnancies, while 10% to 20% of HELLP occur with severe pre-eclampsia.[1] Since hypertensive disorders of pregnancy are associated with an increased risk of Acute Kidney Injury (AKI), any risk factor for hypertension can be considered as risk factors for AKI.[3] Preeclampsia when severe or when associated with HELLP syndrome can lead to AKI and in the setting of HELLP syndrome alone, AKI has been reported to occur in 7–60% of patients.[2,3] In most countries, hypertensive complications of pregnancy are the leading cause of pregnancy-related AKI.[3]

The pathophysiology of HELLP is not entirely clear. There is no known precipitating cause but appears to be associated with pre-eclampsia, the latter of which is known to have pathological vascular lesions in multiple organs, such as the liver, the kidney, and the placenta.[4] Pre-eclampsia is considered to have placental insufficiency leading to inadequate oxygen delivery to the placenta and the release of mediators of endothelial injury.[5] The resultant deficiency of vascular growth factor activity may contribute to hypertenstion, proteinuria, and renal injury.[6] Pre-eclampsia is attributed to an excess of vasoconstrictor over vasodilator impacting systemic circulation which would be expected to lower the glomerular perfusion rate.[7,8] HELLP may also be associated with Disseminated intravascular coagulation (DIC), and in fact some investigators believe that DIC is the primary process that initiates the dysfunction leading to preeclampsia.[9,10] While in developing countries, preeclampsia and the HELLP syndrome are prevalent causes of DIC, the leading causes in the developed countries are placental abruption and postpartum hemorrhage.[9]

Case presentation

A 26 year old primigravida registered at the Jos University Teaching Hospital (JUTH) for antenatal care at a gestational age of 18weeks with no complaints and normal physical and laboratory findings. She had 4 subsequent uneventful follow up visits. She, however, presented in the labour ward of JUTH at a gestational age of 34 weeks with complaints of epigastric pain and frontal headache. There was history of blurring of vision, but no flashes of light or bleeding from any orifices. She felt fetal movements. She was not pale and

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not jaundiced but bleeding from her gums. She had a pulse rate of 86beats per minute and a blood pressure of 190/130 mmHg. She had a uniformly enlarged abdomen, the liver was not palpably enlarged and both kidneys were not ballotable. The symphysiofundal height was 35cm.There was a singleton foetus in longitudinal lie, cephalic presentation. There were no palpable uterine contractions. The foetal heart sound was present. The urine from catheter was coke coloured. Pelvic examination revealed blood smeared vulvovagina with closed cervix. An initial assessment of severe preeclampsia and possible abruptio placenta(a complication of severe preeclamsia was made. The patient was counseled on the diagnosis and she consented for admission. She had blood investigations (Full blood count, packed cell volume, urea, electrolytes with creatinine, liver function tests and clotting profile), grouping and crossmatching of 2 units of blood. Bed side clotting time was 7minutes and urinalysis revealed protein of 2+.She had an abdominal ultrasound which revealed an active foetus in cephalic presentation, anterofundal placenta with estimated foetal weight of 1.92kg.The liver was of normal size and both kidneys also were of normal sizes with good corticomedullary differentiation. She was resuscitated with intranasal oxygen and had a loading dose of magnesium sulphate (intravenous 4gm,intramuscular 5gm on each buttock). She also had intravenous hydrallazine 10mg slowly over 10 minutes and oral antihypertensives (nifedipine and α-methyl dopa). She had an emergency caesarean section due to severe preeclampsia with unfavourable cervix 4 hours after resuscitation. She was delivered of a live male neonate that weighed 1.8kg with APGAR scores of 6 and 8 in the 1st and 5th minutes respectively. There was no retroplacental clot. After returning the uterus to the abdomen, pooling of blood in the pelvis was noted. Upon re-exploration of the surgical field, additional oozing of blood was noted from the subcutaneous tissue, though no source of bleeding was identified. The uterus was well contracted after surgery. The estimated blood loss was 450mls.

She was admitted into the Intensive Care Unit(ICU) where she had 2 units of fresh whole blood transfused. She was also noticed to be bleeding from the operation site with abdominal distension. She was reviewed by the Haematology unit 4 hours after the surgery. A repeat full blood count at the end of C-section revealed a haematocrit of 32%(She had received 2 units of blood).The platelet count had decreased further from 60,000 to 52 000 cells/mm^3 and further increase in Prothrombin Time (PT) from 18 to 20 seconds and activated partial thromboplastin time(aPTT) from 70 to 122 seconds.Two units of fresh frozen plasma was requested to be transfused but the patient did not receive this because this was not available in the hospital and the patient had financial challenge in procuring it.

Postoperatively, the patient's urine appeared to be concentrated and dark red with a decreased output from 450mls on admission day to 52mls on 1st day post surgery. This warranted a review by the nephrologist by the 2nd day post surgery. She was noticed to have a tinge of jaundice, and her total bilirubin had increased from 108.7 umol/L on admission to 125 umol/L. Her ALT remained high from admission value of 402U/L to 432 U/L at post C-section. Her serum creatinine had increased from 283mg/dL to 518mg/dL. Serum urea had also increased from 4.40mMol/L to 10.0mMol/L and uric acid was 732uMol/L. By this time, the results of investigations were consistent with acute kidney injury from severe preeclampsia with ongoing haemoperitoneum.

She had 2 more units of fresh whole blood transfused on the 3rd post surgery and then had haemodialysis also on the 3rd post surgery and paracentesis abdominis with drainage of 1200mls of unclotted blood. This was done because she was having mild abdominal distension with respiratory difficulty(tachypnoea of 36 cycles/minute with Sp02 of 95%. This was followed by transfusion of 3more units of fresh whole blood on the 4th day post surgery. There was still challenge in getting frozen plasma as requested by the haematologists as haemoperitoneum persisted.

By the 5th day post surgery, after 7 units of blood had been transfused and she had a session of haemodialysis, there was increasing abdominal distension from haemoperitonuem with Prothrombin time (PT) and activated partial thromboplastin time (aPTT) which remained prolonged with consequent respiratory difficulty. The haemoperitoneum from coagulopathy was responsible for this respiratory difficulty. She then had exploratory laparotomy with drainage of 3 litres of blood in conjunction with the general surgeons. Surgical drains were left in both paracolic gutters which remained insitu for the next 5 days. Fresh frozen plasma was finally procured and she received two units about 4 hours after the procedure.

She was noticed to be lethargic, irritable with some level of disorientation. She also had asterixis and not making adequate urine by the 6th day post surgery (urine input/output in 24 hours was 2900/350 mls). A diagnosis of Uraemic encephalopathy was entertained due to the deranged serum Urea, electrolytes and creatinine with oliguria. She had two more sessions of haemodialysis subsequently. She improved gradually within the next four days and by the 10th day post caesarean section she was essentially symptom free.

She spent a total of 12 days in the ICU where she received a total of 13 units of fresh whole blood and 5 units of fresh frozen plasma. She also had 3 sessions of

Table 1: Vital signs and laboratory parameters of patient

Parameters	Day1:Admission(baseline)	Day 2:24 hours after C-section)	Day5:After receiving 7th unit of blood and 1st dialysis	Day 9:After laparotomy &drainage .Also had received 13th unit of blood,5th unit of FFP and 3rd dialysis	Day 14:13 days post C/S
Heart rate (bpm)	86	106	92	92	108
Blood pressure (mmHg)	190/130	158/88	142/89	144/92	135/87
Respiratory rate (breaths/min)	30	34	38	18	19
SpO2 (%)	98 on room air	96 on intranasal oxygen	100 on intranasal oxygen	98 on intranasal oxygen	97 on room air
Temperature (°C)	36.6	36.3	36.9	37.1	37.3
Urine input/output mls (24hrs)	2500/450	3000/52	2950/332	2900/405	3300/1920
Haematocrit (%)	25.0	32.0	31	31.8	30.3
WCC (cells/mm3)	1800	1500	1700	5600	9100
Platelets (cells/mm3)	60 000	52 000	48 000	95 000	145 000
Sodium (mEq/L)	136	130	148	141	145
Potassium (mEq/L)	3.2	3.3	4.4	4.3	4.5
Chloride (mEq/L)	102	97	115	110	117
Bicarbonate (mEq/L)	24	22.2	17.2	22.3	19.1
Uric acid (uMol/L)	918	732	533	280	160
Urea(BUN) mMol/L	4.4	10.0	26.0	15.8	10.8
Creatinine (mg/dL)	283	518	962	274	151
PT(secs)	18	20	22	15	12
aPTT(secs)	70	122	127	62	35
Albumin (g/dL)	42	45	33	2.6	2.4
ALT (U/L)	402	432	330	186	33
AST (U/L)	1216	598	421	245	34
Total bilirubin (umol/L)	108.7	125	137	8.2	7.6
Peripheral smear		Normocytic normochromic red blood cells,. Decreased platelets, giant platelets. Macrocytes, eccynocytes. Neutrophilia.			

ALT-alanine transaminase; AST- aspartate aminotransferase; PT-prothrombin time; aPTT-activated partial thromboplastin time; WCC-white cell count

haemodialysis before she was moved to the ward for further care on the 13th day post caesarean section. Both surgical drains were removed because they ceased to drain any effluent. She commenced making adequate urine (input/output in 24 hours=3300/1920mls)

She developed surgical site infection on the 15th day post caesarean section while on the ward and spent further 9 days where she had wound dressing and antibiotics. She was placed on intravenous ceftriaxone with metronidazole. She was discharged home on the 24th day post caesarean section with her wound well healed. She was given a week appointment to be seen for review.

Her baby was fine all through the mother's admission. He received his immunization and at the mother's discharge, he weighed 2.01kg.When she was seen a week after discharge in the post natal clinic, she had no complaints, her wound was well healed and her baby was doing well. She was subsequently seen two weeks and one month later at the postnatal and nephrology clinics and her clinical and laboratory parameters were normal. She had resumed menstruation and is still being followed up in the nephrology clinic.

Discussion

We presented a case of severe preeclampsia with HELLP that was complicated with DIC and acute kidney injury. She was admitted in the ICU and spent 12days there and a total of 25 days in the hospital. She received 13 units of fresh whole blood, 5 units of fresh frozen plasma and had 3 sessions of haemodialysis.

Typically, signs and symptoms of HELLP syndrome develop between 28 and 36 weeks of gestation (70%) or within 48 hours postpartum (30%).[4] This was seen in this patient who presented at 34 weeks of gestation with elevated blood pressure and epigastric pain. Haemorrhage noticed during her surgery later became uncontrollable which led to other events subsequently.

Jaundice is a rare symptom of the HELLP syndrome and is diagnosed in only 5% of the patients with this condition. However, jaundice is related to severe

presentation of the disease and associated with higher mortality.[9,10] This was noticed about 24 hours after the surgery in this patient and it is at variance to the first symptom seen in a case report of a 24 year primigravida where jaundice was the first symptom noticed.[11] So, this sign should be looked out for with a high index of suspicion for the possibility of impending complication when managing patients with similar situations.

A gradual improvement of her condition was observed after drainage of intraperitoneal haematoma with subsequent haemodialysis. This is similar to the case reported by Dybkowska K et al.[12] This drainage of haemoperitoneum was necessary since our patient was in respiratory distress with tachypnoea which improved significantly subsequently. This is similar to the report by Tsukahara E et al[13]where a patient with intraabdominal haematoma was reoperated for the control of DIC and the patient showed a satisfactory outcome after the procedure.

Thrombocytopenia is common during pregnancy and the likely aetiologies make diagnostic possibilities fraught with challenges.[14] Though there was persistent thrombocytopenia, we were able to make an early diagnosis in our patient based on the prompt availability of laboratory results. Our patient developed rapidly worsening hemodynamic instability in the post partum period requiring admission to the ICU. Disseminated intravascular coagulopathy has been reported to be significantly associated with acute kidney injury.[15] There was progression to acute kidney injury in our patient. Treatment of DIC consists of replacement of volume, blood products, and coagulation components and cardiovascular and respiratory support with elimination of precipitating factors which when identified early, reduces the morbidity for patients.[14,16] This was the situation in this patient where she did not make good recovery after the first session of haemodialysis with blood transfusion. She only did after she had 3 sessions of haemodialysis.

Treatment of DIC is centered on two principles. The first is the identification and treatment of the underlying disorder.[14,16] The second goal of treatment is that obstetric complications such as uterine atony or lacerations must be controlled simultaneously with prompt blood and component replacement for a curative outcome.[16] These were carried out dutifully in this patient along with haemodialysis to the benefit of the patient. Even though this patient was not in labour a pelvic examination was done to rule out any genital injuries.

Massive haemorrhage is among the most challenging issues in critical care and obstetric patients.[17] We had challenges getting fresh frozen plasma on time due to financial constraints on the part of the patient and its unavailability in the hospital when initially requested. The promptness in administering this has been found very beneficial in patients with coagulopathy.[17,18]

Conclusion:

This case describes how unexpectedly HELLP syndrome may occur, from an uneventful pregnancy with no identifiable risk factors to multiorgan failure within hours despite delivery of the fetus. Obstetric DIC is a condition that is associated with high maternal and perinatal morbidity and mortality. Prompt recognition, teamwork and treatment with timely administration of blood products along with other supportive care is crucial in the management of this life-threatening and challenging condition.

Conflicts of interest: none
Financial support and sponsorship: none

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