Case Report

Eclampsia and HELLP Syndrome: A Case Report

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Abstract

Background: The HELLP syndrome is characterized by hemolysis (H), elevated liver enzymes (EL) and low platelet count (LP). This syndrome in general complicates 0.2–0.6% of all pregnancies but its incidence increases to 4–12% in severe preeclampsia. In about 15% cases, HELLP syndrome presents without definitive criteria for preeclampsia (atypical preeclampsia).

Case presentation: We describe a 38-year-old second gravida woman who presented with signs of HELLP, with seizures at 32 weeks of gestation, and required emergent delivery of the foetus. The patient also sustained ARDS, cardiac arrest, acute kidney injury requiring haemodialysis and a prolonged intensive care management. With supportive care, clinical status, all laboratory derangements, including renal function, normalised after 4 weeks.

Conclusion: Timely diagnosis and appropriate intervention facilitated recovery and discharge of, the patient, in good general condition, after 32 days of hospitalization.

Key words: HELLP syndrome, Eclampsia, microangiopathic hemolytic anemia, acute kidney injury, multiorgan failure.

Background

Hypertensive disorders in pregnancy represent a large spectrum of disorders where the HELLP syndrome is on the severe end. The incidence of pre-eclampsia in hospital practice in India varies from 5 to 15% and that of eclampsia about 1.5% [1]. Eclampsia continues to be a major cause of maternal and perinatal morbidity and mortality worldwide. The maternal mortality rate is approximately 4.2% [2] (Table 1).

Table 1. Hypertensive disorders of pregnancy

Systolic blood pressure (SBP) is ≥140 mmHg, diastolic BP (DBP) is ≤90 mmHg, or both, on two occasions (at least 4 h apart)			
Pre-pregnancy	**<20 weeks**	**>20 weeks**	**≤12 weeks postpartum**
Chronic hypertension of pregnancy: diagnosed pre-pregnancy or ≤ 20 weeks gestation or persists >12 weeks postpartum			
		Gestational hypertension Diagnosed >20 weeks gestation, without proteinuria, resolves ≤12 weeks postpartum	
		Pre-eclampsia HTN >20 weeks ≤12weeks postpartum, proteinuria ≥300 mg/24 h, protein-creatinine ratio ≥0.3, or ≥1+ urine dipstick specimen Severe pre-eclampsia: Blood pressure ≥ 160/110 mm Hg • Thrombocytopenia (platelet count less than 100,000/mm^3) • Serum creatinine concentration greater than 1.1 mg/dL or greater than 2 times the baseline serum creatinine concentration • Pulmonary edema • New-onset cerebral or visual disturbances • Impaired liver function Eclampsia New onset seizures	

Classification of Hypertensive Disorders in Pregnancy [3,4]

(1). Gestational hypertension

(2). Preeclampsia

   a. Preeclampsia without severe features

   b. Severe preeclampsia

(3). Chronic hypertension

(4). Chronic hypertension with superimposed preeclampsia

HELLP syndrome is a serious complication of pregnancy related to pre-eclampsia. It was first suggested by Weinstein (1982) [5]. This syndrome in general complicates 0.2–0.6% of all pregnancies but its incidence increases to 4–12% in severe preeclampsia. HELLP syndrome occurs mostly in antepartum (70%), and few in postpartum (30%). It is associated with higher maternal morbidity

(Table 4) [6]. The pathophysiology of HELLP syndrome is ill defined. Generally, the disorder is considered a placenta-instigated, liver-targeted acute inflammatory condition, with elements of disordered immunological processes [7].

Eclampsia is defined as the new onset of seizures or unexplained coma during pregnancy or the postpartum period in a woman with signs and symptoms of preeclampsia and without a pre-existing neurologic disorder [3]. The hospital incidence in India ranges from 1 in 500 to 1 in 30. It is more common in primigravidae (75%), five times more common in twins than in singleton pregnancies and occurs between the 36th week and term in more than 50% [8]. Since eclampsia is a severe form of pre-eclampsia, pathophysiology is similar to pre-eclampsia. The mechanism of eclamptic seizures is poorly understood. It may involve a loss of the normal cerebral autoregulatory mechanism, resulting in hyper-perfusion and leading to interstitial or vasogenic cerebral edema and decreased cerebral blood flow [3]. Eclampsia can occur antepartum (50%), intrapartum (25%), or postpartum (25%) [8]. Until proven otherwise, the occurrence of seizures during pregnancy should be considered eclampsia. The majority of eclamptic women have evidence of severe preeclampsia, but in 10 to 15% of cases, hypertension is absent or modest and/or proteinuria is not detected [2] (Table 2–4).

Table 2. Diagnostic criteria for HELLP syndrome

Hemolysis (at least two of these)
(1). Abnormal peripheral smear (schistocytes, burr cell, echinocytes, etc)
(2). Increased total bilirubin (mostly indirect form) >1.2 mg/dl
(3). Low serum haptoglobin level
(4). Drop in haemoglobin level unrelated to blood loss
Elevated liver enzymes
(1). Increased transaminases (AST and ALT) >70 IU/L (twice the upper limit of normal)
(2). Increased lactate dehydrogenase >600 IU/L
(3). Increased total bilirubin >1.2 mg/dl
Thrombocytopenia
(1). Platelet count <100,000

Table 3. Differential diagnosis of HELLP syndrome [5]

(1).	Acute fatty liver of pregnancy
(2).	Appendicitis
(3).	Cholestasis of pregnancy
(4).	Diabetes insipidus
(5).	Gallbladder disease
(6).	Gastroenteritis
(7).	Glomerulonephritis
(8).	Hemolytic-uremic syndrome
(9).	Hepatic encephalopathy
(10).	Hyperemesis gravidarum
(11).	Idiopathic thrombocytopenia
(12).	Nephrolithiasis
(13).	Peptic ulcer disease
(14).	Systemic lupus erythematosus
(15).	Thrombotic thrombocytopenic purpura
(16).	Viral hepatitis

Table 4. Maternal complications associated with HELLP syndrome [4]

Complications	**Incidence**
DIC	15
Abruptio placentae	10–15
Marked ascites	10–15
Wound hematoma or infection	14
Pulmonary edema	8
Pleural effusions	6–10
Acute renal failure/acute tubular necrosis	3
Subcapsular hematoma/infarction/failure	<2

Laryngeal edema	1–2
Retinal detachment, vitreous haemorrhage and cortical blindness	01
Death	01
Others-Adult respiratory distress syndrome, sepsis, stroke, pancreatitis, myocardial infarction and diabetes insipidus	01

Management

Immediate goals are to stop convulsions, establish a patent airway, and prevent major complications (e.g., hypoxemia, aspiration). Further obstetric management includes antihypertensive therapy, induction or augmentation of labor, and expeditious (preferably vaginal) delivery. Due to the progressive nature of the disease, these patients should always be hospitalized with strict bed rest and care in labor and delivery due to the potential for sudden deterioration of maternal or fetal condition. After assessment and stabilization of maternal status, the fetus is evaluated by fetal heart rate tracing, biophysical profile and or doppler studies. The assessment of maternal and fetal status helps determine when delivery is required or imminent, as delivery is the only true cure for this syndrome [9].

Case Presentation

A 38 years old female, a booked case of gravida: G3, para: P1, living:1, abortion:1, at 32 weeks of gestation, came to our emergency department in evening with complaints of nausea, headache, epigastric pain and one episode of seizures in her residence. Patient had repeat episode of seizure associated with tongue bite in our emergency department. Immediately first aid medications were given. On examination the patient was drowsy, arousable and obeying commands. Her blood pressure 140/100mmHg, pulse rate 100 beats/min and hematuria were noted. She had a past medical history of seizure disorder seven years back and took medication for past five years and stopped. There was history of elective caesarean section for first pregnancy delivery without any history of hypertensive disorder of pregnancy (Table 5).

Table 5. Baseline investigation (Day 0)

Haemoglobin	12.8 g/dl
WBC count	22560/cmm
Platelet count	52000/cmm
INR	1.12
Total bilirubin	3.5mg/dl
ALP	182U/L
SGOT	211U/L
SGPT	104U/L
Creatinine	1.2mg/dl

Multidisciplinary team of doctors consisting of emergency physician, obstetrician, anaesthesiologist, paediatrician and intensivist were involved. With a working diagnosis of eclampsia and HELLP syndrome, emergency LSCS was planned. Packed cells, FFP, platelets were arranged, patients' husband was counselled about the critical situation and high-risk consent was taken. On administration of labetalol and magnesium patients BP had optimised gradually to 138/95 mmhg.

Patient underwent emergency pre-term lower segment caesarean section under general anesthesia in view of low platelet count (Day 0). Rapid sequence induction was done with cricoid pressure, propofol, and succinyl choline. Intubated with no 6.5 cuffed endotracheal tube. Maintenance was done with 50% oxygen, nitrous oxide and 0.8 mac sevoflurane. After baby extraction fentalin and paracetamol were given. Magnesium infusion was continued. Intra operatively 1 pint packed red blood cells was transfused. In view of HELLP syndrome and eclampsia elective post-operative ventilation in ICU was decided. The new born was preterm and was shifted to PICU (Table 6).

Table 6. Repeat investigations

Repeat investigations	Day 1	Day 2	Day 3 17.11.20	Day 27 11.12.20
Haemoglobin (g/dl)	8.1	5.6	7.8	10.6
WBC count (cmm)	24750	25410	28310	8620
Platelet count (cmm)	44000	87000	93000	374000
INR	1.37	0.9	–	–
Total bilirubin (mg/dl)	3.6	1.2	–	0.5
ALP (u/l)	86	74	–	79

SGOT (u/l)	690	147	–	32
SGPT (u/l)	170	99	–	24
Creatinine (mg/dl)	1.8	3.4	4.10	0.7
LDH (IU/ml)	–	1105	–	–

Course in ICU

On Day 1 patient was doing well and hence extubated. Blood and blood products were administered for correction of anaemia, thrombocytopenia and coagulation abnormalities. In the evening patient developed breathing difficulty and so was put on NIV. Her renal function started deteriorating. Her ongoing issues were acute kidney injury, acute respiratory distress syndrome, anaemia and thrombocytopenia which are some of the common complications of eclampsia and HELLP syndrome. As these worsened, on day 4, patient had to be intubated and put on mechanical ventilation. She also had a cardiac arrest and got revived after about 2 min of CPR. There was severe ARDS, sepsis with MODS (multi organ dysfunction syndrome) and worsening AKI (acute kidney injury). Neurologist, nephrologist, cardiologist, gynaecologist, physiotherapist and nutritionist worked along with intensivists to form a multidisciplinary care team. Prone ventilation, haemodialysis, neuroimaging, escalation of antibiotics, steroids, anticonvulsants, nutritional support, prompt physiotherapy support and blood product transfusions were done. Tracheostomy was performed for thorough respiratory therapy.

Patient gradually improved with multidisciplinary treatment. Tracheostomy was decannulated and shifted to ward. In the ward, patient recovered well and was discharged on 32nd post-operative day.

Discussion

This case report suggests the pre partum development of HELLP syndrome in combination of eclampsia. Apart from eclampsia, advanced maternal age is also one of the risk factors for developing HELLP syndrome.

Anaesthetic management of eclampsia and HELLP syndrome

Pre-anaesthesia evaluation for assessment of seizure control and neurologic status, with particular attention to signs of increased ICP

and focal deficits, coagulation abnormalities, intravascular volume status should be done. Intracranial imaging is typically not warranted unless focal neurologic signs persist, or the diagnosis is uncertain. This patient's seizure and high blood pressure was controlled with magnesium loading dose of 10 g IM, labetalol 10 mg IV intermittently, and in infusion doses, were started. Continues monitoring of fetal heart rate, maternal heart rate, saturation, and blood pressure were done.

Intraoperative management

Eclampsia is not an absolute contraindication to neuraxial anaesthesia. In fact, neuraxial anaesthesia is the method of choice in patients with no focal signs of neurologic deficits, alert mental status, and normal coagulation parameters. Neuraxial procedures may be initiated in pregnant women without other risk factors if the platelet count is higher than 80,000/mm^3. Platelet counts below 50,000/mm^3 precludes the administration of neuraxial anaesthesia. When platelet count is between 50,000 and 80,000/mm^3, the risks and benefits of neuraxial anaesthesia must be weighed against the risks associated with general anaesthesia for the individual patient if emergency caesarean delivery is required, including whether anatomic features of the patient's airway are favourable. In our patient platelet count was low hence general anaesthesia was chosen. The concerns regarding a difficult airway in pregnancy are magnified in a patient with eclampsia. A difficult airway cart with various sizes of tracheal tubes, laryngeal mask airways, etc. Were kept ready. Rapid sequence induction was done to avoid aspiration risk. Proper attenuation of laryngoscopic response and gentle intubation was done as intracranial haemorrhages during intubation was a leading cause of maternal mortality in women with pre-eclampsia. As patient was on magnesium sulphate titrated doses of non-depolarising neuromuscular blockers was administered.

Postpartum women are at significant risk for pulmonary edema, sustained hypertension, stroke, venous thromboembolism, airway obstruction, and seizures, and should receive close monitoring of oxygenation, blood pressure, fluid intake, and urinary output. A potential complication of HELLP syndrome is subcapsular liver hematoma, which has over 50% maternal and fetal mortality. In ICU our patient was monitored closely for all these complications. AKI is more commonly associated with HELLP, complicating 7–15% of cases, as opposed to pre-eclampsia

alone without HELLP at 1%. One study reports the incidence of AKI from 7.7 to 60% among patients with HELLP syndrome [10].

Here the patient developed AKI, pulmonary edema and ARDS which were promptly managed by renal replacement therapy and prone mechanical ventilation.

Conclusion

HELLP syndrome is a serious complication in pregnancy and it also increases the risk in patient with preeclampsia. Timely diagnosis and management of preeclampsia in earlier pregnancy will reduce the risk of convulsive episodes, multi-organ failure and reduces the feto-maternal morbidity and mortality.

Competing interests:

The authors have no competing interests to declare.

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