Case Report

HELLP Syndrome

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Abstract

The case presented here is of HELLP syndrome which poses a great challenge especially when it is managed in a peripheral hospital. The patient was managed promptly with urgent caesarean section and supportive treatment at Pakistan Aeronautical Complex (PAC) Hospital Kamra and was shifted to tertiary care hospital where the patient was treated successfully in high dependency unit for subsequent complications. With this case report, the authors aim to point out the importance of continual intensive clinical follow-up, laboratory monitoring and corresponding therapeutic procedures in the successful management of this relatively serious condition.

Keywords: HELLP syndrome, Pre-eclampsia, Pregnancy.

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Introduction

Haemolysis, Elevated Liver enzymes, and Low Platelet count (HELLP) syndrome is a severe manifestation of a hypertensive disorder of pregnancy called pre-eclampsia. It carries significant foetal and maternal morbidity and mortality, which increases with the severity of syndrome. Maternal complications include cerebrovascular haemorrhage, pulmonary oedema, retinal detachment, hepatic hematoma or rupture, acute kidney injury (AKI) and intravascular coagulopathy. Foetal complications include placental abruption, intrauterine growth restriction and intrauterine death.

The management of HELLP syndrome includes prompt delivery, adequate blood pressure control, control of seizures and blood and blood products transfusion. Acid-base balance, haemodialysis and ventilatory support may also be needed. Multisystem involvement needs prolonged hospital stay; such patients are prone to depression that needs psychological support and treatment.

Case Report

A 36-year-old woman G4P3, was admitted through obstetric outpatient department with gestational amenorrhea of 35 weeks and 5 days. The patient was suffering from severe epigastric pain and vomiting for 6 hours. The patient previously had three caesarean sections with alive and healthy daughters. The patient had no family history of hypertension. On admission, pulse rate was 100bpm and blood pressure (BP) was 120/70mmHg to 130/90mmHg, taken thrice in following two hours. There was significant epigastric tenderness and exaggerated deep tendon reflexes. The fundal height was consistent with 34 weeks. Her haemoglobin (Hb), total leukocyte count (TLC) and platelet count were 13.3g/dl, $13.3x10^9$/l and $86x10^9$/l respectively. Liver function tests showed bilirubin 19µmol/l and alanine aminotransferase (ALT) 795 IU/L. Lactate dehydrogenase (LDH) was 905 U/l. Coagulation profile, renal function tests, serum amylase and blood sugar were within normal limits. Electronic foetal heart monitoring was reactive. The obstetric ultrasonography revealed foetus corresponding to 33 weeks and 4 days

with adequate liquor. Umbilical artery Doppler assessment revealed increased diastolic resistance. On abdominal ultrasonography (USG), additional finding of cholelithiasis was confirmed. Within two hours of admission, a provisional diagnosis of HELLP syndrome and decision of immediate caesarean section was made. The patient had not provided urine sample since admission in spite of request. At bladder catheterization, only a few ml of blood-stained urine was obtained. The report showed 2+ proteinuria with numerous red blood cells per high power field. The laboratory was requested for arrangement of blood. Almost two hours after admission the electronic foetal heart rate tracing started showing absent variability and patient had a sudden surge of BP to 200/120mmHg. Injection magnesium sulphate and injection hydralazine were given. The patient was immediately taken to operation theatre where the patient developed blurring of vision and had first and the last eclamptic fit. Lower uterine segment caesarean section with bilateral tubal ligation was performed due to massive adhesions, under general anaesthesia (GA). Findings included peritoneal fluid mixed with blood but no evidence of liver hematoma or rupture. The outcome was a male child with Apgar score of 5/10, weighing 2.2kg and serum lactate levels of 7.1mmol/l who died on first postnatal day due to respiratory distress and gastrointestinal bleed in spite of ventilatory support and fresh frozen plasma (FFP) transfusions. One red cell concentrate (RCC) and 6 platelet concentrates were transfused during surgery. Patient was shifted to intensive care unit (ICU) on mechanical ventilation. Her investigations, one hour after surgery, revealed deterioration in condition as shown in Table I.

In 12 hours following the surgery, 4 RCC, 1 fresh whole blood, 8 platelets concentrates, 10 FFP and 3 cryoprecipitate were transfused. The patient remained oliguric. The next morning, peritoneal drainage increased up to 2000ml. Due to the limitations of overall setup at PAC Hospital Kamra, the patient was shifted to Military Hospital (MH) Rawalpindi 16 hours post operatively. At MH ICU, double lumen central venous catheter and nasogastric tube were passed. Laparotomy was done 24 hours after the caesarean section. No definite point of haemorrhage was observed except generalized bruising. Internal iliac arteries were ligated. The patient was managed with magnesium sulphate infusion 1g/hr to 2g/hr and injection hydralazine intermittently for next 10 days. Blood and blood products including 10 RCCs, 22 FFPs, 4 Mega platelets, 10 platelet concentrates and 3 cryoprecipitate were transfused. Injectable antibiotics were given according to culture sensitivity patterns. Haemodialysis was performed 6 times. Nasogastric feeding was started on the third day. The patient remained on different modes of mechanical ventilation for next 10 days. Liver enzymes became normal on 5th day and AKI started recovering on 11th day. Chest physiotherapy was started and the patient was shifted back to PAC hospital Kamra after 22 days on treatment, including calcium channel blocker, alpha-blocker and angiotensin-converting enzyme

Table I. Summary of laboratory tests during the course of the disease

Lab Reports	Operation Day		1st Post-operative Day	20th Post-operative Day	Normal
	Pre-Op	Post-Op			
Blood Picture					
Hb (g/dl)	11.4	8.5	7.1	9.4	10.5 – 12.0
Platelet Count (/l)	86x10^9	46x10^9	40x10^9	194x10^9	150 – 400 x10^9
Prothrombin Time					
Patient (sec)	13	14	16	14	0 – 14
Control (sec)	12	12	14	14	0 – 14
Partial Thromboplastin Time Activated with Kaolin					
Patient (sec)	34	36	37	32	0 – 32
Control (sec)	32	32	32	32	0 – 32
Liver Function Tests					
Serum Total Bilirubin (µmol/l)	19	22	26	5	2 – 17
Serum ALT (IU/l)	795	1200	1310	32	0 – 42
Serum Alkaline Phosphatase (U/l)	170	170	177	148	65 – 306
Renal Function Tests					
Serum Urea (mmol/l)	8.9	13.2	22.9	6.2	2.1 – 7.1
Serum Creatinine (µmol/l)	102	220	360	111	53 – 97
Muscle Enzymes					
Serum LDH (U/l)	905	1020	7059	653	230 – 460

inhibitor. Laboratory reports during the course of the disease are summarized in Table I.

Discussion

The acronym HELLP describes a variant of severe pre-eclampsia characterized by Haemolysis, Elevated Liver enzymes, and Low Platelets. It was first described in 1982 by Weinstein.[1] The incidence of HELLP in pre-eclampsia is 5% to 20% although 20% to 50% of pre eclamptic women have mild derangement of liver functions.[2] Maternal and perinatal mortality associated with HELLP syndrome is 1% to 24% and 9% to 60% respectively. The diagnostic and therapeutic challenges posed by HELLP syndrome are diverse. Of all the women with HELLP syndrome, 18% may not be hypertensive and 13% are non-protienuric at the time of presentation. It may arise postnatally in 30% of the patients.[3]

HELLP syndrome is diagnosed by Tennessee criteria comprising of haemolysis, LDH> 600 U/L, aspartate transaminase (AST) or ALT >70U/L and platelet count < 100 $x10^9$/L. Its severity is further classified from class I to III based on platelet nidar by Martin and Mississippi.

The presented patient suffered from acute onset of HELLP syndrome with its classical symptom of severe epigastric pain and tenderness.[4] Paradoxically, her abdominal USG revealed gall stones. All these initial findings could delay the diagnosis but presence of oliguria and the results of supportive investigations helped to reach at the diagnosis of HELLP syndrome while ruling out acute fatty liver, thrombotic thrombocytopenic purpura and cholecystitis. Emergency caesarean section was performed after controlling BP and seizure under GA. Regional anaesthesia was avoided because of eclampsia and thrombocytopenia.

The University of Mississippi Protocol for HELLP syndrome includes corticosteroids but 2010 systematic review by Cochrane Collaboration found no clear evidence of any effect of steroids.[5,6] According to latest recommendations dexamethasone or betamethasone, given antenatally for foetal lung maturity, are the only steroids which are beneficial in the management of maternal disease as well. Postnatal steroids are not proven to be effective. Central venous catheter could not be passed at PAC hospital Kamra because of the risk of haemorrhage. As has been described in the literature, after expedited delivery, the patient deteriorated regarding hepatic and coagulation profiles and developed AKI. The patient was shifted to tertiary care hospital for the management under multidisciplinary intensive care.

The probability of development of disseminated intravascular coagulation and pulmonary oedema in patients with AKI is 20% and 44% respectively. The patient was managed for all these complications in next 22 days. The patient recovered according to the natural course of disease; her liver and renal functions returned to normal in next 15 days. Anti-hypertensive therapy continued for further 3 weeks and anxiolytics were given for 3 months.

One of the other treatment options under trial is Eculizumab, a targeted inhibitor of complement protein C5, which may result in marked clinical improvement and normalization of laboratory parameters.[7]

Oral contraceptives are safe in women with prior HELLP syndrome. Long-acting reversible contraceptives such as intrauterine devices or subdermal implants are the most effective forms of contraception.[9] The incidence of recurrent HELLP syndrome is only 5% but of pre-eclampsia after HELLP is 20%.[10] As the patient had undergone sterilization, there was the least concern about the mentioned risks.

Conclusion

HELLP syndrome, in its severe form, is a life-threatening condition which needs untiring efforts of a multidisciplinary team. The course of the disease takes a couple of weeks for its recovery with ongoing changing treatment including blood transfusion, ventilatory support and dialysis.

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Corrigendum

The name of the third author (**Rubina Irfan**) of the previous two articles titled 'Role of Internal Iliac Artery Ligation in Severe Obstetric Haemorrhage: Still Alive' published in Vol 6(1)Jan-Mar 2016:12-16 and 'Comparison of Fetomaternal Outcome Between Scarred and Unscarred Uterus in Placenta Parevia Cases' published in Vol 6(3)July-Sept 2016:102-105, were the typo errors.

The correct name of the third author of these two published articles should be read as **Rubina Ashraf**

The typo errors have been corrected in the online version of both manuscripts as well as the article citation. The correct citation of both articles should be read as

Abid R, Begum A. **Ashraf R**, Shaheen F. Role of Internal Iliac Artery Ligation in Severe Obstetric Haemorrhage: Still Alive . J. Soc. Obstet. Gynaecol. Pak. 2016; Vol 6(1):12-16.

Iqbal K, Abid R, **Ashraf R**, Shaheen F. Comparison of Fetomaternal Outcome Between Scarred and Unscarred Uterus in Placenta Parevia Cases.J. Soc. Obstet. Gynaecol. Pak. 2016; Vol 6(3):102-105.