HELLP Syndrome : Report of Two Cases

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Introduction

The acronym HELLP was coined in 1982 to describe a syndrome consisting of Haemolysis Elevated Liver enzyme levels and Low Platelet count [1]. The syndrome considered a variant of preeclampsia, can occur on its own or in association with preeclampsia. Pregnancy induced hypertension (PIH), preeclampsia and HELLP are related and overlap in their presentation. Maternal and foetal morbidity and mortality are significant in HELLP syndrome [2]. Various life threatening complications such as placental abruption, pulmonary oedema, cerebral haemorrhage, hepatorenal failure and disseminated intravascular coagulation (DIC) can occur in these patients. We present two cases of HELLP syndrome with vague presenting complaints. First patient developed HELLP in association with severe preeclampsia and in the second patient HELLP led to foetal death. We discuss the surgical and anaesthetic implications during peri-operative period.

Case Report-1

A 21 year old primigravida at 35 weeks of gestation was admitted with labour pain, headache, epigastric pain and blurring of vision. On examination there was altered consciousness, pulse 86 per minute, blood pressure 170/110 mm Hg, breath rate 24 per minute, and brisk tendon jerks. Based on obstetrical examination delivery by vaginal route was planned. Baseline investigations showed haemoglobin 11.9gm%, platelets 1,60,000/mm³, blood urea 26mg%, serum creatinine 0.9mg%, serum bilirubin 0.9mg%, alanine aminotransferase (ALT) 40 units per litre, aspartate aminotransferase (AST) 28 units per litre. To treat hypertension oral nifedipine 10mg and magnesium sulphate by Pritchard's regime was started. After 4 hrs of admission, repeat examination revealed pulse rate of 84 per minute, blood pressure of 160/100 mm Hg and urinary output was 50 ml. Because of falling urinary output magnesium sulphate was withheld and oral nifedipine continued. After 2 hrs patient had an episode of generalised tonic clonic seizures, for which diazepam 10 mg intravenously was administered. It was then

decided that the pregnancy be terminated by emergency caesarean section.

General anaesthetic technique with acid aspiration prophylaxis was selected. Pre-induction fentanyl 20 µg and esmolol 10 mg administered to decrease the pressure response to laryngoscopy and endotracheal intubation. After preoxygenation a rapid sequence induction-intubation technique was used to facilitate endotracheal intubation. Anaesthesia was maintained with gas, oxygen, isoflurane and atracurium. Additional doses of fentanyl and esmolol were given to keep haemodynamic parameters within 20% of preoperative values. A live male baby was delivered. Due to weak cry, newborn was given 300 µg of naloxone. Neostigmine and glycopyrrolate were given for reversal of muscle relaxant. Following extubation patient was drowsy and disoriented hence observed in intensive care unit. Over the next 12 hours patient had four generalised seizures despite continuing intramuscular magnesium sulphate. Blood pressure ranged from 160 to 200 mm Hg systolic and 110 to 130 mm Hg diastolic with heart rate between 120 to 130 per minute. At this time oliguria and tender hepatomegaly was noticed. Because of persistent seizures and hypertension a continuous infusion of magnesium sulphate 1 gm/hour and labetalol 20 mg/hour was started and monitored by knee jerk response. Repeat investigations after 24 hours revealed haemoglobin 6.8 gm%, platelets count 86,000/mm³, prothrombin time (PT) 07 seconds and partial thromboplastin time (PTT) was 08 seconds more than control values. The blood urea was 60mg %, serum creatinine 1.9 mg %, serum bilirubin 3.4 mg %, ALT 1040 units per litre, AST 646 units per litre, lactate dehydrogenase (LDH) 658 units per litre and peripheral smear showed evidence of haemolysis. Antiphospholipid antibodies were negative. Both infusions were continued for 36 hours after last seizure. A total of 260 mg of labetalol was used. Blood and fresh frozen plasma was transfused appropriately. Over the next two days patient improved clinically, however laboratory parameters took 10 days to return to preoperative values. On 12th day patient was discharged home uneventfully.

Case Report-2

A 26 year old lady with twin pregnancy presented at 33 weeks of gestation, with intra-uterine death of one foetus at

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the time of admission and was taken up for emergency caesarean under subarachnoid block. On examination she was drowsy, pulse 120 per minute, blood pressure 100/56 mm Hg, had pedal and sacral oedema. Preoperative investigations showed haemoglobin of 11.2gm %, platelet count 2,10,000/mm³, blood urea 24 mg %, serum creatinine 0.8 mg %, serum bilirubin 3.0 mg%, ALT 184 units per litre, AST 158 units per litre. After adequate preloading, 12 mg of bupivacaine was injected intrathecally. Both the babies delivered were still born and the intraoperative course was uneventful. In the postoperative period renal function deteriorated with blood urea of 36 mg % and serum creatinine 1.8mg %. There was evidence of haemolysis on peripheral blood smear and LDH was raised to 1110 units per litre. Coagulation profile was deranged with PT 12 secs and PTT 23 secs more than the control values and platelets count fell to 1,95,000/mm³. On first postoperative day, patient developed tachycardia (heart rate 130-140/min) and hypertension (170/100 mm Hg) which was treated with oral atenolol. With supportive care patient recovered and was discharged after 2 weeks.

Discussion

HELLP is a multi-system disease, resulting in generalised vasospasm, microthrombi formation and coagulation defects [3]. The syndrome seems to be the final manifestation of insult that leads to micro vascular endothelial damage and intravascular platelet aggregation. Significant symptoms and signs in any patient with preeclampsia include headache, blurred vision, altered consciousness, clonus, increasing serum creatinine level, consumptive coagulopathy with thrombocytopenia, and abnormal liver function tests [4]. Both our patients had altered consciousness, possibly an early but subtle sign of developing HELLP syndrome.

The HELLP syndrome occurs in 4-18% of patients with preeclampsia. Upto 30% patients develop HELLP syndrome after parturition, typically appearing within 48 hours. In fact, there may be no evidence of preeclampsia before or during labour in 20% of cases. The serum transaminase levels may be elevated and platelet counts can drop to as low as 6000/mm³. Platelet count is the best indicator of HELLP. Progressive isolated thrombocytopenia may be one of the first clues to the diagnosis [5]. Both the above mentioned patients developed renal dysfunction, consumptive coagulopathy and thrombocytopenia. Excessive fluid overload during anaesthetic management can result in cerebral or pulmonary oedema. A positive D-dimer test in the setting of preeclampsia has recently been reported to be predictive of patients who will develop HELLP syndrome [6].

The laboratory abnormalities typically worsen after

delivery and peak at 24 to 48 hours postpartum and resolve by three to four days [7]. Patients with HELLP syndrome should be treated prophylactically with magnesium sulphate to prevent seizures. In Intensive Care Unit (ICU) setting magnesium sulphate and labetalol can be used as an intravenous infusion for the treatment of seizures and hypertension. This also reduces the risk of maternal cerebral haemorrhage. The mainstay of therapy is supportive management and delivery is the definitive treatment. These patients may develop placental abruption and require emergency caesarean section. Although hypertension is not a contradiction to regional anaesthesia [8] but patients with altered consciousness and platelet counts of less than 70,000/mm³ should not be given spinal or epidural anaesthesia. A high degree of suspicion should be maintained whenever patients with preeclampsia present with nonspecific features. The first patient developed eclampsia despite prophylactic magnesium sulphate therapy. The cause for foetal death in second patient could be severe coagulopathy and haemolysis. Women with a history of HELLP syndrome are considered to be at increased risk for complications in future pregnancies.

Conflicts of Interest

None identified

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