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Case Report

HELLP syndrome - A life threatening complication of severe pre- eclampsia

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Abstract

HELLP Syndrome is a life threatening complication of severe pre eclampsia ,which is characterized by evidence of hemolysis, elevated liver enzymes and low platelet count during second half of pregnancy.If untreated , it carries high risk of maternal and perinatal morbidity and mortality due to hepatic and renal complications. A case of HELLP Syndrome with severe pre eclampsia in a primigravida at 39 weeks of pregnancy is reported. She was a known case of severe pre eclampsia and was taking treatment in a private nursing home.She was refered with a picture of HELLP Syndrome.Caesarean section under general anesthesia for fetal distress was carried out following single donar platelet transfusion. She had gross derrangement of hepatic and renal functions and thrombocytopenia following caesarean section. She was treated aggresively in intensive care unit with transfusion of whole blood ,fresh frozen plasma, multiple single donar platelet transfusions ,antihypertensive agents and broad spectrum antibiotics.Patient required multidisciplinary team approach for management.Patient was discharged after fifteen days from hospital.
Keywords: HELLP Syndrome , Severe pre eclampsia ,Thrombocytopenia,Component therapy

1. Introduction

HELLP syndrome is a group of symptoms that occurs in pregnant women who have pre-eclampsia or eclampsia and who also show signs of liver damage and abnormalities in blood clotting. It is characterised by: H- Haemolysis, EL- Elevated liver enzymes , LP- low platelet count. It occurs in 0.5 to 0.9% of all pregnancies and in 10-20% of cases with severe pre-eclampsia[1]. 80% of women with HELLP syndrome present before term. Patients usually present with symptoms of progressive nausea and vomiting, upper abdominal pain, headache and visual problems. The usual signs are jaundice ,upper abdominal tenderness, especially in the right upper quadrant, hepatomegaly and easy bruising/purpura. If HELLP syndrome is not treated early, up to 25% of women may develop serious complications. Without treatment there is a significant mortality. The mortality rate among babies born to mothers with HELLP syndrome varies and depends mainly on gestation and birth weight. The common complications of HELLP syndrome include maternal liver haemorrhage or rupture[2], coagulopathy, postpartum hemorrhage,permanent liver damage or necrosis[3], which may need transplantation, intraventricular haemorrhage with subsequent hydrocephalus has been reported[4]. Retinal detachment and other eye problems have been reported.[5] Transient diabetes insipidus may follow HELLP syndrome.[6]

2. Case Report

Twenty three year unbooked primigravida reported from private nursing home with 38 weeks of pregnancy with severe pre eclampsia and thrombocytopenia for further management. Patient was taking antihypertensive agents(Labetalol

100 mg twice a day) since 20 days of admission. Patient had swelling over body, frontal headache, blurring of vision and epigastric pain. On examination, she was conscious and oriented. Her blood pressure was 160 /104 mm Hg. She had high BMI value of 28.Per abdominal examination revealed a full term uterus with large size baby in cephalic presentation with approximate baby weight of 3.4 kgs. Internal examination revealed a long uneffaced cervix with borderline cephalo pelvic disproportion. She was kept in critical care unit and was treated with Tab. labetalol 100 mg twice a day. Her laboratory investigations revealed platelet count of 51,000/cumm, Haemoglobin - 14.2 grams, Prothrombin time -17.2sec, INR-1.32, Partial thromboplastin time-54 sec. Her S.LDH levels were 1556 IU/L. After 12 hours of admission,her blood pressure increased to 160/120 mm Hg. She was treated with intravenous Labetalol 20 mg and 5 grams of prophylactic intramuscular magnesium sulphate. Her fundus examination revealed serous retinal detachment. In view of her low platelet count and possible need of caesarean section, she was transfused pre operatively with one unit of single donar platelet. Following this transfusion, patient went in labour and developed severe fetal distress. Emergency caesarean section was carried out under general anesthesia after counselling and explaining the risk of operative intervention to the relatives. There was no intra operative complication like post partum haemorrhage. A male child with birth weight of 3.3kg was delivered with low APGAR score. Baby required resuscitation and intensive neonatal care. Patient was kept in intensive care unit during post operative period. Her platelet count,liver function and renal functions showed gross deterioration following caesarean section. Her Hb level dropped to 6.9 grams,S Bilirubin raised to 11mg /dl,S.SGOT -4850IU/L, B.Urea to 167mg/dl,S. Creatinine to 2.3mg/dl.She was transfused with 8 units of fresh frozen plasma,2 units of fresh blood and 2 units of single donar platelets during first post operative day.Her general condition deteriorated and she showed early signs of disseminated intravascular coagulopathy**. (Fig 1 and 2)** She developed hematuria, petechial hemorrhages and intraperitoneal bleeding. Her platelet count dropped down to 24,000/cumm. Liver enzymes, bilirubin levels,blood urea, s.creatinine values showed gross rise. Her blood pressure remained in the range of 150/100mm Hg. She was managed aggressively by fresh blood and single donar transfusions ,injection vitamin K 10 mgs. The blood urea, S. creatinine,liver enzymes, S bilirubin values started coming down on 5th post operative day. Her urine output was normal throughout the post operative period. Patient started showing clinical signs of improvement on 6th post operative day.Her platelet count values ,liver function ,renal functions returned to normal on 12 post operative day. Repeat fundus examination showed signs of resolving hypertensive retinopathy. She was discharged on antihypertensive agents on 15th post operative day.

Figure 1 Petechial haemorrhage on skin due to Thrombocytopenia

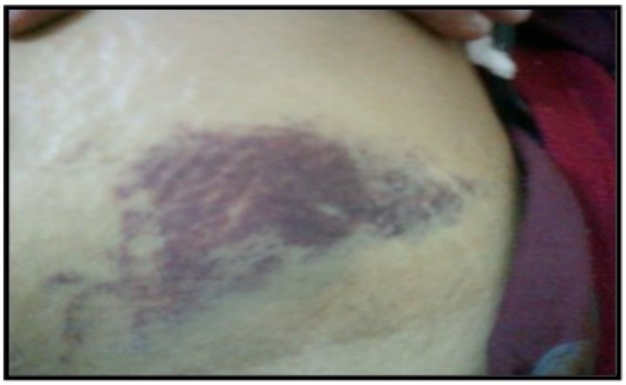
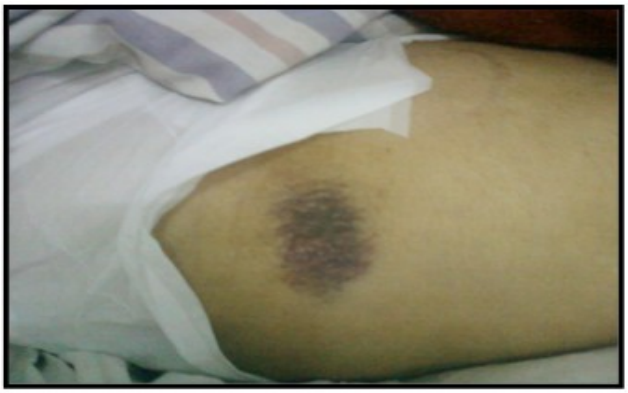


Figure 2 . Injection site ecchymosis due to coagulopathy



3. Discussion

The HELLP syndrome, a serious condition in its complete form, is associated with substantial risk for the mother and her foetus[7-10]. Diagnosis of the complete form of the HELLP syndrome requires the presence of all 3 major components, while partial or incomplete HELLP syndrome consists of only 1 or 2 elements of the triad (H or EL or LP)[11,12]. A wide range of complications may arise and the condition represents diagnostic and therapeutic problems; timing

and method of delivery are important. Haemolysis, one of the major characteristics of the disorder, is due to a microangiopathic haemolytic anaemia (MAHA). Red cell fragmentation caused by high-velocity passage through damaged endothelium appears to represent the extent of small vessel involvement with intima damage, endothelial dysfunction and fibrin deposition. Presence of fragmented (schizocytes) or contracted red cells with spicula (Burr cells) in the peripheral blood smear reflects the haemolytic process and strongly suggests the development of MAHA[13]. Polychromatic red cells are also seen in blood smears, and increased reticulocyte counts reflect the compensatory release of immature red cells into peripheral blood. Destruction of red blood cells by haemolysis causes increased serum lactate dehydrogenase (LDH) levels and decreased haemoglobin concentrations[14]. Haemoglobinaemia or haemoglobinuria is macroscopically recognizable in about 10% of the women[15]. Liberated haemoglobin is converted to unconjugated bilirubin in the spleen or may be bound in the plasma by haptoglobin. The haemoglobin-haptoglobin complex is cleared quickly by the liver, leading to low or undetectable haptoglobin levels in the blood, even with moderate haemolysis[16]. Low haptoglobin concentration (< 1 g/L – < 0.4 g/L) can be used to diagnose haemolysis and is the preferred marker of haemolysis[17]. Thus, the diagnosis of haemolysis is supported by high LDH concentration and the presence of unconjugated bilirubin, but the demonstration of low or undetectable haptoglobin concentration is a more specific indicator.

Elevation of liver enzymes may reflect the haemolytic process as well as liver involvement. Haemolysis contributes substantially to the elevated levels of LDH, whereas enhanced asparate aminotransferase (AST) and alanine aminotransferase (ALAT) levels are mostly due to liver injury. Plasma glutathione S-transferase-a1 (α-GST or GST-a1) may provide a more sensitive indicator for acute liver damage than AST and ALAT, and allow earlier recognition[18]. Thrombocytopenia (platelets (PLTs) < $150{\cdot}10^9$/L) in pregnancy may be caused by gestational thrombocytopenia (GT) (59%), immune thrombocytopenic purpura (ITP) (11%), preeclampsia (10%), and the HELLP syndrome (12%)[19] PLTs < $100{\cdot}10^9$/L are relatively rare in preeclampsia and gestational thrombocytopenia, frequent in ITP and obligatory in the HELLP syndrome (according to the Sibai definition). Decreased PLT count in the HELLP syndrome is due to their increased consumption. Platelets are activated, and adhere to damaged vascular endothelial cells, resulting in increased platelet turnover with shorter lifespan[20].

In general, there are two major options for the management of women with severe preeclampsia and HELLP syndrome. These include: 1) Immediate delivery which is the primary choice at 34 weeks' gestation or later.2) Delivery within 48 hours after evaluation, stabilization of the maternal clinical condition and corticosteroid (CS)treatment. At 27 to 34 weeks of gestation, this option appears appropriate and rational for the majority of cases[21-24].

The present case was attending antenatal clinic regularly at a private nursing home. She developed severe hypertension after 36 weeks of pregnancy. She was put on antihypertensive agents for control of hypertension by the treating obstetrician. Her condition worstened after 37 weeks. The pregnancy should have been terminated by appropriate route at 37 weeks to avoid the subsequent development of HELLP Syndrome. Because the HELLP syndrome can be associated with a bleeding tendency secondary to a deficiency of platelets, it may be necessary to administer platelet transfusions. This may be particularly important before undertaking any surgery, such as a Caesarean section. Too much of conservative approach after 36 weeks in this case has resulted in the deterioration of the condition. Fortunately ,the blood and component therapy was available in the referral institute and the relatives were able to arrange for costly medicines including multiple units of single donar platelets.

4. Conclusion

HELLP syndrome is a life threatening complication of severe pre eclampsia.It is associated with serious risk of coagulation failure, renal failure, hepatic failure and fulminant sepsis.It can be prevented by early recognition of the condition and immediate termination of pregnancy. Management requires multidisciplinary team approach involving anesthesiologist, neonatologist, intensivist, physician and good blood bank facility having availability of component therapy.

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