CASE REPORT

HELLP Syndrome : A Report Of Two Cases

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ABSTRACT

Background: *HELLP Syndrome is a major disorder of the second half of pregnancy that is associated with high risk of maternal and neonatal morbidity and mortality. HELLP is an acronym for Haemolysis, Elevated Liver enzyme levels and Low Platelet Count. As an obstetric complication, it is frequently misdiagnosed at the initial presentation. The purpose of this report is to bring to focus this life threatening disorder that could occur in the second half of pregnancy.*

Methods/Result: *Two cases of HELLP Syndrome are reported. Their conditions were diagnosed and managed accordingly. The first case was delivered by caesarean section while the second case had a vaginal delivery following induction of labour. They fully recovered from the illness and were discharged home from hospital.*

Conclusion: *HELLP Syndrome, being an uncommon major disorder of the second half of pregnancy, early diagnosis is critical to avert a high morbidity and mortality, which have been reported to be as high as 25%.*

KEYWORDS: *HELLP Syndrome; Management.*

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INTRODUCTION

Louis Weinstein first named HELLP Syndrome in 1982 and it is characterized by Haemolysis of the red blood cells, Elevated Liver enzymes and Low Platelet Count[1]. The condition is considered a unique variant of pre-eclampsia but it can occur on its own or in association with pre-eclampsia and can be fatal to both the mother and the baby[2]. The recognition of this syndrome is of value because it draws the clinicians' attention to the wider ramifications of the disease process, which may be present even when blood pressure is not grossly elevated. These patients, regardless of the level of the blood pressure, must be considered as severe disease.

The clinical features of HELLP syndrome are not very specific and could easily be misdiagnosed as peptic ulcer disease, gall bladder disease, acute hepatitis, amoebic hepatic disease, malaria, hyperemesis or acute fatty liver of pregnancy.

CASE REPORTS

CASE 1

A 28-year-old lady Gravida 1, P0 $^{+0}$ at approximately 36 weeks gestation presented with complaints of malaise, epigastric pain, nausea and low-grade fever. Her blood pressure was normal, so was her urinalysis. Repeated antimalarial therapy with chloroquine, fansidar and paludrine were not helpful. Firty-eight hours later, she became febrile with a temperature of 38.4°c and her clinical state worsened. A differential diagnosis of Enteric Fever was made. Therapeutic trial initially with amoxycillin 500mg tds then Ceftriaspone 1gm daily yielded no improvement.

Review of her laboratory indices revealed evidence of haemolysis, [haemoglobin level dropped from 12.4gm to 8.4g/dl], elevated liver enzyme levels [AST. 77 U/L, ALT.84 U/L] and low platelet count [55,000/l]. With these laboratory results, the diagnosis of *HELLP SYNDROME* was made.

An emergency lower segment Caesarean section was carried out and a live male infant was delivered. The patient's condition improved remarkably and all the indices gradually returned to normal. She was discharged home on the 8th post-operative day.

CASE 2

A 29-year-old lady, Gravida 3 Para 2^{+0} at approximately 37weeks gestation presented with history of epigastric pain, nausea/vomiting, headache, fever and restlessness. Her blood pressure was normal and urine analysis was also normal. She was treated for malaria and gastritis with chloroquine and an antacid. She was also given diazepam 5mg as a tranquilizer. On the 3rd day of admission, her condition did not improve and a tinge of jaundice was noticed.

Full blood count (FBC) and Liver function tests(LFT) were done and on review showed low haemoglobin level [6.3gm%], low platelet count [100,000/l] and elevated liver enzymes [AST. 81 U/L, ALT. 92U/L].

From the experience of the case I, the diagnosis of HELLP syndrome was made. The delivery was, therefore, hastened by "ripening" the cervix with 100µg of misoprostol tablet (inserted *per vaginam*) and the labour was augmented with syntocinon infusion. She had a spontaneous vaginal delivery of a live female infant. Her condition improved rapidly

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and was discharged home on the 5th day post-partum.

DISCUSSION

The pathogenesis of HELLP syndrome is not well understood. Sibai *et al* [2] attributed the symptoms and signs of the disease to abnormal vascular tone, vasospasm and coagulation defects, but no common precipitating factor has so far been found. These abnormalities lead to microvascular endothelial damage and platelet activation which excite a cascade of release of thromboxane A and serotonin causing vasospasm, platelet agglutination and aggregation and thus further endothelial damage[2]. The thrombocytopaenia, haemolysis of the red cells and the abnormal liver enzymes are attributed to the cascade of events. They are only terminated with delivery of the baby.

HELLP syndrome occurs in approximately 0.2 to 0.6 % of all pregnancies[3]. Although it commonly occurs in the 3rd trimester of pregnancy, in 11% of patients it occurs at less than 28weeks of gestation[4]. Both of our patients presented in the 3rd trimester-36weeks and 37weeks. Most of the reported cases presented ante partum (69%) and an estimated 31% presented post partum[5].

HELLP syndrome presents vaguely. A high index of suspicion is required to make a diagnosis. The usual presenting complaints are malaise (90%), epigastric discomfort/pain (65%), nausea and vomiting (30%) and headache and fever (31%)[3]. In this environment, the differential diagnosis includes malaria, typhoid fever, hepatitis, peptic ulcer and cholecystitis. Early diagnosis of this syndrome is critical as late diagnosis is detrimental to both the fetus and the mother. In our reported cases, the diagnosis was initially missed in Case 1 but in Case 2, the diagnosis was made easier by the experience gained from Case 1. As a safe guard, any pregnant woman who presents with malaise or viral-like illness in the 3rd trimester should be evaluated with a complete blood count and liver function tests[6].

The main stay in making a diagnosis of this condition are the abnormal laboratory findings. The haemoglobin level drops when compared with levels during the antenatal visits. The serum ransaminase levels are usually elevated. The levels in our patients got up to 140 units/L and 77 units/L in cases 1 and 2 respectively. The platelet levels in our patients dropped to 100,000/l and 50,000/l in the 2 cases. The platelet count is the best indicator of HELLP syndrome and should be suspected in any patient who shows a significant drop in the platelet count during the antenatal period[7].

HELLP syndrome could be classified into 2 types. The first is based on the number of abnormalities observed[8] and the second is based on the platelet count[9]. In the first type, patients are said to have partial HELLP syndrome, when they have one or two abnormalities but when all three abnormalities are present it is a full HELLP syndrome. Our patients had the three abnormalities of haemolysis, elevated liver enzymes and low platelet count, hence the full HELLP syndrome.

The second type is further divided into class I with platelet count less than 50,000, class II with count from 50,000 to 100,000 and the class III with count from 100,000 to 150,000. Patients with class I HELLP syndrome are at a higher risk for maternal morbidity and mortality than classes II and III[5].

Our cases were in the class II and III groups and recovered fully from the illness.

Weinstein[1] in his description recommended a prompt delivery on making the diagnosis of HELLP syndrome but recent researchers suggest that morbidity and mortality do not increase when these patients are treated conservatively[10]. Conservative measures are an advantage to the infant, since prolongation of the pregnancy means less time in the neonatal intensive care unit, a decrease incidence of respiratory distress syndrome and a decrease incidence of necrotizing enterocolitis[11]. The management approach should be based on the estimated gestational age and the condition of the mother and fetus[12]. Although, in the past and currently in most centres, patients with HELLP syndrome are delivered routinely by Caesarean Section, this is now recommended for patients with severe disease, or with superimposed disseminated intravascular coagulopathy [DIC] or a gestation of less than 32 weeks[2]. Patients with mild to moderate disease and are at 32 weeks of gestation or more and have a favourable cervix should have a trial of labour[2].

High dosage of corticosteroids, 10mg dexamethazone given intravenously 12 hourly, has been shown to markedly improve the abnormal laboratory indices associated with HELLP syndrome[13]. These abnormalities resolve faster in those who continue to receive the steroids postpartum[7].

It is recommended that patients with HELLP syndrome should be treated prophylactically with magnesium sulphate to prevent seizures, whether hypertension is present or not and those with hypertension should be treated with

antihypertensive drugs[14].

Maternal complications include abruptio placentae, hepatorenal failure, hepatic rupture, pulmonary oedema, adult respiratory distress syndrome, pleural effusion, DIC and death[2]. Patients with platelet count less than 20,000 x 10^9 should receive blood and blood products especially those undergoing Caesarean Section.

Finally, patients who have had HELLP syndrome have a 19-27% risk of developing the disease in subsequent pregnancies and 43% risk of developing preeclampsia in another pregnancy[15].

CONCLUSION

HELLP syndrome is an uncommon life-threatening complication of pregnancy and does not have an abrupt onset. Clinicians must therefore be vigilant to recognize the early signs and prognostic indices of the disease so as to initiate prompt treatment or referral to tertiary centre to reduce maternal and perinatal morbidity and mortality.

REFERENCES

1. Weinstein L. Syndrome of hemolysis, elevated liver enzymes and low platelet count: a severe consequence of hypertension in pregnancy. Am J Obstet Gynecol 1982;142(2):159-67.
2. Sibai BM, Ramadan MK, Usta I, *et al.* Maternal morbidity and mortality in 442 pregnancies with hemolysis, elevated liver enzymes and low platelets (HELLP syndrome). Am J Obstet Gynecol 1993;169:1000-6.
3. Wolf JL. Liver disease in pregnancy. Med Clin North Am 1996;80:1167-87.
4. Gleeson R, Farrell J, Doyle M, *et al.* HELLP syndrome: a condition of varied presentation. Ir J Med Sci 1996; 165:265-7.
5. Sibai BM. The HELLP syndrome (hemolysis, elevated liver enzymes and low platelets): much ado about nothing? Am J Obstet Gynecol 1990;162:311-6.
6. Tomsen TR. HELLP syndrome (hemolysis, elevated liver enzymes and low platelets) presenting as generalized malaise. Am J Obstet Gynecol 1995;172:1876-90.
7. Magann EF, Perry KG Jr, Meydrech EF, *et al.* Postpartum corticosteroid: accelerated recovery from the syndrome of hemolysis, elevated liver enzymes and low platelets (HELLP). Am J Obstet Gynecol 1994;171:1154-8.
8. Audibert F, Friedman SA, Frangieh AY, *et al.* Clinical utility of strict diagnostic criteria for the HELLP (hemolysis, elevated liver enzymes and low platelets) syndrome. Am J Obstet Gynecol 1996;175:460-4.
9. Martin JN Jr, Blake PG, Lowry SL, *et al.* Pregnancy complicated by preeclampsia-eclampsia with the syndrome of hemolysis, elevated liver enzymes and low platelet count: how rapid is postpartum recovery? Am J Obstet Gynecol 1990;76(5 pt 1):737-41.
10. Visser W, Wallenburg HC. Temporising management of severe pre-eclampsia with and without the HELLP syndrome. Br J Obstet Gynaecol 1995;102:111-7.
11. Sibai BM, Mercer BM, Schiff E, *et al.* Aggressive versus expectant management of severe preeclampsia at 28 to 32 weeks' gestation: a randomized controlled trial. Am J Obstet Gynecol 1994;171:818-22.
12. Sibai BM, Frangieh AY. Management of severe preeclampsia. Curr Opin Obstet Gynecol 1966;8:110-3.
13. Magann EF, Bass D, Chuhan SP, *et al.* Antepartum corticosteroids: disease stabilization in patients with the syndrome of hemolysis, elevated liver enzymes and low platelets (HELLP). Am J Obstet Gynecol 1994;171:1148-53.
14. Waisman BD, Mayorga LM, Camera MI, *et al.* Magnesium plus nifedipine: potentiation of hypotensive effect in preeclampsia? Am J Obstet Gynecol 1988;159:308-9.
15. Sullivan CA, Magann EF, Perry KG, *et al.* The recurrence risk of the syndrome of hemolysis, elevated liver enzymes and low platelets (HELLP) in subsequent gestations. Am J Obstet Gynecol 1994;171:940-3.