**Case Reports**

# Two cases of HELLP syndrome with fatal outcomes

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**Summary**

Severe preeclampsia and HELLP syndrome are still the leading causes of maternal and perinatal morbidity and mortality. We present two cases of pregnancies which were complicated by HELLP syndrome at 31 weeks of gestation and 25 weeks of gestation, the first one with maternal and the second with perinatal fatal outcomes. The aim of this report is to draw attention to the life-threatening complications that might occur in cases of preeclampisa and HELLP syndrome. The importance of early diagnosis with implications for management is also discussed.

*Key words:* Hellp syndrome; Outcome; Early diagnosis.

## Introduction

HELLP syndrome (hemolysis, elevated liver enzymes, and low platelets) has been recognized as a complication of preeclampsia/eclampsia for decades [1]. Recognition of this syndrome in women with preeclampsia is increasing because of the frequency of blood test results that reveal unexpected thrombocytopenia or elevated liver enzymes [1, 2]. During the past decade the increasing awareness of obstetricians and physicians in other disciplines have led to a significant reduction of maternal mortality (< 1%) and perinatal mortality (9.4-16.2%) in cases of HELLP syndrome [2, 3]. Because of the high risk of maternal and perinatal mortality, early diagnosis, aggressive treatment with maternal stabilization and expeditious delivery have been advocated [3]. In recent years, many studies have been published in an attempt to refine the diagnostic criteria, to identify risk factors for adverse pregnancy outcome, and to treat women with this syndrome. Despite the voluminous literature, the diagnosis and management of HELLP syndrome remain controversial [1-3].

We present two pregnancies complicated by HELLP syndrome aiming to draw attention to the life-threatening complications that might occur. The importance of early diagnosis with implications for management are also discussed.

## Case 1

A 29-year-old woman, gravida 3, para 2, had no history of hypertension or vascular disease before pregnancy. Her current pregnancy course was uneventful. At 31 weeks' gestation, she had pretibial edema and proteinuria with hypertension (170/110 mmHg). After she was hospitalized, she began complaining of headache and epigastric pain. Her laboratory data revealed mild elevation of transaminase (aspartate aminotransferase (AST), 50 U/l, alanine aminotransferase (ALT), 30 U/l, lactic acid dehydrogenase (LDH), 510 U/l, and platelets 143,000/mm$^3$). Immediate stabilization of the mother's condition by means of anticonvulsive prophylaxis with intravenous magnesium sulphate and well controlled reduction of blood pressure by the administration of nifedipine was started. Her vital signs and urinary output were monitored. She underwent emergency cesarean section because of a non-reassuring fetal heart rate pattern. A female infant, weighing 1,330 g, with Apgar scores 1 and 5, at 1 and 5 min, respectively, was delivered. The mother remained hypertensive (185/115 mm Hg) after the operation, and her laboratory tests showed markedly elevated transaminases and lowered platelets (AST 530 U/l, ALT 382 U/l, LDH 1100 U/l, platelets 62,000/mm$^3$). She was diagnosed with HELLP syndrome. At that time she was clearly conscious and aware. About ten hours later, she developed oliguria and her blood pressure was 160/100 mmHg and laboratory tests revealed further progression of HELLP syndrome (AST 831 U/l, ALT 503 U/l, LDH 3120 U/l, platelets 38,000/mm$^3$). Controlled volume expansion was done. Fresh frozen plasma was given for the treatment of coagulation disorders. Platelet transfusion was given to correct the trombopenia. We had planned to give 12 mg dexamethasone every 12 hour for two doses but she fell into a stuporous state 16 hours after admission and was transferred to the intensive care unit. Unfortunately, she rapidly developed a comatose condition and died one hour after the onset of coma. Because of the immediate and serious development of her shock condition, there was no chance to get a computed tomography scan to exclude possible intracranial hemorrhage.

## Case 2

A 36-year-old woman, gravida 2, para 0, abortion 1, was referred to our hospital at 25 weeks of gestation because of intrauterine growth restriction, hypertension (200/120 mmHg) and proteinuria. Her past medical history revealed antiphospholipid syndrome and an embolic cerebrovascular event four years

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before. She had been on anticoagulant therapy since that time and also during the pregnancy. Her laboratory data revealed elevation of transaminase and lowered platelets AST, 192 U/l, ALT, 221 U/l, LDH, 1191 U/l, platelets 86,000/mm$^3$. She was diagnosed with HELLP syndrome and immediate stabilization of the mother's condition was initiated. Anticonvulsive prophylaxis with intravenous magnesium sulphate and well controlled reduction of blood pressure by administration of nifedipine were started. Her vital signs and urinary output were monitored. Oxytocin induction was started to terminate the pregnancy. Unfortunately the fetus died intrapartum. A female infant, weighing 410 g, was delivered. The postpartum course of the mother was uneventful. Her transaminases and platelet counts returned to normal levels gradually within three days.

## Discussion

The ultimate goal of any protocol for the management of HELLP syndrome is to reduce maternal mortality and morbidity rates, followed by delivery of a healthy and full-term baby [3, 4]. Since 1982, maternal as well as perinatal mortality has decreased due to careful assessment of the maternal and fetal status [5]. An important aspect of patient care is early diagnosis. Cases that are diagnosed and referred to an intensive care unit at a late stage of disease have a very poor prognosis [4]. In our first case, if we could have followed-up the patient in the intensive care unit earlier the prognosis might have been better. The diagnosis of HELLP syndrome requires the presence of hemolysis based on examination of the peripheral smear, elevated indirect bilirubin levels, or low serum haptoglobin levels in association with significant elevation of liver enzymes and a platelet count below 100,000/mm [3] after ruling out other causes of hemolysis and thrombocytopenia [1].

In Sibai *et al's.* series maternal death in HELLP syndrome was due to various complications: one from ruptured liver hematoma, one from pulmonary embolism, and three from diffuse hypoxic encephalopathy [5]. Our first case most probably died beceause of intracranial bleeding, but a cranial CT was not performed. Raval *et al.* reported a maternal death from disseminated intravascular coagulation [6], and Weinstein [7] reported a maternal death in a patient with severe microangiopathic hemolytic anemia, marked hyperbilirubinemia, and massive ascites.

Pregnancy affected by severe HELLP syndrome is an extremely high-risk situation, therefore these patients must be followed-up and managed in critical care units. Aggressive management can significantly change the outcome for these patients [3].

Patients whose pregnancies are complicated by HELLP syndrome are at a higher risk of renal failure, consumptive coagulopathy, abruptio placentae, pulmonary and cerebral edema, subcapsular liver hematoma, and hypovolemic shock [8]. As HELLP syndrome frequently occurs before term with an unfavorable cervix, cesarean section is a common mode of delivery, with rates ranging from 42% to 98% [5]. Disseminated intravascular coagulation (DIC) has been described in up to 15% of patients, half of whom had placental abruption [3]. Wound hematoma or infection is a common complication after cesarean section and presents in 7% to 14% of HELLP syndrome patients [3]. Acute renal failure has been reported as a complication in 8%, the majority secondary to acute tubular necrosis. Ecclampsia is seen in 4% to 9% of HELLP patients [5]. Other complications reported include severe ascites (8%), pleural effusion (6%), pulmonary edema (6%), and subcapsular liver hematoma requiring laparotomy (1%) [5].

Infant mortality rates range from 5% to nearly 20% [5]. Neonatal survival in infants born to mothers with HELLP syndrome is mainly dependent on gestational age and birth weight at delivery. It does not appear that HELLP syndrome, independent of gestational age, increases neonatal mortality [5].

Nevertheless in our second case the baby died, most probably beceause it was only a 25-week-old fetus with IUGR. Weinstein reported hematologic abnormalities (thrombocytopenia, leukopenia, and abnormal peripheral smears) in neonates born to mothers with HELLP syndrome [3]. Neonatal thrombocytopenia has been reported in up to 50% of pregnancies complicated by HELLP syndrome, but no correlation has been demonstrated between maternal and neonatal platelet counts. Infants affected by HELLP syndrome are more likely to experience intrauterine growth restriction, like in our second case, and respiratory distress syndrome [3].

To achieve a decrease in maternal mortality from HELLP syndrome we emphasize the importance of an early and correct diagnosis of the syndrome which improves maternofetal prognosis. The high maternal and perinatal mortality and morbidity reported with the presence of HELLP syndrome requires maternofetal follow-up in a tertiary center where intensive maternal and neonatal care are available [3, 7]. The timely allocation to a perinatal center and intensive monitoring of mother and child after admission are mandatory for successful management of these patients [1, 2]. The aim of therapy is immediate stabilization of the mother's condition by means of anticonvulsive prophylaxis with intravenous magnesium sulphate and well controlled reduction of blood pressure [9]. Also controlled volume expansion and adequate treatment of coagulation disorders by giving fresh frozen plasma (not heparin) should be carried out. Immediate delivery is the method of choice in cases of HELLP syndrome at a gestational age of 34 months or more [1, 8, 9].

Recent studies suggest that some women with partial HELLP syndrome may be treated with expectant management or corticosteroid therapy. We had planned to give dexamethasone to our first case but she only received the first dose of corticosteroids [1, 10, 11]. However, expectant management in patients with HELLP syndrome remote from term and the use of corticosteroids to improve postpartum maternal outcome remain experimental [1, 11].

We suggest more aggressive and intensive care of HELLP syndrome patients. Management requires careful obstetric supervision and should be multidisciplinary.

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