CEOG Clinical and Experimental Obstetrics & Gynecology

Severe ascites as the primary symptom of fulminant postpartum HELLP syndrome: a case report

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Summary

Previous studies show that HELLP syndrome occurring in postpartum period was more dangerous to both fetus and the mother than if presented before delivery. So far, there is a lack of screening or predictive tests for postpartum HELLP syndrome in clinical practice. *Case:* Here the authors report a rare case of postpartum HELLP syndrome with severe ascites as the primary symptom. The patient was diagnosed with severe preeclampsia and fetal growth retardation at 34+2 weeks gestation and received anti-hypertensive therapy. Severe ascites were found intraoperatively during emergency caesarean section. On the second postoperative day, complete HELLP syndrome was diagnosed. This case gradually complicated by hypoproteinemia, acute renal failure, severe anaemia, and infection and required renal haemodialysis, blood transfusion, and other supportive treatments for about one month. *Conclusion:* Although this case has a fulminant and long course, it has a well clinical prognosis and also shows that severe ascites may be a clue for postpartum HELLP syndrome in patient with severe preeclampsia.

Key words: HELLP syndrome; Sever ascites; Renal failure; Severe preeclampsia; Postpartum.

Introduction

The acronym of HELLP syndrome was devised by Weinstein in 1982 and was a serious, life-threatening severe preeclampsia complication [1]. H stands for hemolysis indicating microangiopathic hemolytic anemia, EL for elevated liver enzymes indicating a pathological increase of liver enzymes, and LP for low platelet count indicating thrombocytopenia. HELLP syndrome afflicts many pregnant women and results in a large percentage of maternal and perinatal complications [2]. Maternal mortality of the syndrome is 3.9% and perinatal mortality varies between 7.7% and 37% in worldwide [3]. Selcuk *et al.* reported that HELLP syndrome is the most frequent cause of acute renal failure in pregnancy [4].

The most common used classifications of HELLP syndrome were developed at the Universities of Tennessee and Mississippi. The Tennessee Classification [5] defined the "complete HELLP syndrome" which met all of the following criteria: (1) platelets 100,000/ml or less, (2) AST 70 IU/L or greater, and (3) abnormal peripheral smear in addition to either total serum LDH 600 IU/L or greater or bilirubin 20.4 umol/L or greater. The patient who displays some but not all of these was defined as "partial HELLP syndrome ".The syndrome also can be divided into three classes or groups primarily according to the platelets' count [6]: class I requires severe thrombocytopenia (platelets' count is less than 50,000/ml), class II requires moderate thrombocytopenia platelets' count is 50,000 to less than 100,000/ml), and class III requires mild thrombocytopenia when (platelets' count is 100,000 to 150,000/ml).

This syndrome accounts for 0.17% to 0.85% of all live births [7]. Preeclampsia or eclampsia can increase the incidence of the syndrome [8]. The classical presentation of HELLP syndrome is right upper quadrant pain or epigastric pain accompanied with malaise, nausea or vomiting, and these symptoms accounts for 82-96% of the cases [9]. When the condition arises in the postpartum period, it will pose more serious risk to both fetus and the mother than before delivery [10]. Therefore, it is important for clinicians to make an accurate diagnosis of the syndrome to prevent any adverse outcome. Its occurrence is rare in the postpartum period, even rarer without any malaise except for severe ascites.

Woods *et al.* reported that severe ascites portend a patient at high risk for cardiopulmonary complications [11]. However, severe ascites as the primary symptom of HELLP syndrome is rare. Here the authors report a class I fulminant postpartum HELLP syndrome with severe ascites as a primary symptom complicated by renal failure in a patient who was under intensive care for severe preeclampsia in obstetrics department.

Case Report

The patient was a 35-year-old women, Yellow race, gravida 3, para 1. Her antenatal care was conducted in the maternity and children's healthcare center in Xindu City. She was seen regularly by the doctors at this center. The care was uneventful until 30 weeks

Revised manuscript accepted for publication June 25, 2014

Clin. Exp. Obstet. Gynecol. - ISSN: 0390-6663
XLII, n. 5, 2015
doi: 10.12891/ceog1939.2015

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plus six days gestation when the doctor found her blood pressure was 189/120 mmHg and prescribed nifedipine and magnesium sulfate for her in the center for seven days. Then, she went home and did not continue her therapy. In this period, her blood pressure was about 150/90 mmHg (measured by her family members). After 10+ days, she went to the center again and the doctor referred the patient to the present hospital with blood pressure fluctuated at a high level (178-203/113-120 mmHg) and 3+ of proteins in the urine. The patient was admitted to the present hospital at 34 weeks plus two days gestation with a chief complaint of elevated blood pressure and albuminuria. She had no past medical or surgical history. On admission, her blood pressure was 180/120 mmHg and her the reflexes were brisk. The blood cell count, creatinine, serum urea nitrogen, urates, albumin, and creatinine clearance rate was normal with only mild raised liver enzymes. Both lower extremities showed mild pitting edema and the uterine fundus height was less than that of a date. Obstetric ultrasound showed that biparietal diameter and femur length of fetus was 7.7 cm and 5.9 cm, respectively, which was below the 5th percentile for the gestational age. The ultrasound test did not find any ascites. Overall, the diagnoses were severe preeclampsia and intrauterine growth retardation. The authors administered intensive care in obstetrics department and ten mg of nifedipine three times per day, magnesium sulfate, and ten mg of dexamethasone once per day for three days. However, the blood pressure also ranged between 140/100 and 170/120 mmHg.

Continuous fetal monitoring showed that the non-stress test (NST) indicated fetal distress. Therefore, the authors give the patient an emergency lower segment caesarean section immediately after ten days from admission. The process of the operation was smooth except for about 2,000 ml clear yellowish ascites which was inconsistent with mild pitting edema in her both lower extremities. The patient gave birth to an unhealthy baby with Apgar's scores (3-5-7/1-5-10 minutes) and a birth weight of 1,730 g and the baby required neonatal care. The baby was then transferred to Children's Centre of Chengdu. The patient had a postpartum haemorrhage of 300 ml. In this operation the authors give the patient 1,500 ml liquid (normal saline + colloid).

After operation the patient was asymptomatic and had normal blood cell count (Hb:126 g/L: PLT:100×10^9/L), creatinine, serum urea nitrogen, urates, creatinine clearance rate, and liver enzymes were also normal with ALT: 54 IU/L, AST: 44 IU/L, GGT: 336 IU/L, ALP: 165 IU/L, TBA: 2.9 IU/L and the urine output was normal. However, the serum albumin dropped to 26.4 g/L. The blood pressure of the patient was still at a high level and the highest was 180/120 mmHg. The authors called cardiovascular specialist for a consult who suggest to give the patient furosemide and double increment of nifedipine dosage to control hypertension. At the end of the first post-operative day, the urine of the patient became haemorrhagic with reduced output and complained of abdominal distension. Six hours urine was only ten ml. The emergency accessory examination showed hyponatremia and the platelets' count dropped to 51 and serum albumin dropped to 24.6 g/L and raised liver enzymes (ALT: 113 IU/L, AST: 253 IU/L), and total bilirubin (20.4 umol/L), WBC (28.2×10^9/L), and creatinine (412.5 umol/L). The authors asked all related departments in the hospital for an immediate consult .

On the morning of the second postoperative day, the liver enzymes continued to rise and the platelets' count dropped to 23 and hemoglobin dropped to 98 g/L. A peripheral blood smear showed blood cell shrink, damage, and heterocyst which indicated microangiopathic haemolysis in the patient's body. HELLP syndrome was diagnosed. The patient was still anuria and went into renal failure which required haemodialysis (CRRT). Therefore, the authors transferred the patient to the general ICU. In general ICU the patient was given ten mg of dexamethasone twice per day for two days, antibiotics, liquid infusion, plasma transfusion, and haemodialysis (CRRT) for about a week and the liver functions gradually normalized. However, the platelets' count reduced to 21 and hemoglobin dropped to 54 g/L. The patient still had severe ascites. The authors give the patients packed red cells, fresh frozen plasma, and platelet transfusion and ascites drainage. One month later, the renal function and the blood profile were normal and then the patient was discharged home in a good clinical condition.

Discussion

Although the reported incidence of HELLP syndrome has considerable differences among the world [8], China has a lesser number of people that suffer from this condition, and it accounts for about 0.11% of all pregnancies and approximately 1.03% of pregnancies with hypertension [12]. Among severe preeclampsia cases, this condition comprises 10% and approximately 50% in all eclampsia cases [10]. However, these data cannot accurately represent the Chinese population. To the best of the present authors' knowledge, there was no study focus on the incidence of HELLP syndrome among Chinese pregnancies complicated by preeclampsia or eclampsia.

The majority of HELLP syndrome occur between 32 and 34 weeks' gestation. Signs and symptoms with HELLP syndrome are variable which depend largely on the stage of the patient's disease. Usually, it manifests in the right upper quadrant or epigastric pain, nausea and/or vomiting or malaise [6]. Only about 13-30% manifests itself on postpartum up to six days after delivery [9, 12]. The present case was diagnosed with sever preeclampsia before delivery, on the first day after cesarean section, and the patient had acute renal failure. Both sever ascites and acute renal failure complicating HELLP syndrome is not infrequent, in the Rath *et al.* review, the incidence was 8% respectively. However, in all cases of the syndrome generalized edema account for 50%-69% [9, 13]. Some doctors argue that severe ascites are caused by hypoproteinemia which may be a reason for the ascites according to the clinical experience. However, it was not inconsistent with general edema. Currently we are still not sure of the accurate etiology of the ascites and the difference between generalized edema and severe ascites. Perhaps severe ascites are predictors of postpartum HELLP syndrome.

HELLP syndrome cases with low platelet counts have been found to be at increased risk for adverse maternal outcomes. Among all the deaths in patients with HELLP syndrome, 60% of cases were class I [2, 14]. The present case was a complete and classs I HELLP syndrome, however at the beginning, it was not typical of complete HELLP syndrome which is sometimes referred to as partial HELLP syndrome.

According to the present authors' previous knowledge, the only cure for HELLP syndrome is delivery and all the other treatments are palliative, even at early gestational ages. They also discuss the early delivery for this case in the present hospital. However, pediatricians in the hospital did not suggest

to conduct the delivery for the safety of the baby according to the poor conditions of neonatal care in the present district. Basama *et al.* also reported a postpartum HELLP syndrome case and they reported that this type had casted some doubt about the traditional idea and highlighted that there are no current means of prediction and early detection the syndrome [15]. The present case is consistent with their statement, but in the present case severe ascites may have been the early prediction of the syndrome. Then the present authors recognised the increased of the liver enzymes (AST, ALT), anuria, and acute renal failure. They also observed hematuria which indicated haemolysis in this case.

The pathophysiology of HELLP syndrome is still unknown; among forming hypothesises, role of imbalance between vasoconstrictive and vasodilation hormones was very important in this condition. Previous study showed that abnormal placentation resulting in placental ischemia and then produced some circulating toxic factors which react aggressively with endothelium causing endothelial injury, especially the increased production of the vasoconstrictor thromboxane A2 [16]. These toxic factors can promote platelet aggregation and results in endothelial lesions, local vascular constriction, and thrombocytopenia. In the present case, these vasoconstrictive substances may contribute to the difficulties in controlling blood pressure. The acute renal failure and severe ascites in the present case can be explained by the fact that endothelial lesions mainly occur in organs with high blood flow and lead to severe maternal complications.

Even though a recently systemic review shows that there is no sufficient evidence to support using the corticosteroids for HELLP syndrome [17], the present authors still used aggressive corticosteroids treatment besides symptomatic treatment in this case according to some studies that concluded that aggressive corticosteroids clearly benefit the mother with HELLP syndrome according to stage of disease, especially for the cases of class I or II [14].

Conclusion

In summary, the authors reported a rare case of postpartum HELLP syndrome with severe ascites as the primary symptom. Although the case had a fulminant and long course, it had a well clinical prognosis with multidisciplinary ICU approaches. So far, there is a lack of screening or predictive tests for postpartum HELLP syndrome in clinical. This case shows that severe ascites may be a clue for postpartum HELLP syndrome in patient with severe preeclampsia.

Acknowledgments

The authors are grateful to the staff of the Department of Obstetrics and Gynaecology and General ICU, the First Affiliated Hospital of Chengdu Medical College for their work. They thank the patient and her legal representative, both of them agreed and supported us to report this medical condition. At last, they also would like to thank Dr. Qiukui Hao in West China Hospital for editing the English language in this article.

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