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Case report

A rare case of normotensive HELLP syndrome complicated with massive ascites: Spontaneous resolution

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KEYWORDS

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Abstract HELLP develops in approximately 0.1–0.8% of pregnancies overall and as many as 15–20% of patients with HELLP syndrome do not have antecedent hypertension or proteinuria. The risk factor for development of ascites is extensive structural damage of the microvasculature in patients complicated by HELLP. The aim of this study is to report a case with HELLP syndrome complicated with massive ascites after vaginal delivery that eventually resolved spontaneously.

1. Introduction

HELLP is an acronym that refers to a syndrome characterized by Haemolysis with a microangiopathic blood smear, Elevated Liver enzymes, and a Low Platelet count [1]. It probably represents a severe form of preeclampsia, but the relationship between the two disorders remains controversial. HELLP develops in approximately 0.1–0.8% of pregnancies overall and as many as 15–20% of patients with HELLP syndrome do not have antecedent hypertension or proteinuria, leading some authorities to believe that HELLP is a separate disorder from preeclampsia [2]. Both severe preeclampsia and HELLP syndrome may be associated with serious hepatic manifestations, including infarction, haemorrhage, and rupture.

A previous history of preeclampsia or HELLP is a risk factor for HELLP syndrome. A variety of genetic variants associated with an increased risk of HELLP syndrome have been reported, but have no role in clinical management. In contrast to preeclampsia, nulliparity is not a risk factor for HELLP syndrome [3,4]. Half or more of affected patients are multiparous.

In preeclampsia patients, capillary leakage may be associated with abnormal collections of fluid in the pleura or peritoneum. The most important factor for its development seems to be reduced PCOP of proteins. However, it has been documented that increased mean arterial pressure, extensive structural damage of the microvasculature in patients complicated by HELLP syndrome, intracapillary coagulopathy, platelet count $\leqslant$100,000/mm, proteinuria $>$5 g/24 h, serum creatinine (Cr) $\geqslant$120 mOsm/l, portal hypertension, presence of acute cardiogenic pulmonary oedema, uncontrolled systolic arterial pressure (SAP) and adult respiratory insufficiency syndrome (ARIS) may favour their formation [3,5,6].

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The aim of this study is to report a case with HELLP syndrome complicated with massive ascites after vaginal delivery that eventually resolved spontaneously.

2. Case report

28 years old, G2P1, with history of normal vaginal delivery and one living child was admitted in our emergency department at 36 weeks of gestation in labour. She was irregular in her antenatal checkups and last visit was since two months. Her antenatal investigations and the blood pressure recordings were normal. The first and second trimester ultrasound scans were corresponding to the period of gestation.

On presentation, she has an icteric tinge in the eyes with no history of blood or hepatic disease and with no history suggestive of impending eclampsia. The examination of the patient showed, pulse rate of 80/min and blood pressure recording of 100/70 mmHg. The abdominal examination showed a nearly full term sized uterus with longitudinal lie and breech presentation. The per vaginal examination showed fully dilated cervix MI breech deeply engaged. The urine examination revealed proteinuria + + + with dark liquorice urine.

She delivered a viable female baby by normal vaginal delivery. There was no evidence of retro-placental haemorrhage.

Immediate postoperative period was uneventful. Her blood pressure was recorded 100/70 mmHg and pulse of 90 beat/min. The investigations at this time showed proteinuria + + +, platelet count of 60,000/cubic millimetre, serum bilirubin 3.8 mg%, with direct bilirubin 3 mg%, ALT and AST were 100 and 150 respectively, INR 5, serum uric acid 9.5 mg%, serum BUN 22 mg%, serum creatinine 2.5 mg%, HB 7 g%, serum albumin 0.9 g%, serum LDH was 400 IU/l and there was evidence of haemolysis in the peripheral smear. The patient had a normal UOP. She was suspected to be developing HELLP syndrome and was transfused with fresh frozen plasma and blood. The patient was started on injection dexamethasone 24 mg/day in three divided doses for the benefit in cases of HELLP syndrome. The patient developed distension of abdomen (Fig. 1). So, she was started on injection frusemide to ensure adequate urinary output, and the central venous pressure was normal. The ultrasonography showed huge ascites with normal liver scan (Fig. 2). The echocardiography showed no pericardial effusion with normal ejection fraction. Her follow-up laboratory investigations were as follows: HB 3.5 g%, PLT 50,000 cell, cubic millilitre, PT 30%, INR 2, serum BUN 23 mg%, serum creatinine 3 mg%, serum albumin 1 g%, serum AST ALT 80, 90 respectively. Her vital signs were stable. So, blood, FFP were continued to be transfused and also the steroids.

The patient general condition started to resolve. The patient had adequate urine output. The CVP was normal. She continued to have the similar pulse and BP records. The follow-up investigations were HB 8 g% that was normalized over 2 days, BUN, S bilirubin, ALT and AST were normal. The peripheral blood smear was negative for haemolysis and platelet count 70,000, PT 100, INR 1, serum albumin 1.5 g %, SGOT SGPT 30, BUN 20 mg%, serum creatinine 2 mg %, furosemide discontinued. She was discharged from the hospital on tab prednisone with the instructions for weekly follow-up at 2 weekly follow-up at gynaecology. She became normal after 10 weeks.

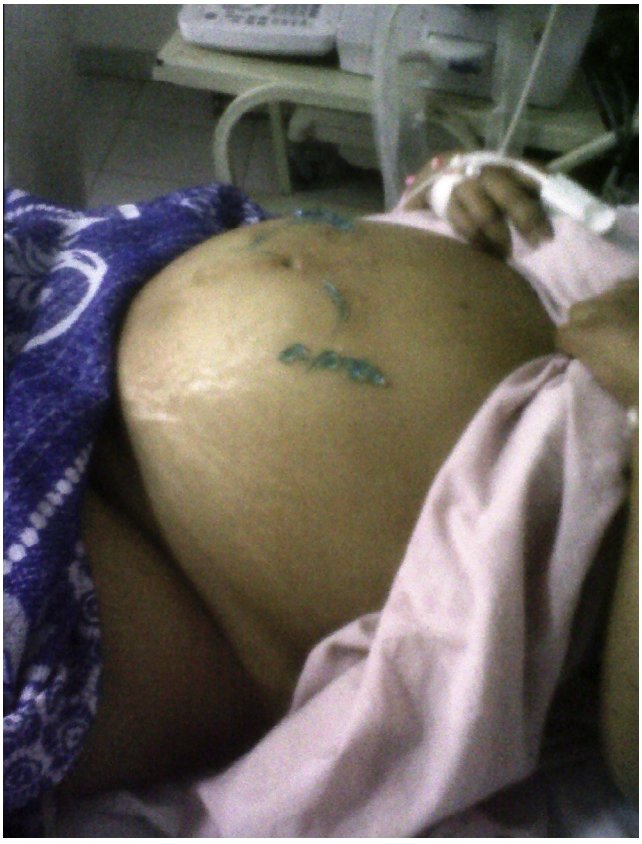

Figure 1 Abdominal distension postpartum after vaginal delivery.

3. Discussion

Pre-eclampsia is a condition involving all the organ systems. The basic patho-physiology involves the release of vasoconstrictive agents, endothelial damage, hyperpermeability of the capillaries and microangiopathic haemolysis. This results in the presentation of the condition in various forms of hypertension, HELLP syndrome, visual symptoms, seizures, pulmonary oedema, proteinuria, etc. [7].

Ascites has been reported at caesarean section in severe pre-eclampsia cases. The analysis of this fluid has showed it to be transudate [7]. The exact aetiology of the same is not known, but intrahepatic portal hypertension is one possible explanation [8]. The case presented above had a varied presentation with the collection of the fluid in the abdominal cavities and HELLP syndrome.

Woods et al. [9] in a retrospective study found, the incidence of large-volume ascites in patients with HELLP syndrome who had caesarean was about 10%. They compared HELLP syndrome patients without ascites, to those with HELLP associated ascites and found sixfold increase in the incidence of congestive heart failure and a ninefold increase in the incidence of adult respiratory distress syndrome. In a series of cases reported the requirement of early termination of pregnancy in cases of women with ascites in pre-eclampsia. They observed a rapid deterioration in maternal condition with rise in blood pressure and increasing proteinuria.

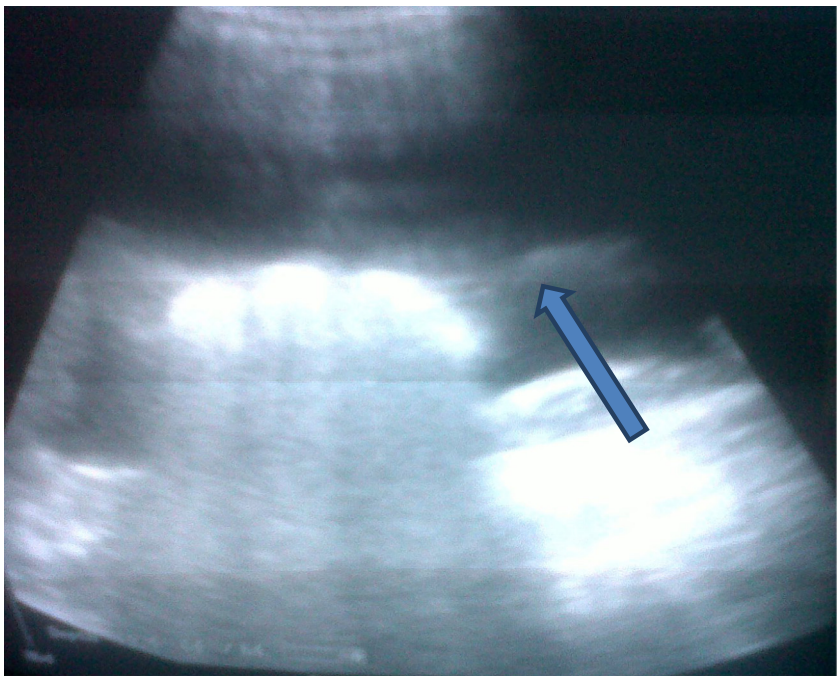

Figure 2 Ascites on ultrasound.

In most women with a HELLP syndrome, the maternal PLT counts continue to decrease immediately post-partum with an increasing trend on the third day [10]. About 30% of the HELLP syndromes develop after birth, the majority within the first 48 h. However, the time of onset might range from a few hours to 7 days after delivery [10]. In women with post-partum HELLP syndrome, risk of renal failure and pulmonary oedema is significantly increased compared to those with an antenatal onset. Since early post-partum administration of high dose CS might accelerate recovery, its routine administration is highly advocated (10 mg of dexamethasone every 12 h) [11].

However, a randomized study showed that adjunctive use of intravenous dexamethasone for postpartum patients with severe preeclampsia did not reduce disease severity or duration [12]. Moreover, the benefit of dexamethasone in post-partum HELLP syndrome was not verified in a randomized placebo controlled trial on 105 women with postpartum HELLP syndrome. There was no difference in maternal morbidity, duration of hospital stay, need for rescue scheme or the use of blood products between the groups, nor was there any difference with respect to the pattern of PLT count, recovery, AST, LDH, haemoglobin or diuresis. These findings did not support the use of dexamethasone in the puerperium for recovery of women with HELLP [13].

Women with a HELLP syndrome who demonstrate progressive elevation of bilirubin or creatinine for more than 72 h after delivery may benefit from plasma exchange with fresh frozen plasma [14]. In the case of continuing haemolysis, persistent thrombocytopenia and hypoproteinaemia, post-partum erythrocyte and thrombocyte substitution, as well as albumin supplementation, are standard treatment regimens. In a recent study of women with class 1 HELLP syndrome adding of platelet transfusion to standard CS administration did not increase the recovery rate. Ertan et al. treated women with diuresis problems in the postpartum period with furosemide and applied prophylaxis with antithrombin or low-dose heparin for DIC. A meta-analysis concluded that furosemide was not beneficial to prevent or treat acute renal failure in adults [15]. Too little fluid can exacerbate an already vasoconstricted intravascular volume and lead to renal injury in severe preeclampsia or in the HELLP syndrome. A bolus intravenous fluid of 250–500 ml is advocated if oliguria persists, and, if necessary, central monitoring of the patient.

Some patients with the HELLP syndrome, especially those with DIC, may demonstrate delayed resolution or even deterioration in the post-partum period. Therefore, the use of heparin has been proposed for patients with preeclampsia, HELLP syndrome and DIC. A retrospective analysis of women with DIC in the post-partum period revealed that 6 of 9 developed post-partum bleeding including retroperitoneal haematoma. Treatment with heparin was discouraged for post-partum bleeding. Thus, most authors oppose the routine use of heparin. The increased capillary leak may be due to a combination of increased postcapillary resistance and endothelial disruption resulting in disordered protein movement and reduced intravascular oncotic pressure. The low colloid osmotic pressure results in effusions such as ascites. According to Cong the incidence of ascites was found to be 21.6/1000 in severe preeclampsia and it was concluded that ascites in severe preeclampsia indicated termination of pregnancy as it cannot be cured by medical treatment [16].

Woods et al. have reported 10% incidence of large-volume ascites with haemolysis, elevated liver enzymes and low platelets (HELLP) syndrome. Neither the obstetric text books nor the case reports available give a clear definition of massive ascites, although they describe massive ascites in subjective and semiobjective terms of 2 or more litres. Vaijyanath and associates concluded that massive ascites developing in preeclampsia is an unusual complication and massive ascites leading to maternal respiratory compromise calls for active termination of pregnancy within 24–48 h. The paucity of

reports in the literature may be due to under-reporting and also to the difficulty of recognizing this condition clinically. A high index of suspicion and careful clinical and ultrasonographic examination to detect ascites will help in elucidating the true incidence of this complication. The incorporation into clinical practice of evaluating the amount of ascites in preeclampsia might alert the obstetrician towards more intensive and more frequent maternal and foetal surveillance to avoid maternal and foetal hypoxia [17].

4. Conclusion

Women with HELLP syndrome in the presence of ascites require intensive monitoring to avoid complications during the intra- and postpartum period. The management of this case with use of high dose steroids which were started for the ascites indicates the response and their use.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflict of interest

There is no conflict of interest.

Acknowledgments

I acknowledge the cooperation of Shatby Hospital residents who participated in appointing the patient and following up. We also appreciate the commitment and compliance of the patient who reported the required data and attended for the regular follow-up.

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