Case Report

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Therapeutic Plasma Exchange in the Management of Postpartum HELLP Syndrome: A Case Report

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ABSTRACT

Haemolysis, Elevated Liver enzymes and Low Platelet count (HELLP) syndrome is a serious obstetric complication occurring during pregnancy or postpartum. It often co-exists with other Thrombotic Microangiopathies (TMAs), including atypical Haemolytic Uraemic Syndrome (aHUS). Differentiating between these conditions is crucial due to varying treatment approaches. Postpartum aHUS, a rare but potentially fatal condition, involves unchecked complement activation leading to severe renal failure, hypertension and microangiopathic haemolysis. A multidisciplinary approach and early diagnosis are vital for optimal patient outcomes. A 29-year-old woman presented on postoperative day 1 after an emergency Lower Segment Caesarean Section (LSCS) with oedema, proteinuria, hypertension, sudden visual impairment and persistent abdominal pain. Laboratory findings suggested HELLP syndrome with concurrent postpartum aHUS (decreasing platelet count, increased liver enzymes, decreased renal function and microangiopathic haemolysis). Critical care management included intravenous fluids, antihypertensives, antibiotics and analgesics. Therapeutic Plasma Exchange (TPE), initiated due to TMA and severe renal impairment, significantly improved clinical outcomes. Haematological issues were addressed with corticosteroid therapy (initially intravenous methylprednisolone, gradually transitioned to oral prednisolone). Diuretics managed fluid control and close monitoring continued. The patient was discharged with supportive treatment, controlled hypertension and stable renal function. This case highlights the importance of early detection and a tailored multidisciplinary approach in managing postpartum HELLP syndrome with aHUS. Further research is needed to improve diagnostic algorithms and treatment plans due to overlapping clinical presentations.

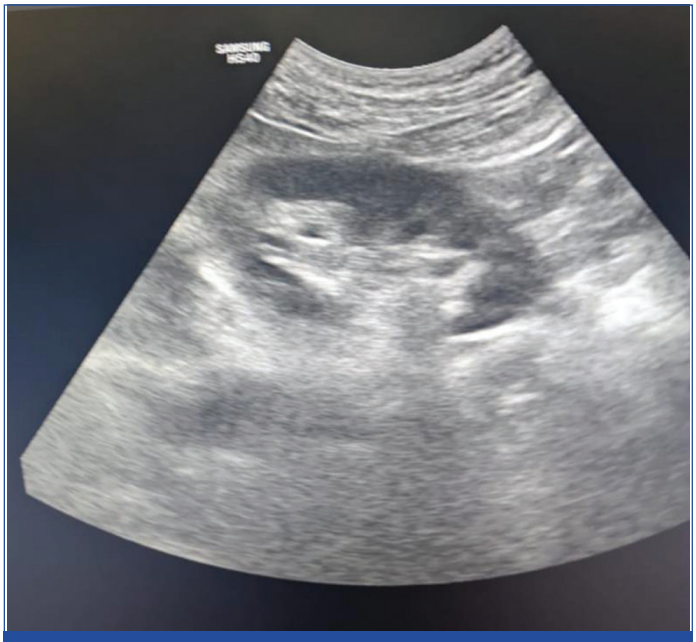
Keywords: Acute kidney injury, Haemolysis, Hypertension, Immunosuppressive agents, Microangiopathies, Thrombocytopenia

CASE REPORT

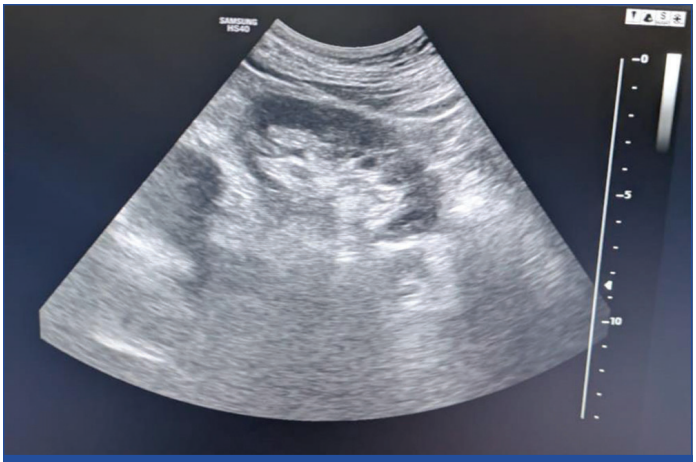
A 29-year-old primiparous woman, following an emergency LSCS for oligohydramnios, presented to a tertiary care centre on postoperative day 1. She reported sudden-onset blurred vision (lasting one hour) and persistent abdominal pain since the previous day. She had a history of High Blood Pressure (BP) on postoperative day 1; however, there was no reported headache, nausea, vomiting, or epigastric pain. Intraoperative records from the previous hospital indicated an uneventful procedure, although they revealed a Systolic Blood Pressure (SBP) of 130 mmHg and urine output <30 mL/hr, treated with intravenous Lasix. There was no history of diabetes, hypertension, substance abuse, alcoholism, smoking, or renal disease.

On admission, BP was 146/94 mmHg. The patient was conscious and well-oriented. Pallor, bilateral pedal oedema and facial and periorbital oedema (present for two months) were noted. Urine examination showed albuminuria (+3) with a urine output of approximately 100 mL. Cardiovascular examination revealed normal heart sounds (S1S2) with equal bilateral air entry. Abdominal examination showed a well-retracted uterus with a dry dressing and no visible bleeding. Fundoscopy did not reveal hypertensive retinopathy.

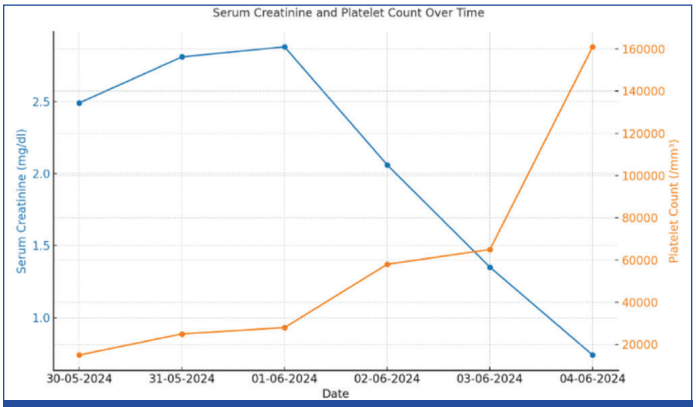
Laboratory investigations showed a platelet count decrease from 202,000/mm^3 to 67,000/mm^3 within a day and further to 15,000/mm^3, indicating worsening thrombocytopenia Normal-(>150,000/mm^3). Blood tests revealed a haemoglobin level of 8.1 g% (Normal- >11 g%) and leukocytosis (total leukocyte count 25,300/mm^3). Serum creatinine levels were rising, indicating renal impairment, while liver function tests showed elevated SGOT and SGPT. Urine analysis revealed severe proteinuria. Abdominal Ultrasonography (USG) showed bilaterally elevated renal cortical echogenicity ([Table/Fig-1] right kidney; [Table/Fig-2] left kidney). Lactate Dehydrogenase (LDH) levels and coagulation profile were assessed to evaluate HELLP syndrome. [Table/Fig-3] shows changes in serum creatinine levels and platelet count over time, illustrating the patient's recovery.



[Table/Fig-1]: Right kidney showing raised renal echogenicity finding on Ultrasonography (USG).



[Table/Fig-2]: Left kidney showing raised renal echogenicity finding on Ultrasonography (USG).



[Table/Fig-3]: Trends of serum creatinine and platelet count over time. The left y-axis represents Serum Creatinine levels (mg/dL), while the right y-axis represents platelet count (/mm³). The data points are plotted against the dates from 30-05-2024 to 04-06-2024, illustrating the patient's recovery progress.

Initially, the patient's high BP, proteinuria and fluid retention (facial, periorbital and pedal oedema) suggested postpartum hypertensive disease with potential renal involvement. Oliguria and impaired renal function further supported renal involvement. A comprehensive differential diagnosis identified the underlying condition. Postpartum preeclampsia was initially considered (oedema, proteinuria, hypertension); however, eclampsia was ruled out due to the absence of seizures. Haemolysis, elevated liver enzymes and thrombocytopenia strongly suggested HELLP syndrome, later confirmed by test results. The patient's oliguria and elevated creatinine levels were interpreted as signs of hypertensive complications leading to Acute Kidney Injury (AKI). Microangiopathic haemolysis, thrombocytopenia and renal failure raised the possibility of TMA, necessitating further investigation. Idiopathic Thrombocytopenic Purpura (ITP) was excluded (isolated thrombocytopenia). Systemic Lupus Erythematosus (SLE) was considered unlikely due to the absence of serological markers. Thrombotic Thrombocytopenic Purpura (TTP) was ruled out due to the absence of neurological symptoms and normal ADAMTS13 levels. HUS was excluded due to the absence of prior infection. Acute Fatty Liver of Pregnancy (AFLP) was considered less likely given the postpartum presentation and rapid recovery after corticosteroid therapy and supportive management.

Postpartum aHUS, characterised by severe hypertension, renal failure and microangiopathic haemolysis, complicated the HELLP syndrome (hypertension, proteinuria, thrombocytopenia, elevated liver enzymes and haemolysis). The patient received multidisciplinary care (obstetricians, nephrologists, intensivists and ophthalmologists) in critical care. Supportive treatment included intravenous fluids (haemodynamic stability), antibiotics (infection control) and analgesics (pain management). Antihypertensives controlled BP; anaemia and thrombocytopenia were treated with one unit of packed red blood cells and eight units of random donor platelets. The patient underwent three cycles of therapeutic Plasma Exchange (PLEX) due to the risk of TMA and severe renal failure (approximately 2000 mL plasma replaced with fresh frozen plasma per cycle). One pint of packed red blood cells was additionally administered during the second cycle. Strict monitoring managed associated risks. Immunosuppressive therapy began with intravenous methylprednisolone, gradually reduced to oral prednisolone. Diuretics aided oedema resolution and fluid management. Close monitoring resulted in stabilisation and improved systemic and renal function. Clinical and laboratory results improved steadily during hospitalisation. Serum creatinine decreased, urine output increased (improved renal function). Antihypertensives normalised BP, oedema subsided and neurological symptoms (blurred vision) resolved. Platelet counts increased and liver enzyme levels normalised. The patient was discharged on postoperative day 8 with supportive treatment, prednisolone and oral antihypertensives. Over four weeks, she maintained stability, with normal renal function and controlled BP. Urine protein levels decreased and fundoscopy confirmed resolution of hypertensive retinopathy. Long-term nephrology and obstetric follow-up were recommended.

DISCUSSION

HELLP syndrome is a serious pregnancy complication sometimes mistaken for preeclampsia. Weinstein's 1982 description [1] indicates it affects 0.5–0.9% of pregnancies and 10–20% of preeclampsia cases. It is associated with significant maternal and neonatal morbidity and mortality, more common in severe preeclampsia [2,3]. TPE, a category III indication, has been studied as a treatment, particularly in refractory postpartum cases [4]. TPE effectively removes toxins (coagulation factors, albumin, ammonia, endotoxins, bilirubin, inflammatory cytokines) by replacing the patient's plasma with donor plasma [5]. This improves coagulation parameters and neurological, hepatic and renal function by removing vasoactive mediators (renin, angiotensin) [6,7].

This case report highlights TPE's role in treating a postpartum patient with HELLP syndrome. The patient showed rapid clinical improvement after this intervention. The atypical presentation led to several differential diagnoses before confirmation. TPE may prevent maternal morbidity and mortality and control Disseminated Intravascular Coagulation (DIC), especially in cases refractory to conventional therapy by replacing toxic plasma components. This removes vasoactive substances (renin, angiotensin), inflammatory cytokines and other mediators contributing to the syndrome's pathophysiology [4].

Aggressive supportive management is crucial in severe postpartum HELLP syndrome, particularly with TMA and renal dysfunction. In this 29-year-old primiparous woman, a multidisciplinary approach (intravenous fluids, antihypertensives, platelet and blood transfusions, corticosteroid therapy and diuretics) led to significant clinical improvement. Studies by Vafaeimanesh J et al., and Chowdhry M et al., highlight timely intervention's importance in managing thrombocytopenia, liver dysfunction and hypertension [8,9]. The patient's recovery highlights the effectiveness of comprehensive supportive therapy in postpartum HELLP cases, as evidenced by restored renal function, stabilised BP and resolution of neurological symptoms. Patients with similar conditions (severe thrombocytopenia, microangiopathic haemolysis, multi-organ failure) have shown therapeutic benefits with PLEX.

Several studies support the timely use of PLEX in refractory HELLP syndrome to prevent irreversible organ damage and improve maternal outcomes. Ramadan MK et al., supported early plasmapheresis in cases of diagnostic uncertainty, highlighting the diagnostic challenges due to overlapping features of TTP and HELLP syndrome [10]. Similarly, Kojima N et al., documented PLEX's effectiveness in stabilising haemodynamics and reversing

organ dysfunction in patients with severe HELLP syndrome and multiorgan failure [11]. Taj S et al., showed that plasmapheresis prevented liver transplantation in a critically ill HELLP patient by improving coagulation, renal function and hepatic recovery [12].

Using TPE to differentiate HELLP syndrome from TTP and other TMAs can lead to better prognoses and fewer complications [13,14]. TPE can stabilise patients with multiorgan failure and may avoid liver transplantation in extreme cases [11,12]. PLEX has been shown to improve liver function and platelet counts, especially with early intervention [13]. It has been beneficial in cases with similar TTP and HELLP syndrome presentations, supporting its use in complex clinical settings [14].

TPE is increasingly recognised as an effective treatment for postpartum patients with HELLP syndrome, particularly in severe or refractory cases [8,9,13]. In this case, TPE led to rapid clinical improvement, demonstrating its role in managing atypical presentations. Differential diagnoses (DIC and TTP) were carefully investigated because HELLP syndrome shares similarities with other TMAs [10,14]. TPE is particularly beneficial in severe HELLP cases complicated by DIC, facilitating the removal of abnormal complement pathway components and stabilising coagulation dysfunction [10,12]. Studies show that TPE significantly reduces maternal mortality and morbidity in HELLP syndrome, with documented improvements in platelet counts and liver function [8,9,13]. Patients with Class 1 HELLP syndrome and concurrent DIC often require multiple plasmapheresis sessions (up to 22 treatments reported) [8]. Early TPE initiation, particularly in refractory postpartum cases, is associated with better clinical recovery and improved maternal outcomes [10,13]. These findings emphasise TPE's crucial role in a multidisciplinary approach in managing severe HELLP syndrome, particularly in cases with persistent multiorgan dysfunction [11,12]. This case underscores the effectiveness of early intervention in severe postpartum HELLP syndrome, leading to substantial clinical improvement, organ recovery and favourable maternal outcomes.

CONCLUSION(S)

HELLP syndrome remains a significant cause of maternal and perinatal morbidity and mortality, requiring timely and effective management. This case highlights TPE's clinical significance as a valuable intervention, particularly in postpartum cases refractory to conventional treatment. A multidisciplinary approach (TPE, intravenous fluids, antibiotics and corticosteroids) was crucial for favourable outcomes. Future research should focus on optimising treatment protocols, identifying ideal candidates for TPE and determining the optimal timing of intervention. Early recognition and diagnosis are essential to improve prognosis, emphasising the need for standardised strategies to enhance postpartum maternal and foetal health and safety.

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