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Research Article

POST-PARTUM THROMBOTIC THROMBOCYTOPENIC PURPURA WITHHEMOLYTIC UREMIC SYNDROME - A CASE STUDY

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Abstract

Introduction: Thrombotic Thrombocytopenic Purpura (TTP) is a rare and potentially life-threatening condition, particularly in the postpartum period. While TTP affects approximately 3.7 per million individuals, postpartum TTP is even less common. This disorder is characterized by microvascular thrombi formation, resulting in endothelial damage and organ dysfunction, including the kidneys, although significant acute kidney injury (AKI) is more characteristic of Hemolytic Uremic Syndrome (HUS). **Case presentation:** A 31-year-old, P1L2 woman presented postpartum with hypertension, shortness of breath, and seizures. Initial investigations revealed anemia, thrombocytopenia, and renal dysfunction. The patient underwent emergency cesarean section in outside hospital and was referred here for further management. **Clinical findings and investigations:** Initial investigations revealed anemia, thrombocytopenia, and renal dysfunction. She subsequently developed TTP, confirmed by clinical and laboratory findings, including a negative ADAMTS13. Posterior Reversible Encephalopathy Syndrome (PRES) was also diagnosed. Schistocytosis was observed on peripheral smear. **Interventions and outcome:** Her treatment consisted of plasma exchange, hemodialysis, and corticosteroids, Anti epileptics, Anti hypertensives and diuretics leading to a complete recovery. **Conclusion:** Early diagnosis and treatment with Plasma Exchange could be the vital factors in reducing maternal mortality due to postpartum HUS. This case highlights the challenges in diagnosing postpartum TTP, especially with atypical presentations and negative ADAMTS13, and the importance of prompt diagnosis and intervention.

Keywords: Thrombotic thrombocytopenic purpura, Thrombotic microangiopathy, ADAMTS13, PRES, Hemolytic Uremic Syndrome (HUS), Schistocytes, plasmapheresis.

INTRODUCTION

Thrombotic Thrombocytopenic Purpura (TTP) is a rare and potentially life-threatening condition, particularly in the postpartum period. While TTP affects approximately 3.7 per million individuals, postpartum TTP is even less common. This disorder is characterized by microvascular thrombi formation, resulting in endothelial damage and organ dysfunction, including the kidneys, although significant acute kidney injury (AKI) is more characteristic of Hemolytic Uremic Syndrome (HUS). Timely diagnosis and treatment are essential, as untreated TTP can have a high mortality rate. This report details the case of a 31-year-old woman who developed severe AKI secondary to TTP after delivery, requiring dialysis and plasmapheresis before achieving a full recovery.

CASE PRESENTATION

A 31-year-old woman, gravida 2 para 1, conceived via in vitro fertilization (IVF), with no significant past medical history, was referred to our emergency department due to elevated blood pressure and dyspnea. She had undergone an emergency lower segment cesarean section (LSCS) at an outside hospital on July 24, 2024. Following spinal anesthesia, she experienced generalized tonic-clonic seizures, necessitating intubation and conversion to general anesthesia for the LSCS. Postoperatively, she exhibited respiratory distress.

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Laboratory tests at the referring hospital demonstrated elevated serum lactate dehydrogenase (LDH), abnormal liver and renal function, and a declining platelet count. She received transfusions of fresh frozen plasma (FFP) and packed red blood cells (PRBCs). Due to persistently abnormal LDH, liver, and renal function, along with worsening thrombocytopenia, she was transferred to our institution. Upon arrival, the patient was drowsy (GCS: E3V4M5), with a pulse of 94 beats per minute, a respiratory rate of 18 breaths per minute, and an SpO2 of 98% on 3 liters of oxygen. Her blood pressure was 160/100 mmHg, and labetalol infusion was initiated. Arterial blood gas analysis revealed metabolic acidosis with hyperkalemia. She was admitted to the intensive care unit (ICU) for further management. Initial blood work showed anemia (Hb 10.5 g/dL), leukocytosis (TLC 12,200 cells/mm3), severe thrombocytopenia (23,000 platelets/µL), and impaired renal function (creatinine 2.4 mg/dL). Liver function tests showed hyperbilirubinemia (total bilirubin 4.5 mg/dL), hypoalbuminemia, and elevated transaminases (SGPT 1495 U/L, SGOT 2275 U/L). Electrolytes showed hyperkalemia (K 5.9 mmol/L). Urine analysis revealed pyuria and 3+ proteinuria. A chest X-ray showed right upper zone infiltrates. Serum LDH was markedly elevated (4660 U/L). A peripheral blood smear demonstrated schistocytes. Due to the suspicion of thrombotic microangiopathy (TMA), a nephrology consultation was obtained, and plasmapheresis was planned. An echocardiogram revealed normal cardiac function (ejection fraction 64%). An abdominal ultrasound identified a collection, possibly a hematoma, in the right paramedian abdominal wall at the surgical site. Empiric antibiotics were

started after obtaining blood and urine cultures. The family was informed about the patient's condition and the need for plasma exchange. The patient underwent six sessions of plasma exchange, one session of double filtration plasmapheresis (DFPP), and one session of hemodialysis. Importantly, the ADAMTS13 activity was negative. Low-dose corticosteroids were administered. The patient's mental status deteriorated, and a CT brain scan revealed changes consistent with Posterior Reversible Encephalopathy Syndrome (PRES). A neurologist opinion was obtained and AED was added. Blood pressure was meticulously controlled with anti hypertensives. Serial monitoring of LDH, platelet count, and renal function demonstrated improvement. The patient was transferred to the ward. On August 2nd, the patient developed severe abdominal pain. Serum amylase (307 U/L) and lipase (490 U/L) levels were elevated. A CT abdomen was performed, which was reported as non-contributory. A gastroenterology consultation was obtained, and conservative management was continued. The patient experienced shortness of breath during transfer for the CT scan and was found to be hypoxic. She was returned to the ICU and placed on BiPAP with oxygen support. Diuretics were administered. A repeat echocardiogram showed moderate pulmonary artery hypertension (PAH). A cardiology consult was obtained, and diuretics and antihypertensives were continued. The patient's condition improved, oxygen support was weaned, and she was transferred back to the ward. Her general condition continued to improve, and she was subsequently discharged.

DISCUSSION

Postpartum TTP is a rare but critical condition. This case presented diagnostic challenges due to the patient's atypical presentation and the negative ADAMTS13 result. While the classic pentad of TTP includes microangiopathic hemolytic anemia, thrombocytopenia, neurological abnormalities, renal dysfunction, and fever, this patient's presentation was dominated by seizures, hypertension, and respiratory distress. The negative ADAMTS13 result is noteworthy, as it suggests that this case may represent a form of TTP distinct from the more common ADAMTS13-deficient type. Atypical HUS, which can also present with TMA, was considered, but the clinical picture and the absence of diarrheal prodrome made it less likely. Drug-induced TMA was also a consideration, but no offending agents were identified. The use of corticosteroids, despite the negative ADAMTS13, was based on clinical judgment, considering the severity of the patient's condition and the possibility of other contributing factors to the TMA. This highlights the complexity of managing TTP, especially in cases with atypical presentations. The differential diagnosis also included preeclampsia and HELLP syndrome, which share some clinical features with TTP, such as hypertension, thrombocytopenia, and elevated liver enzymes. However, the presence of schistocytes, the severity of the thrombocytopenia, and the neurological symptoms pointed toward TTP. The PRES findings are consistent with the patient's seizures and elevated blood pressure. PRES is a recognized complication of TTP. The abdominal pain and elevated amylase/lipase raised concern for pancreatitis. While the CT abdomen was reported as non-contributory, patient was managed conservatively. The moderate PAH observed on repeat echocardiogram could be a consequence of the TTP, although it is also possible that it represents a separate, unrelated finding. Cardiologist opinion was sought and medicines were added accordingly.

Conclusion

This case underscores the importance of considering TTP in the differential diagnosis of postpartum complications, even when the presentation is atypical and the ADAMTS13 activity is negative. Prompt recognition and aggressive management with plasma exchange, hemodialysis, and corticosteroids are crucial for improving outcomes. Further research is needed to better understand the pathophysiology of TTP, particularly in cases with negative ADAMTS13, and to develop more targeted treatment strategies. Clinicians should be aware of the challenges in diagnosing TTP and maintain a high index of suspicion, especially in postpartum patients presenting with neurological symptoms, thrombocytopenia, and evidence of microangiopathic hemolysis.

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