Postpartum Thrombotic Thrombocytopenic Purpura Presenting with Status Epilepticus: A Case-Based Narrative Review

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ABSTRACT

Thrombotic thrombocytopenic purpura (TTP) is a rare but life-threatening thrombotic microangiopathy caused by severe deficiency of ADAMTS-13, a metalloproteinase responsible for cleaving ultra-large von Willebrand factor multimers. Pregnancy and the postpartum period represent well-recognized triggers for TTP, with incidence substantially higher than in the general population. Delayed recognition carries a high mortality risk. We present a case of an 18-year-old postpartum woman who developed status epilepticus as the initial manifestation of TTP, alongside a narrative review of the current evidence regarding pathophysiology, differential diagnosis, risk stratification with the PLASMIC score, and therapeutic management. The patient was successfully treated with plasma exchange, corticosteroids, and antiplatelet therapy. This case underscores the importance of including TTP in the differential diagnosis of any postpartum patient presenting with neurological deterioration and thrombocytopenia.

Keywords: thrombotic thrombocytopenic purpura; postpartum; ADAMTS-13; PLASMIC score; plasma exchange; status epilepticus

INTRODUCTION

Thrombotic thrombocytopenic purpura (TTP) is a thrombotic microangiopathy (TMA) characterized by microangiopathic hemolytic anemia, severe thrombocytopenia, and end-organ ischemia secondary to platelet-rich microthrombi in the microcirculation. The incidence in the general population is estimated at 2–6 cases per million annually, making it an uncommon but diagnostically urgent condition [1]. Untreated, mortality approaches 90%; with appropriate plasma exchange therapy, survival rates have improved dramatically to over 80–90% [2].

Pregnancy and the postpartum period are independently associated with an increased risk of TTP, with the incidence in pregnant women estimated to be up to 10-fold higher than in the

general population [3]. This increased risk is thought to reflect the physiological changes in coagulation and von Willebrand factor (vWF) levels that occur during pregnancy. Importantly, the clinical presentation of TTP frequently overlaps with other obstetric emergencies—including preeclampsia, HELLP syndrome, acute fatty liver of pregnancy, and antiphospholipid syndrome—complicating timely diagnosis [4].

Neurological manifestations, including headache, confusion, seizures, and stroke, occur in up to 60% of TTP cases and may represent the dominant presenting feature. Status epilepticus as an initial presentation is uncommon but has been reported [5]. This case report and narrative review aims to highlight the diagnostic challenges of postpartum TTP, outline the utility of risk stratification tools, and summarize current evidence on treatment strategies.

PATHOPHYSIOLOGY

The central mechanism of TTP involves a severe deficiency—either congenital or acquired—of ADAMTS-13 (a disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13), the metalloproteinase responsible for cleaving ultra-large von Willebrand factor (UL-vWF) multimers. When ADAMTS-13 activity falls below approximately 10% of normal, UL-vWF multimers persist in the circulation and promote spontaneous platelet aggregation, leading to microvascular thrombosis [1,2].

Acquired TTP, which accounts for the majority of adult cases, results from autoantibodies (predominantly IgG) directed against ADAMTS-13. Pregnancy-associated TTP typically involves a combination of physiologically elevated vWF levels alongside pre-existing partial ADAMTS-13 deficiency unmasked by the additional antigenic burden of pregnancy. The postpartum period may precipitate TTP through immune dysregulation, inflammation, and the acute haemostatic changes accompanying delivery [3].

The pathological consequence of microthrombus formation is multiorgan ischemia, preferentially affecting organs with high microvascular density—including the brain, kidneys, heart, and gastrointestinal tract. Neurological involvement is particularly common due to the sensitivity of cerebral microvasculature to platelet-rich occlusion [5].

CASE REPORT

An 18-year-old female patient with no prior medical history was initially evaluated at an external center with complaints of fatigue and was followed with a provisional diagnosis of idiopathic thrombocytopenic purpura secondary to pregnancy. Due to insufficient blood

product support, she was transferred to our institution. On admission, laboratory investigations revealed thrombocytopenia (platelet count 12,000/μL), hemoglobin 7.1 g/dL, LDL cholesterol 542 mg/dL, creatinine 0.93 mg/dL, haptoglobin <0.1 g/L, and ADAMTS-13 activity of 0.02 U/mL, consistent with severe deficiency. The PLASMIC score was calculated at 6, indicating high probability of TTP.

During hospitalization, the patient developed status epilepticus, necessitating orotracheal intubation and transfer to the intensive care unit. Following hepatitis B serological screening, therapeutic plasma exchange was initiated with fresh frozen plasma. Over the course of five plasma exchange sessions, combined with prednisolone 1 mg/kg/day and aspirin 100 mg/day (initiated after platelet count exceeded 150,000/μL), the patient achieved haematological remission. A weaning protocol was successfully completed, and the patient was transferred to the ward and subsequently discharged on oral prednisolone 32 mg/day.

DIFFERENTIAL DIAGNOSIS IN THE OBSTETRIC SETTING

The differential diagnosis of TMA in the peripartum period is broad and clinically challenging. The principal conditions to consider include preeclampsia and HELLP syndrome, hemolytic uremic syndrome (HUS), acute fatty liver of pregnancy (AFLP), antiphospholipid syndrome (APS), and disseminated intravascular coagulation (DIC) [4]. Each of these may present with overlapping features of thrombocytopenia, microangiopathic hemolysis, and end-organ dysfunction.

Several clinical and laboratory features can aid differentiation. HELLP syndrome typically resolves within 48–72 hours postpartum and is associated with elevated liver enzymes and hypertension. HUS predominantly involves renal failure and is more commonly associated with Shiga toxin-producing organisms or complement dysregulation. AFLP is characterized by elevated transaminases, hypoglycemia, and hypofibrinogenemia. In contrast, TTP is uniquely associated with severely reduced ADAMTS-13 activity (<10%), which, when measurable, provides the most specific diagnostic confirmation [1,4].

The PLASMIC score, which incorporates seven readily available clinical and laboratory variables—platelet count, hemolysis markers, absence of active cancer, absence of stem cell transplant, MCV, INR, and creatinine—provides a validated bedside tool for risk stratification prior to ADAMTS-13 results [6]. A score of 6–7 confers a high probability of severe ADAMTS-13 deficiency and should prompt immediate initiation of plasma exchange without awaiting confirmatory results.

TREATMENT

Plasma exchange (PEX) with fresh frozen plasma (FFP) remains the cornerstone of TTP management. PEX acts by removing circulating anti-ADAMTS-13 autoantibodies and UL-vWF multimers while replenishing functional ADAMTS-13. Daily PEX should be initiated as soon as the diagnosis is suspected, prior to confirmation of ADAMTS-13 results, given the mortality risk of delay [2].

Corticosteroids, typically prednisolone at 1 mg/kg/day, are used adjunctively to suppress autoantibody production. Antiplatelet therapy with aspirin is generally initiated once platelet counts recover above 50,000–150,000/μL. Rituximab, an anti-CD20 monoclonal antibody, is increasingly incorporated in refractory or relapsing TTP and in cases with persistently low ADAMTS-13 activity after PEX cessation [7].

Prior to initiating rituximab, hepatitis B serology (HBsAg and anti-HBc total/IgG) should be obtained in all patients. Antiviral prophylaxis should be administered for at least one year following treatment completion in patients with evidence of past hepatitis B infection [7]. Caplacizumab, an anti-vWF nanobody, is a newer agent that reduces platelet consumption and may shorten time to platelet count normalization; its availability in Turkey and other low-to-middle income settings remains limited.

DISCUSSION

This case illustrates several important teaching points in the recognition and management of postpartum TTP. First, the initial misclassification as gestational ITP highlights the diagnostic difficulty in thrombocytopenic postpartum patients. ITP does not cause hemolysis, and the presence of microangiopathic changes on the peripheral blood smear (schistocytes), together with elevated LDH and low haptoglobin, should prompt urgent consideration of TMA.

Second, the presenting feature of status epilepticus is an uncommon but recognized manifestation of TTP-related cerebral microvascular occlusion. Clinicians managing postpartum seizures must include TTP in their differential, particularly in the context of concurrent thrombocytopenia and hemolytic anemia, even in the absence of the full classical pentad (which is now recognized to be present in fewer than 5% of cases at diagnosis).

Third, this case demonstrates the utility of the PLASMIC score in rapidly identifying high-risk patients before ADAMTS-13 results are available. A score of 6 or above should be treated with

the same urgency as a confirmed diagnosis. Early initiation of PEX is the single most important determinant of outcome in TTP.

Finally, the successful clinical course in this patient underscores the efficacy of a systematic, guideline-concordant approach combining PEX, corticosteroids, and antiplatelet therapy. Regular monitoring of platelet counts, ADAMTS-13 activity, and lactate dehydrogenase levels guides treatment duration and helps identify early relapse.

CONCLUSION

Postpartum TTP is a rare but potentially fatal condition that requires a high index of clinical suspicion, particularly when thrombocytopenia is accompanied by hemolytic anemia and neurological manifestations. Status epilepticus may be the presenting feature. The PLASMIC score provides reliable risk stratification when ADAMTS-13 results are unavailable. Prompt initiation of plasma exchange, combined with corticosteroids and antiplatelet therapy, is the mainstay of treatment and significantly improves survival. Increased awareness of TTP among obstetric and critical care teams is essential for timely diagnosis and intervention.

AUTHOR CONTRIBUTIONS

Concept and design: A.T.K. Data collection: A.T.K. Data analysis and interpretation: A.T.K. Manuscript writing: A.T.K. Critical revision: A.T.K. Final approval: A.T.K.

CONFLICT OF INTEREST

The author declares no conflict of interest.

ETHICS STATEMENT

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

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