



Pregnancy-associated Complement-mediated Thrombotic Microangiopathy: diagnostic dilemma

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INTRODUCTION

- Complement-mediated Thrombotic Microangiopathy (c-TMA) is also known as atypical hemolytic uremic syndrome
- Caused by uncontrolled activation of the complement system
- Elevated terminal complement C5b-9 activates platelet and vascular endothelial-cells causing fulminant, systemic microvascular clotting
- Characterized by hemolytic anemia, thrombocytopenia, and renal failure

CASE

- 34 y.o. G3P1 with prior NSVD. Admission CBC was normal and routine labor epidural placed. Hours later fetal bradycardia required emergent cesarean delivery
- Patient developed postpartum hemorrhage and DIC requiring massive transfusion in OR (6 pRBC's, 5 FFP, 2 cryo)
- Post-op patient had persistent thrombocytopenia (nadir 23k) and oliguric AKI (peak Cr 5.6) requiring continuous dialysis

CASE CONT.

- Labs revealed low C3/C4, hemolysis and rhabdomyolysis. ADAMTS13 activity was normal
- A diagnosis of pregnancy-associated c-TMA was made and the terminal complement inhibitor eculizumab (monoclonal Ab to C5) started POD2. The clinical picture quickly improved
- She was discharged on POD14, continued eculizumab for 6 months and received a brief period of outpatient dialysis
- Genetic workup confirmed a missense variant in complement factor I, consistent with c-TMA

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DISCUSSION

- C-TMA commonly presents in the postpartum period, often after a pregnancy complication, such as PPH
- Patients are often critically ill and diagnosis may be delayed due to overlapping features with other TMA syndromes (TTP, HELLP)
- Multidisciplinary collaboration with hematology, nephrology, MFM and anesthesiology was paramount in early recognition and treatment in this case

REFERENCES

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Disease	Classic Features	Treatment
C-TMA	Renal failure, hemolytic anemia, thrombocytopenia	Eculizumab
HELLP	Hemolysis, elevated LFTs, thrombocytopenia	Delivery
TTP	Severe, prolonged thrombocytopenia Reduced ADAMTS13	Plasma exchange