Postpartum Complement Mediated Thrombotic Microangiopathy Caused by Subacute Bacterial Endocarditis

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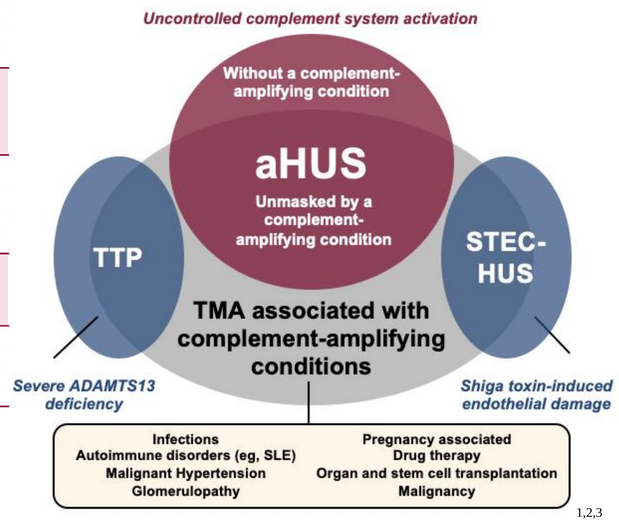
Background

Thrombotic microangiopathies (TMA) are a heterogenous group of syndromes characterized by: microangiopathic hemolytic anemia, thrombocytopenia, and end organ damage.
TMA are classified as primary or acquired, and can manifest subsequent to infectious events, and resolve with underlying disease treatment, however they have high morbidity and mortality.
Here, we report a case of complement-mediated TMA (C-TMA) secondary to Group B strep tricuspid valve bacterial endocarditis (SBE), successfully treated with apheresis, empiric antibiotics, and percutaneous vegetation debulking.

Primary cause and target of coagulopathy		Platelet Count	D-Dimer	PT / aPTT	Fibrinogen
DIC	Macrophage, enodthelial cell	↓	↑	↑	↓
APS	Antiphospholipid antibody	↓	→	PT → PTT ↑	→
aHUS	Complement system	↓	→	→	→
TTP	ADAMTS 13	↓	→	→	→

Table 1. Biomarker changes in various TMAs. *DIC* disseminated intravascular coagulation, *APS* antiphospholipid syndrome, *aHUS* atypical hemolytic uremic syndrome, *TTP* thrombotic thrombocytopenic purpura, *PT* prothrombin time, *aPTT* activated partial thromboplastin time [7]



[1,2,3]

Case Presentation

A 31-year-old healthy female presented four weeks post cesarean delivery with weakness and shortness of breath, after being found hypotensive and tachycardic at a routine visit.

Vitals: Temp 37.0 C, HR 148 /min, RR 22 /min, BP: 80/50, O2 Sat: 97% on RA
Physical exam was significant for mottled bilateral lower extremities, consistent with livedo reticularis
Diagnostic tests:

Hgb / Hct	7.9 g/dL / 26.4 %	↓ / ↓
INR	1.3	↑
Platelet count	17 K/μL	↓↓
Creatinine	2.5 mg/dL	↑

All normal during delivery 1 month ago

LDH	775 U/L	↑
Total bilirubin	1.3 mg/dL	—
Fibrinogen	199 mg/dL	—
Haptoglobin	135 mg/dL	—

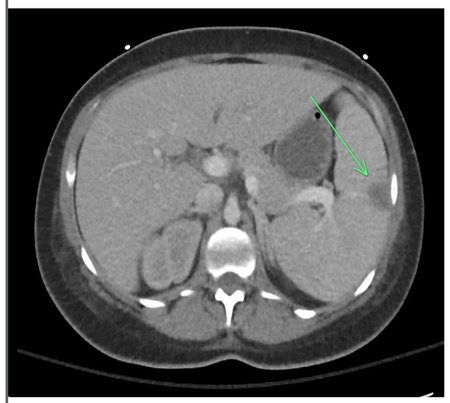
Peripheral smear few shistocytes, evidence of hemolysis

ADAMTS 13 Acitivty	24%	↓, not consistent with TTP (< 10%) [3]
C3	50	↓
C4	70	↓
Total complement (CH50)	11	↓

Low complement levels indicative of complement system activation

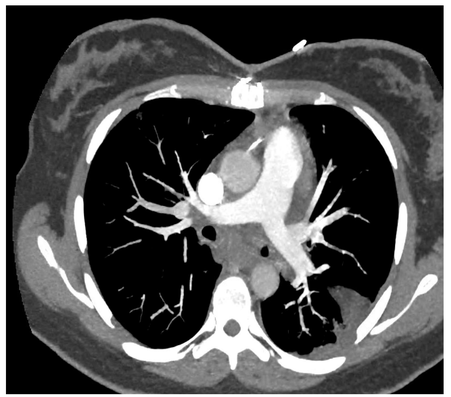
Imaging

CT Abdomen and Pelvis



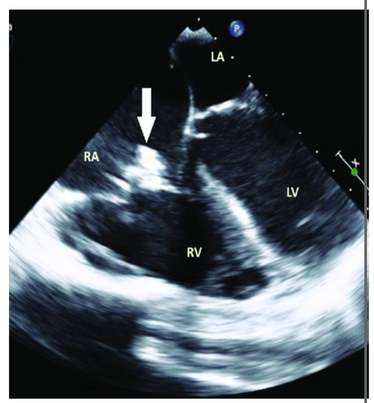
Multiple wedge-shaped peripheral splenic infarcts

CTA Chest



Distal right main PA, bilateral upper and lower lobe segmental arterial emboli with RV strain, and bilateral lower lobe and LUL pulmonary infarcts

TEE



Tricuspid valve vegetation

Hospital Course

On the day of admission, tightly regulated anticoagulation and plasmapheresis were started with suspected diagnosis of TTP and atypical HUS.
After forty-eight hours of plasma exchange, she showed rapid improvement.
Workup for autoimmune hemolytic anemia, systemic lupus erythematosus, catastrophic antiphospholipid syndrome, disseminated intravascular coagulation, typical HUS, heparin-induced thrombocytopenia, drug-induced TMA, and malignancy returned negative.
Blood cultures were positive for Group B streptococcus (prior to delivery GBS testing negative) and TEE showed a large mobile tricuspid vegetation without PFO; antibiotics were initiated. Once stabilized, the patient was referred for percutaneous vegetation debulking and encouraged to pursue genetic testing for complement protein mutations.

Conclusion

This is an unusual presentation of C-TMA provoked by SBE. Had this patient had hereditary C-TMA, her course would have been refractory to plasma exchange and eculizumab would have been initiated.
GBS endocarditis of the tricuspid valve, particularly in patients without a history of IVDA or other predisposing risk factors is a rare entity. Only 24 different cases have been reported thus far in the literature, of which 9 are pregnancy-related and only 2 post c-section cases [4,5]
Early recognition and rapid communication with an interdisciplinary multispecialty team resulted in immediate intervention with plasmapheresis as rescue therapy until the provoking endocarditis was treated.

References

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