Second-Trimester Abortion Complicated by Atypical Hemolytic Uremic Syndrome (aHUS)

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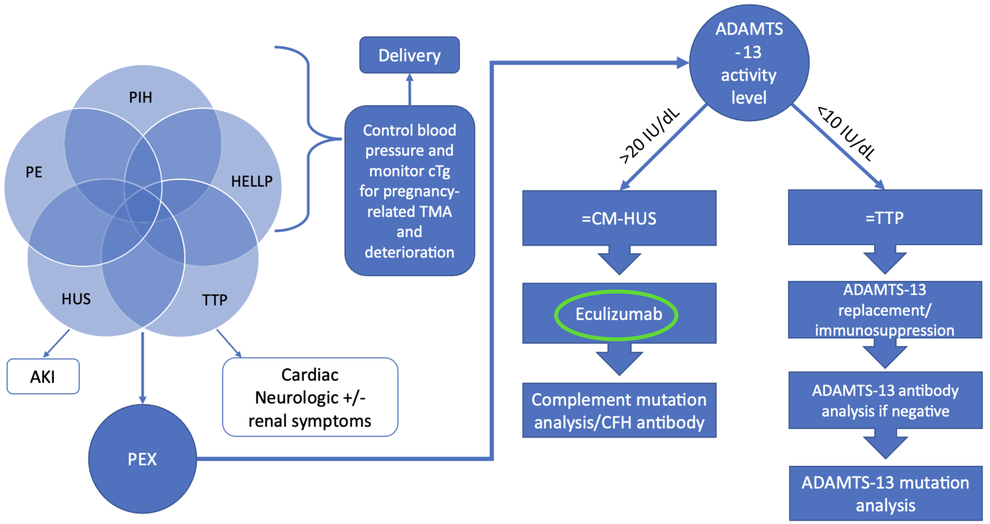
- **Pregnancy-associated thrombotic microangiopathies (TMA)**
  - Microangiopathic hemolytic anemia (Hgb < 10 g/dL, LDH > 1.5 upper limit of normal)
  - Thrombocytopenia (<100 x 10^9 /L)
  - End-organ damage
  - High morbidity & mortality in **aHUS**: 4x risk of ESRD and 6x risk of mortality over 10-year period

- **Subtypes:**

Atypical hemolytic Uremic Syndrome (aHUS)	Thrombotic thrombocytopenia purpura (TTP)	PET/ HELLP
• 1 in 25 000 • Severe AKI • 2/3 need dialysis • Dysregulation in complement alt pathway	• 1 in 200 000 • Mainly neuro +/- cardiac • ADAMTS deficiency	• PET 1 in 20 HELLP 1:1000 • Multi-systemic • Less severe AKI • Imbalance b/t angiogenic & anti-angiogenic factors

- **Treatment:**



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PE, PET = pre-eclampsia, PEX = plasma exchange, cTg = cardiotocography, CM-HUS = complement-mediated HUS

- **Case**
  - 24-year-old woman presented for 24-week dilation and evacuation for undesired pregnancy

- **Intraop course**
  - Anesthesia: Deep sedation with paracervical block
  - Complicated by post-abortion hemorrhage (EBL 1.7 L) and DIC (Hgb 6.4 g/dL, Plt 89 K/μL, INR 1.7, Fib 65 mg/dL)
  - Management
    - Intrauterine balloon
    - Transfusion: 4 U pRBC, 6 U FFP, 1 U platelet, 5 g fibrinogen concentrate
    - Bilateral uterine artery embolization

- **ICU postop course**
  - Rapidly progressive AKI
    - Serum creatinine **0.73 mg/dL (pre-op) → 7.62 mg/dL (peak POD4)**
  - Hematology/nephrology consultations
  - DDx: ATN, contrast nephropathy, TMA
  - TMA workup
    - HELLP, TTP, APLS ruled out
    - aHUS
      - Equivocal complement genetic panel
      - **Elevated soluble complement 5b-9 (332 ng/mL)**

- **Outcome**
  - Transferred to tertiary care center to initiate eculizumab (complement C5 inhibitor)
  - Self-directed discharge before treatment started
  - Re-presented on POD11 for SOB
    - Diagnosed with new heart failure with mildly reduced ejection fraction (EF 40-45%). DDx peripartum- versus TMA/aHUS-associated cardiomyopathy → started on guideline-directed medical therapy
    - **Serum creatinine 1.99 g/dL → deferred eculizumab after risk and benefits discussion**

- **Teaching Points**
  - aHUS
    - Diagnostic challenge due to clinical overlap with other pregnancy-associated TMAs
    - Medical emergency with high risk of rapid progression to ESRD
    - Early involvement of multidisciplinary specialists
    - Prompt treatment with eculizumab improves renal outcomes
  - Eculizumab
    - Complement (C5) inhibitor
    - Appears safe in pregnancy (limited data)
    - Plasma exchange if complement inhibitor therapy unavailable
  - Optimal management of subsequent pregnancies remains unknown
    - Increased risk of recurrence of aHUS or PET
    - C5 inhibitor prophylaxis may be warranted in high-risk cases

References:

1. PMID: 22270271
2. PMID: 39156777
3. PMID: 41477157
4. PMID: 28101432