When Pregnancy's Pro-Thrombotic State Isn't Enough: Refractory Acquired Hemophilia A

Background

Pregnancy-Associated Acquired Hemophilia A (AHA)

- Accounts for 1-8% of all AHA cases.
- Rare acquired bleeding disorder caused by IgG autoantibodies to Factor VIII (FVIII). Typically presents postpartum; however, can rarely arise during pregnancy (< 5% of cases).
- Patients typically present with soft tissue, cutaneous or mucosal bleeding. Postpartum presentation commonly presents with obstetrical/genital bleeding.
- Diagnostics include:
  - Prolong aPTT, mixing study, reduced FVII activity, and positive FVII inhibitor assay
- Management (two goals: hemostatic management and inhibitor eradication)
  - Hemostatic management: recombinant activated Factor VII and activated prothrombin complex concentrate
  - Inhibitor eradication: Corticosteroids/cyclophosphamide. Rituximab for refractory cases.
- Special Considerations:
  - Neuraxial anesthesia requires greater than 50% FVIII levels
  - The recurrence risk is approximately 22% in subsequent pregnancies

The Case

23-year-old G3P2 at 17 weeks gestation

Initial Presentation:

- Dyspnea on exertion, anemia (Hgb 6.8), prolonged PTT
- Workup: FVIII 1%, high FVIII antibodies (200+ BUs)
- Treatment: IVIG/steroids → minimal improvement. Discharged with oral TXA and steroid taper
- Due to social situation, patient presented only once for follow-up where factor activity remained <1%.

Delivery (36 weeks - IOL for cholestasis):

- FVIII 1% with high titers → neuraxial anesthesia contraindicated
- Uncomplicated SVD with recombinant FVIIa during second stage + 24 hours postpartum
- Left AMA on PPD2 with "normal" bleeding

Postpartum Complications:

- PPD 7: Severe anemia (Hgb 4) → D&C with JADA placement
- 2 weeks later: Recurrent bleeding + endometritis → repeat D with JADA
- Persistently high titers and near-absent FVIII activity throughout

Teaching Points

- **Maintain high index of suspicion:** Consider AHA in pregnant patients with prolonged PTT and bleeding events
- **Factor level thresholds matter:**
  - >50% recommended for neuraxial anesthesia
  - 1% associated with severe bleeding complications
- **Refractory disease requires aggressive management:**
  - Bypass hemostatic therapy (recombinant FVIIa or prothrombin complex) for delivery
  - Rituximab used in refractory cases. Remission rates of 60-90%.
  - Emicizumab is an emerging option of hemostatic management; however, use is currently off label
  - Consider prophylactic uterine artery embolization in severe cases
  - Extended postpartum monitoring essential
- **Multidisciplinary coordination is critical:** Tertiary center management with hematology, OB, anesthesia, and blood bank involvement
- **Postpartum vigilance:** Low threshold for mechanical uterine decompression given limited hemorrhage prevention options