

A Parturient with Glanzmann's Thrombasthenia (GT): A Case Report

Sana Rukh MD MPH¹, Maithili Khandekar BS¹, Truc-Anh Nguyen MD²

1. University of Arizona College of Medicine- Tucson Department of Anesthesiology

2: Banner University Medical Center-Tucson

Background

- GT is an autosomal recessive bleeding disorder with a qualitative or quantitative defect in integrin $\alpha\text{IIb}\beta 3$ which leads to a platelet aggregation defect
- Incidence is estimated at 1/1,000,000 worldwide but more common in South Asian, Middle-Eastern, Jewish, French Romanian populations
- Type 1 (<5% of normal protein) is the most common type at ~78%

Patient Presentation

- 34F G1P0 w/ GT diagnosed in childhood with strong family hx (father, brother, sister & carrier mother) and mucocutaneous bleeding with dental manipulation managed with Amicar, OCPs, and Fe supplements
- Physical exam notable for mild bruising otherwise hemodynamically stable
- Lab values: H/H (11.2/33%), MCV 103, platelets 175, bleeding time 8 mins

Clinical Course

- 2nd trimester**
 - Admitted for increased WOB, back pain, and fever and found to have R pleural effusion & RML lung mass on CTA chest (Figure 2) presumed to be a hemothorax after a negative infectious/neoplastic workup
 - Underwent US-guided thoracentesis & chest tube placement with 800cc bloody fluid drained, started on F7 replacement & Amicar, d/c'd on 2L oxygen
- 3rd trimester**
 - Case discussed weekly during Multi-disciplinary Rounds
 - Admitted for mIOL x2, which she ultimately failed & underwent C/S with the decision to undergo GETA instead of neuraxial anesthesia (pre-op Hgb 11.2 & plts 175), bleeding time 8 mins
 - Pre-op: transfused 2u platelets + 1g TXA
 - Intra-op: transfused 1u platelets + 1g TXA
 - Post-op: Observed 5 days w/o hemorrhage or F7 replacement then discharged
 - Post-partum: Placed on OCPs without any major bleeding

Figure 1: GT Pathophysiology

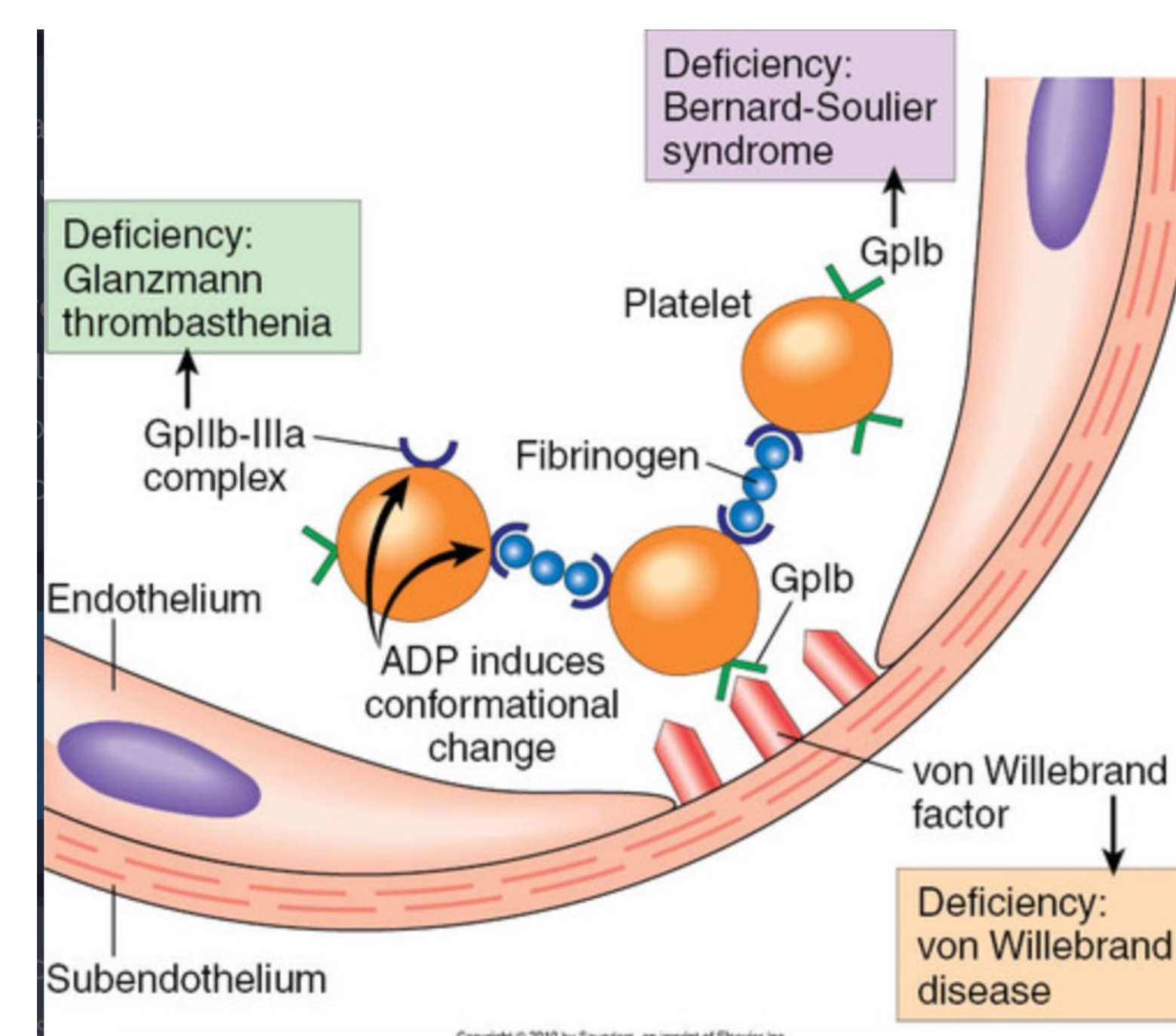


Figure 2: CTA Chest of Patient



References

- Wijemanne, A., Watt-Coote, I., & Austin, S. (2016). Glanzmann thrombasthenia in pregnancy: Optimising maternal and fetal outcomes. *Obstetric medicine*, 9(4), 169–170.
- Peterson, W., Tse, B., Martin, R., Fralick, M., & Sholzberg, M. (2021). Evaluating hemostatic thresholds for neuraxial anesthesia in adults with hemorrhagic disorders and tendencies: A scoping review. *Research and practice in thrombosis and haemostasis*, 5(4), e12491.
- Figure 1: Copyright 2010 by Saunders, an imprint of Elsevier Inc.

Discussion

Signs & Symptoms of GT

- Mucocutaneous bleeding- epistaxis, gingival bleeding, petechiae, purpura, GI/GU bleeding
- Excessive bleeding after trauma and/or invasive procedure

Diagnosis

- Prolonged bleeding time, normal plt count/morphology, aPTT, PT
- Family history & physical showing typical signs of GT

Concerns During Pregnancy

- Greater risk of miscarriage and antepartum & postpartum hemorrhages
- If mother develops anti-platelet antibodies, can transfer over placenta → severe neonatal thrombocytopenia

Treatment

- Avoiding antiplatelet meds, punctures, invasive procedures
- Medications
 - Childbearing age females
 - OCPs to suppress menses, Fe supplements, hysterectomy
 - Prophylactic plts before invasive procedures
 - Childbirth: Ppx w/ F7 replacement & antifibrinolytics
- Managing bleeding episodes symptomatically (ex: nasal packing, TXA)
- Hematopoietic stem cell transfusion if severe

Conclusion

- Importance of involving anesthesiologists early to maximize safety (ex: GETA over neuraxial anesthesia d/t risk of hematomas & permanent neurological damage)
- Multidisciplinary approach required during pre-operative, peri-operative, and post-operative care of GT patients