Neuraxial Anesthesia in Von Willebrand Disease Type 2a

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Background

- Von Willebrand disease (VWD) is the most common inherited bleeding disorder in the United States, affecting up to 1% of the US population.

- Von Willebrand Disease Type 2A is a qualitative defect of vWF, leading to moderate to severe bleeding in affected individuals. Of the various subtypes of Von Willebrand Disease, type 2A accounts for about 10-20% of cases.
  - Qualitative defect is due to a loss-of-function mutation in VWF, leading to poor binding of VWF to Platelet GP1b receptors.
  - This particular type of VWD is inherited in an autosomal dominant pattern.
  - VWD type 2a is identified on laboratory analysis by a significant decrease in VWF activity when compared to VWF antigen (decreased VWF activity:antigen ratio), as well as decreased ristocetin-induced agglutination (VWF:RCO)

- The safe administration of neuraxial anesthesia in such patients poses significant challenges due to the risk of epidural hematoma and potential catastrophic neurologic sequelae, further complicated by the fact that there is sparse evidence of safe neuraxial administration in these patients[2].

- Identifying safe methods to administer neuraxial anesthesia in patients with inherited coagulopathies allows not only for the ability to facilitate surgical procedures without the need for general anesthesia and airway manipulation in the high-risk obstetric population, but it also allows for safe autonomous decision making and potential improvements in the birthing experience for the mother.

The Case

- 40 year old obstetric patient with a significant history of Von Willebrand Disease type 2A, diagnosed in childhood. Patient presented to pre-operative clinic and indicated a preference for neuraxial anesthesia for her repeat scheduled cesarean delivery.

- Preoperative labs obtained by hematology at 36 weeks gestation demonstrated vWF activity <10%, vWF antigen 109%, and factor VIII activity at 77%, indicating a profound qualitative defect of von Willebrand factor.

- To help decrease the risk of epidural hematoma post neuraxial anesthesia, patient was given a bolus of plasma-derived von Willebrand factor/factor VIII prior to the procedure, with a body-weight dosed infusion started perioperatively and continued post-operatively for 7 days.
  - Bolus dose calculation per hematology note: BW in KG x [125% VWF:RCO x 0.5]
  - Infusion was calculated as: [Initial Bolus / 24 hours]
  - VWF panel was checked 4 hours after loading and at every 12 hours until delivery.
  - VWF panel was also checked 4 hours and 12 hours post delivery, with repeat panels every 24 hours thereafter while on infusion.

VWF, Activity	114	85	98	81	102 C	12 ↓ C
von Willebrand Factor, Antigen	314 ↑	313 ↑	274 ↑	292 ↑	280 ↑ C	115 C
VWF Multimers						
Factor VIII, Activity	184	189	160	205 ↑	190 C	70 C

- Neuraxial anesthesia was administered, using 1.6cc of 0.75% hyperbaric bupivicaine with 15mcg of intrathecal fentanyl and 0.1 mg morphine. Cesarean delivery was uneventful, with a total of about 700cc of blood loss. Post-operative course was also uneventful, with no further blood loss, neurologic sequelae, or reactions to plasma derived infusion of von Willebrand factor/Factor VIII complex.

Discussion

- **Preoperative Planning:**
  - **Joint collaboration between surgical, hematologic, and anesthetic teams is essential.**
    - **Patient had pre-operative meetings with the obstetric team, obstetric anesthesiologists, and hematologists to address pre-operative planning, risk discussions, and pre, intra, and post-operative dosing of VWF/FVIII infusion.**
  - **Plasma-derived von Willebrand factor/FVIII complex can be used to safely support neuraxial anesthesia in VWD Type 2A patients.**
- **Safety of Neuraxial Anesthesia:**
  - **With proper management and coordination, spinal anesthesia can be safely performed in patients with VWD Type 2A**
- **Outcome:**
  - **Patient was evaluated post-operatively by the anesthesiology team with no identified neurological sequelae.**
  - **Patient underwent a successful cesarean delivery without hemorrhage and had an uneventful recovery.**
  - **Labs obtained pre-operatively, intra-operatively, and post-operatively indicated a persistently normalized vWF activity while dosing was maintained. No adverse reactions to the infusion were observed.**