Perioperative Management of Suspected von Willebrand Patient

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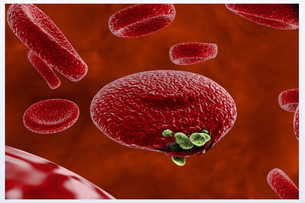

Introduction

Von Willebrand Disease (VWD) is the most common inherited bleeding disorder. There are different variations of the condition, and it is caused by either a decreased quantity or dysfunctional von Willebrand factor (vWF). In any type of VWD, the potential risk of mucocutaneous bleeding, joint and soft tissue bleeding, and postpartum bleeding can occur. In addition, many patients present with limited blood labs and specialized testing on preoperative evaluation.

This case report describes a suspected VWD patient who underwent a cesarean delivery, and our successful management of the patient despite limited preoperative information.

Case Report

33-year-old $G_2T_0P_1A_0L_0$ female with unclear past medical history presents at 37 weeks gestation for a repeat Cesarean section (C-section). She immigrated from Cuba a couple of months ago, and she presents at Kendall Regional Medical Center. In Cuba, her previous pregnancy lasted until 23 weeks. An amniocentesis was performed, and the procedure was complicated by amniotic fluid leakage and abnormally prolonged bleeding. Microsurgical cesarean section was performed and was treated with cryoprecipitate and platelet infusions. She was subsequently diagnosed with hemophilia.

Procedure

The case was performed under general anesthesia, using the modified rapid sequence induction and intubation (RSII). We induced with 50 mcg of fentanyl, 100 mg of lidocaine, 100 mg of propofol, and 100 mg of succinylcholine. After intubation, cesarean section immediately started. After 5 minutes, a baby boy was born. Patient had normal uterine tone, and 30 units of Pitocin was given. No intraoperative complications occurred and vital signs remained stable through the case. The patient was transported to Labor and Delivery.

Outcome

Postpartum complications were followed for five days and no postpartum bleeding was found. Daily CBC was normal. On postoperative day (POD) 0, the patient developed fevers (T_{max} = 38 C). Unfortunately, the patient was tested positive for COVID-19 on POD 3.

Discussion

Perioperative management of suspected vWD obstetric patient is challenging especially with limited information. Typically, vWD is diagnosed based on clinical history of prolonged bleeding, abnormal PTT and bleeding time, and abnormal specialized tests. However, pregnancy can affect the levels of vWF and factor VIII. It becomes even more complex, when the diagnosis of the bleeding disorder before the pregnancy is questionable. Even though this patient was diagnosed with hemophilia in Cuba and has family history of the condition, vWD was a better overall fit. vWD is a more common heritable condition than hemophilia, has autosomal dominance genetics that is more common in females, and can also be treated with cryoprecipitate similar to hemophilia.

Despite the questionable diagnosis, forming a comprehensive perioperative plan for a vWD patient is essential beforehand and preparing for potential complications. For induction, we proceeded with general anesthesia under RSII. Even though the vWF:RCo was normal, it was on the lower end of normal. Due to the potential risk of bleeding and hematoma in the intrathecal space, we did not proceed with spinal catheter placement. If any abnormal intraoperative or postpartum bleeding occurred, cryoprecipitate, which contains vWF, can be used or vWF/factor VIII combination.

References

1. Swystun LL, Lillicrap D. How much do we really know about von Willebrand disease? Current Opinion in Hematology. 2016 Sep;23(5):471-8. doi: 10.1097/MOH.0000000000000272. PMID: 27428891.

2. Von Willebrand disease in women. Committee Opinion No. 580. American College of Obstetricians and Gynecologists. Obstetric Gynecology. 2013;122(6):1368-1373.

3. RCSI researchers find potential new way to improve treatment for von Willebrand disease. (2020, October 29). Pharma World. https://www.pharmaworldmagazine.com/rcsi-researchers-find-potential-new-way-to-improve-treatment-for-von-willebrand-disease/