Anesthetic Management of Von Willebrand Disease Type III

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OBJECTIVES

- Identify the characteristics of different types of Von Willebrand Disease
- Understand the treatment of specific types of Von Willebrand Disease
- Discuss the anesthetic management of Von Willebrand's Type III

BACKGROUND

Von Willebrand Disease type III (VWD3) is the most serious form of the bleeding disorder. VWD type III is when there is a complete absence of Von Willebrand factor.

Von Willebrand factor (VWF) mediates platelet adhesion to broken blood vessels and promotes aggregation. VWF also binds to factor VIII and protects it from degradation by plasma proteases. VWD is an inherited autosomal recessive disease making type 3 very rare but serious due to its clinical implications.

CASE REPORT

Patient is a 38M that presented to the ED due to his vomit having blood streaks and multiple black stools. He has had a decreased appetite and had an episode of red mucous sputum. He has a history of lightheadedness/dizzy episodes. He was a heavy drinker of alcohol and smoked half a pack per day.

Medical History was significant for decompensated hepatic cirrhosis, recurrent GI bleeding, hemorrhoids, and Von Willebrand's Type III.

In the days prior while patient was being transferred to CTA to assess for active GI bleeding, he went into cardiac arrest with PEA. Most likely due to anemia with a hgb of 5.7. He received 4 rounds of CPR, epinephrine, and bicarbonate. Patient received 3 PRBCs, 1 unit of platelets, and emergent humate 3036 units. 1000 mg TXA was also given.

Anesthesia was called after several attempts at intubation were unsuccessful due to an edematous and bloody airway. The patient was actively aspirating. After suctioning the airway, a DL was performed and the airway was secured with an ETT successfully.

Patient arrived to the operating room for a bronchoscopy to remove blood clots in lungs the following day. On arrival the patient was intubated, on pressors (vasopressin, epi, and levophed) and on high PEEP/FiO2 requirements.

RESULTS

The anesthetic plan to perform the bronchoscopy was to keep the 7.5 ETT pending the proceduralist ability to remove the blood clots through the tube due to the prior difficult airway. If blood clots were too large we would attempt to switch the tube using a tube exchanger to ETT 8.0. We continued all pressors and performed an inhalation induction with sevoflurane. The patient's hgb prior to coming down was 6.6 so we immediately gave one unit of PRBCs and had 2 more units avilable in the room.

When the patient arrived we were having a difficult time keep his O2 saturations up due to blockage of the airway from the numerous blood clots. When switched from pressure control ventilation to giving breaths manually with the bag, the patient was able to achieve much larger tidal volumes. We continued this process of ventilation throughout the procedure. Due to the scope's suction limiting the amount of volume we were able to deliver, we had to communicate with the proceduralist to allow us time to give the patient breaths and improve saturation between insertion of the scope.

As the blood clots were removed from blocking the airway, it was much easier to ventilate the patient and larger tidal volumes were able to be delivered. At the end of the procedure we were able to leave the patient on pressure control ventilation and achieve adequate saturations. The patient remained intubated on the ventilator with 100% oxygen and returned to the ICU.

DISCUSSION

Symptoms of Von Willebrand Disease includes easily bruising, epistaxis, and bleeding from mucous membranes. Patients presenting for VWD3 usually have a history of bleeding since childhood, reduced plasma VWF, and autosomal recessive inheritance. It is important to understand bleeding history of family since VWDIII is a recessive trait therefore both parents would have a negative bleeding history.

To determine the best route of treatment the type of VWD the patient has must be determined. The goal of treatment for VWD is to correct the defect in platelet adhesion that is lost with the absence of VWF and anticoagulation due to low factor VIII levels. Numerically perioperative treatment should be aimed to increase VWF and factor VIII to 50-100% of normal levels. DDVAP is commonly used for treatment of VWD because it releases stored VWF and factor VIII. However, this is not a useful for treatment for patients with type III because they have a complete deficiency in VWF and therefore nothing is being stored to release. The most effective treatment for patients with type 3 VWD is plasma derived concentrates of VWF and factor VIII. These should always be on hand for patients undergoing surgery to stop any bleeding episodes that may occur or given prophylactically for surgery.

Preoperative blood tests can be used to determine the amount of VWF to diagnose the type of VWD. Some of these lab tests include VWF antigen, VWF activity, factor VIII activity, and bleeding time. These can be followed with assays to determine the classification.

Type	Underlying disorder	Clinical Presentation/Characteristics
1	Deficient quantity of vWF	Mucocutaneous bleeding, epistaxis, easy bruising, menorrhagia
2A	Defect in quality of vWF	Moderate bleeding
2B	Abnormal vWF	Moderate bleeding; thrombocytopenia; risk of thrombosis
2M	Abnormal vWF binding	Rare; significant bleeding
2N	Inactive vWF binding sites	May see low factor VIII and normal vWF levels
3	Severe deficiency of vWF	Severe bleeding, hemarthroses, muscle hematomas

DISCUSSION

Many factors should be considered before performing an anesthetic on a patient with von Willebrand disease. General anesthesia is preferred over regional due to increased risk of bleeding. When performing neuraxial anesthesia, special consideration should be given due to increase risk of hematomas.

Coagulation tests should be performed prior to see where the patient's factor levels are, especially VWF. Treatment should be given beforehand to reduce bleeding risk with surgeries. Always ensure the patient has a type and screen before surgery and blood is avilable. Consult the patient's hematologist or read their hematology notes before surgery.

Special attention should also be given to the airway to avoid any possible trauma. With an increased risk of bleeding, any trauma to the airway could result in blood in the airway. Using a videoscope may be preferred to avoid this risk.

CONCLUSIONS

When performing an anesthetic on a patient with Von Willebrand Disease, many factors need to be considered. Treatments may vary depending on the type of the disease. It is important to remember VWD type III is the only type that will not respond to DDVAP. Understanding the type and severity of the disease is vital in determining the correct treatment option.

Take all preoperative steps to understand the patient's coagulation status such as blood tests and consulting hematology. Be prepared for serious bleeding by having blood ready and anticoagulation medications specific for the patient. Avoid procedures that increase the risk of bleeding such as regional or neuraxial.

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