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Anesthetic Management of a Patient with Von Willebrand Disease

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Abstract

This case report describes an anesthesia-related issue encountered during a watchman device placement and transesophageal echocardiogram (TEE) where profuse perioperative esophageal bleeding ensued in a patient with Von Willebrand disease (VWD) Type 1. Prolonged bleeding of mucosal surfaces is common in patients with type I VWD and should be considered when attempting any procedure involving bruising or trauma to the oropharynx such as TEE or esophagogastroduodenoscopy (EGD).[1] The patient had also been taking aspirin, which can precipitate bleeding that may not have occurred otherwise. Patients with VWD often need a combination of multiple therapies to treat uncontrolled bleeding. Anesthesia providers should be aware of the risk of bleeding, premedication, and which medications and clotting factors to give next should desmopressin be insufficient.

Anesthetic Management of a Patient with Von Willebrand Disease

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Purpose

- This case report describes an anesthesia-related adverse event encountered in the hospital operating room with regards to profuse esophageal bleeding intra operatively and post operatively in a patient with Von Willebrand disease (VWD) Type 1. Preoperative assessment and intraoperative management are discussed.

Introduction

- This case is unique as it describes the most common inherited bleeding disorder, VWD, which affects up to 1 percent of the population, although only 0.1 to 1 percent of those individuals are clinically symptomatic (0.001 to 0.01 percent of the general population).[1]

Literature Search

- Majority of case studies regarding patients with VWD have type 2 or type 3 instead of type 1. Type 2 and type 3 VWD are considered higher risk of severe or uncontrolled bleeding. Patients with VWD Type 1 have rare cases of severe or uncontrolled bleeding.[7,8,9]

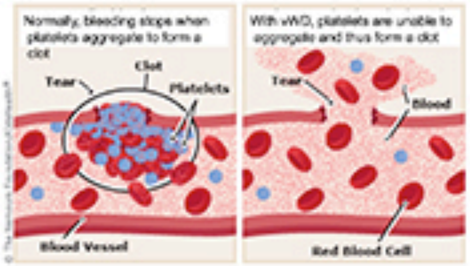
Von Willebrand Disease

- Von Willebrand disease is a bleeding disorder caused by decreased plasma von Willebrand factor (VWF) which affect clotting and hemostasis.[1]
- Three types of VWD with varying quantitative and, or qualitative deficiency of VWF
- Uncontrolled or severe bleeding may occur if the patient is not appropriately premedicated and treated aggressively.
- Antiplatelet and anticoagulants used in patients with atrial fibrillation present an additive risk for bleeding in patients with VWD[1]
- Anesthesia providers should be aware of the risk of bleeding and best possible treatment regimens should desmopressin be insufficient. This report highlights the importance of preoperative assessment, preparation, and optimization of a patient with a known bleeding disorder.

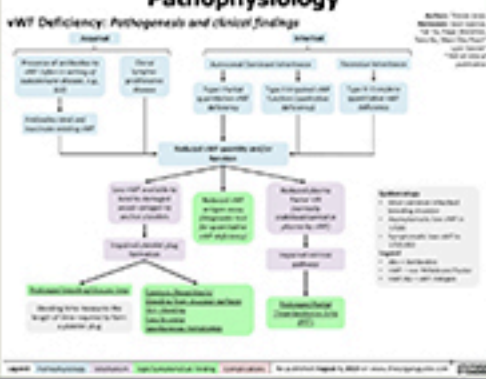
Takeaway

- Draw baseline labs, assess risk of bleeding, pretreat with DDAVP, have Humate-P available, anticipate second line treatments.

Abnormal Bleeding



Pathophysiology



Types of VWD

TYPE	DESCRIPTION	PEOPLE WITH VWD (%)	INHERITANCE PATTERN
Type 1	[illegible]	[illegible]	[illegible]
Type 2	[illegible]	[illegible]	[illegible]
Type 3	[illegible]	[illegible]	[illegible]

Clinical Snapshot

Patient Information

- Patient: 69 M
- Procedure: Watchman and Transesophageal Echocardiogram (TEE)
- PMH: atrial fibrillation, hypertension, and Von Willebrand Disease Type 1
- Medications: aspirin

Preoperative

- Patient had a previous episode of prolonged bleeding and bruising in the esophagus with a prior EGD twenty years ago in which he was hospitalized, but did not recall the treatment he received
- Lab work for activated partial thromboplastin time (aPTT) and VWF activity were not ordered, and the patient did not have a hematologist.
- Patient taking aspirin to prevent clots forming due to atrial fibrillation.
- Patient received one dose of desmopression (DDAVP) prior to intubation

Intraoperative

- Video laryngoscopy used to facilitate atraumatic intubation
- The patient noted to have a small amount of bleeding in the oropharynx
- TEE attempted twice with significant amount of bleeding in the esophagus, and the TEE was aborted
- Heparin 8,000 units given intravenously during the watchman procedure and was reversed with 30mg of Protamine intravenously
- The watchman procedure was completed, and the patient was transferred to recovery while intubated to protect the patient's airway from uncontrolled bleeding

Postoperative

- Two units of cryoprecipitate were transfused in recovery, and a hematologist was consulted
- The hospital facility did not carry plasma-derived VWF concentrate, Humate-P. A courier was sent to retrieve a dose of Humate-P from a surrounding hospital. While waiting for the VWF concentrate, an antifibrinolytic agent, one gram of tranexamic acid (TXA), was given intravenously to stabilize the clot by preventing clot breakdown.[2]
- A bedside EGD was performed to identify the source of bleeding which was a small mucosal tear in the posterior pharynx and upper esophagus area.
- Humate-P was transfused, and the patient transferred to cardiovascular intensive care unit for observation and hemostasis.
- An additional unit of Humate-P was transfused the next day
- Patient remained intubated x 1 day, extubated without complications.
- Patient made a full recovery post operatively

Discussion

- Prolonged bleeding of mucosal surfaces are common in patients with type I VWD and should be considered when attempting any procedure involving bruising or trauma to the oropharynx such as TEE or esophagogastroduodenoscopy (EGD).[1]
- The patient had also been taking aspirin, which can precipitate bleeding that may not have occurred otherwise.
- Patients with VWD often need a combination of multiple therapies to treat uncontrolled bleeding.[3]
- Anesthesia providers should be aware of the risk of bleeding, optimal treatments plan for premedication, and bleeding not responsive to desmopressin.

Treatment

First Line Treatment

Desmopressin (DDAVP)

- Induces synthesis of Von Willebrand Factor (VWF) by endothelial cells[3]

Humate-P

- Plasma-derived VWF
- Has undergone pathogen inactivation procedures[4]

Second Line Treatment

Cryoprecipitate

- Not typically used as a source of VWF as it has not undergone pathogen inactivation procedures that are used for the plasma-derived VWF concentrates such as Humate-P
- Useful in emergency if VWF concentrate is unavailable[2]

Tranexamic acid

- Antifibrinolytic agent that stabilizes the clot by preventing clot breakdown.[2]
- Particularly useful in areas of high fibrinolytic activity including the nose, oropharynx, or urogenital tract.[2]

Conclusion

- The patient was considered high risk for bleeding and should have had hematology consulted prior to surgery with baseline VWF activity, levels, and aPTT established.
- Humate-P should have been available site for use if desmopressin ineffective.
- TXA and Humate-P should have been transfused immediately after the TEE probe caused significant oropharyngeal bleeding and should not have been delayed in recovery.
- The patient had a known episode of oropharyngeal uncontrolled bleeding from a previous EGD and should have been optimized for the TEE procedure.[1,2,3]
- The medications required for hemostasis should have been anticipated and readily available.
- Heparin should have been held since the patient did not have an aPTT result and could have already been at a therapeutic level.

References

[illegible]