GOIANA JOURNAL OF MEDICINE

DOI: 10.29327/2396527.65.65-6

ISSN 0034-9585

SCIENTIFIC ARTICLE - CASE REPORT

GASTROPLASTY IN A PATIENT WITH VON WILLEBRAND DISEASE: CASE REPORT

FELIPE MENDES FARIA[1]; GUSTAVO SIQUEIRA ELMIRO[1,2]; GIULLIANO GARDENGHI[1,2,3]

1. Clínica de Anestesia - Goiânia GO
2. Hospital do Coração de Goiás (HCOR) - Goiânia GO
3. Hospital ENCORE - Aparecida de Goiânia GO

ABSTRACT

Von Willebrand Disease (VWD) is an autosomal hereditary bleeding disorder defined by decreased activity of Von Willebrand Factor in the blood. It may be secondary to a quantitative or qualitative defect. This case report addresses a patient with VWD and obesity who underwent gastroplasty at a Hospital in Goiânia. With a hematologist's recommendation for the use of factor VIII 2500 IU, to be performed 1 hour before the surgical procedure, with a subsequent maintenance dose for 5 days. In the operating room, total intravenous anesthesia was performed, with Propofol and Remifentanil. The procedure was uneventful and after 24 hours of hospital stay, the patient was discharged by the team.

Keywords: Von Willebrand Diseases; Anesthesia; Hematology.

INTRODUCTION

Von Willebrand Disease (VWD) is an autosomal inherited bleeding disorder defined by reduced von Willebrand factor (VWF) activity in the blood. It can be secondary to a quantitative or qualitative defect. VWD is characterized by three main subtypes: type 1, which is characterized by a partial quantitative deficiency of VWF; type 2, which is characterized by a qualitative deficiency; and type 3, which is characterized by a complete deficiency of VWF. [1]

Among the main symptoms are mucocutaneous bleeding, including epistaxis, easy bruising, and even heavy menstrual bleeding, as well as increased bleeding during surgical/invasive procedures. In this case, the patient has VWD and underwent a gastroplasty with a recommendation from the hematologist for factor VIII replacement before the surgical procedure. Another potential therapeutic option would be desmopressin, which stimulates the endothelial release of stored VWF and factor VIII, along with adjuvants such as tranexamic acid.[2]

The present case report aims to describe the clinical/anesthesiological management of a patient with VWD undergoing elective gastroplasty.

CASE REPORT

24-year-old female patient, ASA [2], with obesity, VWD, and generalized anxiety disorder, on continuous use of sodium valproate, lithium carbonate, and fluoxetine. She is willing to undergo gastroplasty

at a hospital in Goiânia. In the pre-anesthetic consultation, a hematologist's report was presented indicating the use of factor VIII 2500 IU, to be administered 1 hour before the surgical procedure, with a subsequent maintenance dose for 5 days. Additionally, in case of bleeding during or at the end of surgery, another dose was allowed immediately postoperatively. Total intravenous anesthesia was administered for the surgery, following all recommendations made by the hematologist.

In the operating room, the patient was properly monitored with a pulse oximeter with plethysmographic curve, electrocardiogram, and non-invasive blood pressure, presenting stable vital signs before anesthetic induction. She received a venous catheter with a 20G catheter in the right upper limb. She underwent total intravenous anesthesia: Preoxygenation with a facial mask with 100% oxygen (6L/min) for 3 minutes. Anesthesia was induced with propofol (150 mg), sufentanil (15 mcg), rocuronium (20 mg), and lidocaine (80 mg). A periglottic block was performed with ropivacaine 0.5%, 5ml of the solution, and the trachea was intubated with a 7.0 cuff tube. Cormack-Lehane laryngoscopic classification 2A (only the posterior portion of the glottic cleft visible), direct and atraumatic laryngoscopy confirming the correct tube placement by capnography, with mechanical ventilation adjusted to maintain an end-tidal carbon dioxide pressure (PETCO2) close to 35 mmHg. Anesthetic maintenance occurred with propofol and remifentanil target-controlled as per the physician's discretion. Adjuvant medications used were dipyrone 2g, cephalexin 4g, nausedrom 8 mg, buscopan compound, dexamethasone 10mg, ondansetron 8mg, pantoprazole 40mg, and precedex 50 mcg.

The intraoperative period proceeded without hemorrhagic complications, and the patient maintained stable vital signs throughout the anesthetic period. At the end of the surgical procedure, neuromuscular blockade was reversed with Sugamadex 200 mcg, and after a few minutes, the patient woke up followed by uneventful extubation. The patient was then transferred to the post-anesthetic care unit (PACU), where morphine 10mg was administered for post-operative analgesia. After 1 hour in the PACU, the patient was transferred to a hospital room, where she stayed for 24 hours, receiving the factor VIII as instructed by the hematologist. Due to her good condition and absence of bleeding, the patient was discharged from the hospital.

DISCUSSION

Von Willebrand Disease (VWD) is the most common autosomal hereditary bleeding disorder along with hemophilia A, with an estimated prevalence of 1 in 1,000 individuals. It can be better determined by the decrease in von Willebrand factor (VWF) activity in the blood, which can be secondary to a quantitative or qualitative alteration. [1,3,4]

The VWF is a multimeric plasma glycoprotein whose main functions are to facilitate platelet adhesion to the injured vascular endothelium by binding to the platelet membrane, as well as to act as a carrier and stabilizer of factor VIII in the plasma. [5]

There are several subtypes of VWD that require individualized treatment based on the specific diagnosis, bleeding phenotype, and specific clinical context. The main symptoms include mucocutaneous bleeding, including epistaxis, easy bruising, menorrhagia, as well as bleeding during surgery and other invasive procedures. [2]

VWD is classified into 3 main categories: partial quantitative deficiency of VWF (type 1), complete deficiency (type 3), and qualitative deficiency (type 2). Type 2 is further classified into subtypes defined by defects in multimerization (type 2A), increased platelet binding (type 2B), defects in VWF-platelet or

VWF:CB binding (type 2M), or defects in factor VIII (FVIII) binding (type 2N). Type 1 is the most common, accounting for about 85% of VWD, while type 3 is the least common, affecting about 1 in 1 million individuals. The diagnosis of VWD includes evaluation of the history of hemorrhagic symptoms, assessment of the family history of bleeding or VWD, and confirmatory laboratory tests. [1]

Clinical laboratory tests for VWD include initially measuring at least VWF:Ag, VWF-platelet binding activity (VWF:RCo, VWF:GPIbM, and VWF:GPIbR), and FVIII levels. Additional tests may be indicated based on the results of the initial test, including low-dose ristocetin-induced platelet aggregation (VWF:RiCo), VWF multimers, and von Willebrand factor propeptide (VWFpp) levels. [1,6]

In a case described by Saurote, a case of a patient with von Willebrand disease (VWD) undergoing a mitral valve replacement is presented. Similar to the reported case, intensive care was provided in the preoperative, intraoperative, and postoperative periods with factor VIII replacement. At the end of the process, the patient progressed without complications and with good clinical evolution, allowing her to be discharged after a few days of hospitalization and observation. [5]

The main therapies include the use of desmopressin to induce the endothelial release of stored von Willebrand factor (VWF) and factor VIII and the use of VWF concentrates, including plasma-derived and recombinant products, as well as adjunctive therapies such as the antifibrinolytic agent tranexamic acid. Management remains challenging due to the significant variability in bleeding symptoms among patients, variability in clinical practice, and lack of high-quality evidence to guide decision-making. [2]

CONCLUSION

VWD is an autosomal hereditary disease, as stated, increasing the risk of bleeding intraoperatively and postoperatively, therefore, it can be concluded that good preventive medical care is essential to avoid complications. Strengthening bonds with assisting physicians, especially hematologists, and with the patient herself in a prior consultation is of notable importance for her care in the surgical center, as with these measures, a more dignified care with fewer risks to the patient can be planned. In the case reported here, the use of factor VIII in the pre-, peri-, and postoperative periods was able to prevent undesirable bleeding.

REFERENCES

1- Sharma R, Haberichter SL. New advances in the diagnosis of von Willebrand disease. Hematology Am Soc Hematol Educ Program [Internet]. 2019(1):596-600. doi: 10.1182/hematology.2019000064

2 - Connell NT, Flood VH, Brignardello-Petersen R, Abdul-Kadir R, Arapshian A, Couper S, et al. ASH ISTH NHF WFH 2021 guidelines on the management of von Willebrand disease. Blood Adv [Internet]. 2021 Jan 12; 5(1):301-25. doi: 10.1182/bloodadvances.2020003264

3 - Mannucci PM. New therapies for von Willebrand disease. Blood Adv [Internet]. 2019 Nov 12;3(21):3481-7. doi: 10.1182/bloodadvances.2019000368

4 - Weyand AC, Flood VH. Von Willebrand disease: current status of diagnosis and management. Hematol Oncol Clin North Am [Internet]. 2021 Dec;35(6):1085-101. doi: 10.1016/j.hoc.2021.07.004

5 - Saroute ANRS, Brandão CMA, Guedes MAV, et al. Patient with von Willbrand disease undergoing mitral valve repair: a strategy for the control of the coagulopathy: Arq Bras Cardiol [Internet]. 2007 Jan;88(1):e3-e5 doi: 10.1590/S0066-782X2007000100022

6 - Fogarty H, Doherty D, O'Donnell JS. New developments in von Willebrand disease. Br J Haematol. 2020 May 12;191(3):329-39. doi: 10.1111/bjh.16681

FELIPE MENDES FARIA - http://lattes.cnpq.br/7891778400395141 - https://orcid.org/0000-0003-1498-906X

GUSTAVO SIQUEIRA ELMIRO - http://lattes.cnpq.br/4765163399934337 - https://orcid.org/0000-0003-2113-8757

GIULLIANO GARDENGHI - http://lattes.cnpq.br/1292197954351954 - https://orcid.org/0000-0002-8763-561X

ADDRESS

GIULLIANO GARDENGHI
CET – CLIANEST, R. T-32, 279 - St. Bueno, Goiânia - GO, Brasil, CEP: 74210-210
E-mail: coordenacao.cientifica@ceafi.edu.br

Library Review - Romulo Arantes
Spell Check: Dario Alvares
Received: 28/03/24. Accepted: 02/04/24. Published in: 26/04/24.