Case Report

Von Willebrand Disease in Colostomy Stoma Closure – A Case Report of Perianaesthetic Management

Mishuk Dutta[1] , Salahuddin Al Azad [2], Nahida Parveen Nimmi[3], Azharul Islam[4], Lutful Aziz[5]

[1]Registrar, Dept of Anaesthesia & Pain Medicine, Apollo Hospitals Dhaka, [2]Registrar, Dept of Anaesthesia & Pain Medicine, Apollo Hospitals Dhaka, [3]Sr. Registrar, Dept of Anaesthesia & Pain Medicine, Apollo Hospitals Dhaka, [4]Senior Consultant, Dept of Anaesthesia & Pain Medicine, Apollo Hospitals Dhaka, [5]Senior Consultant, Dept of Anaesthesia & Pain Medicine, Apollo Hospitals Dhaka

Abstract

Von Willebrand disease (VWD) is the most common inherited bleeding disorder that is caused by deficiency or dysfunction of von Willebrand factor (VWF), a plasma protein that mediates the initial adhesion of platelets at sites of vascular injury and also binds and stabilizes blood clotting factor VIII (FVIII) in the circulation.[1] The prevalence of VWD is about 1% of the population & approximately twice in female then man because of their unique potential of menorrhagia.[2] We are reporting a successful perioperative anaesthetic management of 33year female, a known case of Von Willebrand disease (VWD) who underwent routine colostomy closure.

(JBSA 2019; 32(2): 83-)

Introduction

Von Willebrand disease (vWD) is the most common inherited coagulopathy. The disease results from either a deficiency or abnormality in von Willebrand factor (vWF), a glycoprotein found in blood plasma which is necessary for platelet aggregation and maintenance of factor VIII levels, which ultimately lead to clot formation at the point of the endothelial lining injury. Complications of vWD include abnormal bleeding from minor cuts and procedures, anemia, bruising, epistaxis, heavy menstrual bleeding, swelling, and joint pain. Bleeding episodes are usually less frequent than in severe haemophilia & excessive haemorrhage may be observed only after trauma or surgery. Within a single family, the disease has variable penetrance, so that some members may have quite severe & frequent bleeds, whereas others are relatively asymptomatic.[3]

Case Summary

A 33year old woman, weighted 61 kg & 160 cm height admitted to the hospital for a routine Closure of Colostomy stoma. She was diagnosed as heterozygous Von Willebrand disease (VWD) type 1 & 2, three year back when she came to the hospital with the complain of paleness & severe menorrhagia. She gave history of consanguinity & confirmed that her father was diagnosed as hemophilia A & unfortunately passed away followed by factor VIII transfusion after trans urethral resection of prostate(TURP) surgery .She didn't have any other medical co morbidity.

She attempted IVF for five times of which 4th time was successful but ended with miscarriage. She gave history of medical admission for back pain & haematuria two years ago & diagnosis was portal vein thrombosis. The reason behind it was allergic reaction from post IVF female hormone therapy for which she was treated for low molecular weight heparin & the follow up CT after two months was normal.

Her surgical history includes, cystectomy for polycystic ovary in 2013, She had undergone total abdominal hysterectomy & left sided sulfingooophorectomy, right sided oophorectomy, adhesiolysis & repair of rectum with colostomy because of pelvic endometriosis,severe adhesion & rectum injury. Last surgery was event full, required multiple bag of cryoprecipitate, apheretic platelet & whole blood. She gave no history of drug or food allergies. Iron infusion, PPI & multivitamins were the regular taking medication.

Multidisciplinary management comprising haematologist, surgeon, anaesthetist were dealing the patient. preoperative anesthesia evaluation revealed the patient had a Mallampati class 1 airway. Preoperative laboratory values were hb 13.7gm/dl, platelet 166000/L, TLC 4740/L, BT 4min 30 sec, CT 8min 30 sec, PT 12sec, APTT 32.2 sec, TT 16.6 sec, INR 1.00. Blood grouping & cross matching done & liaison with laboratory services ensures appropriate factor concentrates are available and in sufficient quantity will be ready before the procedure. Haematologist advised to infuse 1unit aphaeretic platelet,4unit cryoprecipitate before procedure & inj. Tranexamic acid 500 mg 6 hourly. NPO status was confirmed. Patient received inj glycopyrrolate .2 mg half hour before procedure. After administration of the blood product patient was shifted to operation theatre & then her pulse rate was 82beats/min, B/P 110/60mmHg & room air oxygen saturation was 98%. Standard monitors were attached. General anaesthesia was the choice of technique.

Preoxygenation was done by 100% oxygen with tight fitting face mask & was confirmed by ETO2 more than 90%. Co-induction was done by propofol, midazolam, fentanyl. Intubation was facilitated with rocuronium I/V 30 mg. After obtunding the airway reflex, intubation done with 7.5 ID PVC tube by video laryngoscope without any airway injury. Analgesia was maintained by fentanyl, I/V paracetamol & local infiltration of local anaesthetic in the operation field. Maintenance of anaesthesia was continued by inhalational anaesthetic agent(sevoflurane), intermittent administration of muscle relaxant(rocuronium) & judicial use of fluid to maintain the hemodynamic status. The surgery was very complex because of severe adhesion & continuous oozing from operation site & lasted for two hours. Another 6unit cryoprecipitate & 1unit whole blood was transfused to combat the situation. After the procedure patient was reversed & deep extubation was done with inj neostigmine & glycopyrrolate. Vitals were stable throughout the surgery Patient was shifted to ICU for close monitoring.

In ICU patient's hemodynamic status was stable & she was conscious, oriented. Postoperative analgesia was maintained by morphine PCA (patient-controlled analgesia) & all platelet inhibiting medication & I/M injections are strictly avoided. Hematologist advised to transfuse 5unit cryoprecipitate 12hourly total 10unit per day for the next four days.

She was observed in ICU for 24 hour & then shifted to word as her condition was stable & was no other complication. Her coagulation profile was monitored daily. Her factor level was stable & within the normal range, so she was discharged in her sixth postoperative day.

Discussion

Pathophysiology:

Von Willebrand disease (VWD), first described by Erik von Willebrand in 1926, is the most common inherited autosomal dominant bleeding disorder in humans is the most common but usually mild bleeding disorder caused by either a quantitative or qualitative defect in von Willebrand's factor (VWF). VWF is a plasma glycoprotein which plays a vital role in platelet adhesion, aggregation and, acts as a carrier for factor VIII and thereby decreasing its clearance from plasma. VWF is synthesized in bone megakaryocytes and vascular endothelium and stored in Weibel–Palade bodies in the endothelial cells.it normally forms a multimeric structure that is essential for its interaction with subendothelial collagen & platelets. von Willebrand factor act as a carrier protein for factor VIII, to which it is noncovalently bound.so deficiency of von Willebrand factor lowers the plasma factor VIII level. von Willebrand factor also form bridges between platelet & subendothelial components, allowing platelets to adhere to damaged vessel walls.[4] Deficiency of VWF therefore leads to impaired platelet plug formation resulting easy bruising from trauma; in particular, bleeding from mucosal surfaces, that is, epistaxis, gums, and bowel. Blood group antigens (A & B) are expressed on vWF, reducing its susceptibility to proteolysis: as a result, people with blood group O tend to have low VWF levels than individuals with non O groups.[3]

Classification

There are three major types of VWD disease. Type 1, the most frequent form, is characterized by a partial quantitative deficiency in von Willebrand

factor (VWF). Type 2 is a qualitative deficiency, and Type 3 is a virtually complete deficiency. Type 2 VWD is divided into four subtypes. Type 2A includes variants with decreased platelet adhesion caused by a selective deficiency in high-molecular weight VWF multimers (HMWM). Type 2B includes qualitative VWF variants with increased affinity to platelet glycoprotein Ib. Type 2M includes variants with decreased platelet adhesion, but without HMWM deficiency, and type 2N includes variants with markedly decreased affinity for factor VIII.[5]

Diagnosis

The diagnosis of VWD requires clinical and laboratory components, a personal history of excessive mucocutaneous bleeding, a family history of excessive bleeding, and a laboratory evaluation that is consistent with a quantitative and/or qualitative defect in VWF. An initial hemostasis laboratory evaluation usually includes a platelet count and complete blood count (CBC), PTT, prothrombin time (PT), and optionally either a fibrinogen level or a thrombin time (TT). This testing neither "rules in" nor "rules out" VWD, but it can suggest whether coagulation factor deficiency or thrombocytopenia might be the potential cause of clinical bleeding. If the mucocutaneous bleeding history is strong, consider performing initial VWD assays (VWF:Ag, VWF:RCo,VWF:RCo and/or VWF:CB and FVIII) at the first visit.A definite diagnosis of VWD type 1 is performed when VWF:Ag is < 30 IU/dL, in association with bleeding symptoms.An abnormal VWF:RCo/VWF:Ag ratio (< 0.6) is a simple way to suspect type 2 VWD.[1,5]

Treatment

VWD therapies follow three general strategies. The first aims at increasing plasma concentration of VWF through an endogenous release by desmopressin. The second strategy uses agents that improve haemostasis (tranexamic acid, amino-caproic acid), without modifying plasma levels of VWF. The third approach aims at replacing VWF by human plasma-derived, virus-inactivated concentrates.[2]The appropriateness of therapeutic choice depends on VWD severity and type, severity of the haemostatic challenge, and nature of the actual or potential bleeding. For minor surgery, prophylaxis should achieve VWF:RCo and FVIII:C levels e" 50 IU/dL on the day of surgery and during the first postoperative day, and > 30 IU/dL during 2 to 5 days thereafter. For major surgery, such as cardiac or neurosurgery, the levels of VWF:RCo and FVIII:C should be around 100 IU/dL on the day of surgery and during the first postoperative day, and should be maintained e" 50 IU/dL for 7 to 14 days or until healing is complete.[5]

Desmopressin

Desmopressin stimulates VWF release through its agonist effect on vasopressin V2 receptors. FVIII levels also increase acutely following its administration. Desmopressin is usually effective in Type 1 VWD. Type 2A patients rarely respond relevantly. Type 2B patients were previously considered as a contraindication to desmopressin. The reason was a frequent fall in platelet count after desmopressin stimulation.[5]

Antifibrinolytic agents:

Currently, tranexamic acid (TXA) is the most widely used antifibrinolytic agent. The drug inhibits the conversion of plasminogen into plasmin, thereby stabilizing previously formed clots. TXA can be used orally or intravenously. Dose and administration mode vary among teams. Intravenously, the bolus dose of TXA is 10-15 mg/Kg repeated every 8-12 hours or followed by a maintenance infusion of 10 mg. Kg-1.h-1.[6]

Replacement therapy:

Cryoprecipitate is administered in a dose of 1 unit/5-6 kg, which raises the Factor VIIIC level by 15-20%. During the intraoperative period, the consumption of vWF is increased, and may require the administration of cryoprecipitate as frequently as every 6-8 hours in contrary to every 8-12 hours postoperatively, after reviewing the bleeding time and the clinical response.

Fresh frozen plasma (FFP) corrects the abnormality similar to cryoprecipitate, but administration may be associated with circulatory volume overload. Usually 20 ml.kg-1 FFP every eight hours will control clinical bleeding.

Transfused platelets do not correct the BT but may be required in addition to cryoprecipitate in a patient who is thrombocytopenic (<50-80 x 109 L-1) secondary to blood loss or plasma dilution during surgery.[6]

Each patient should be managed individually, on a case-by-case basis, according to his/her sub-type of VWD, severity, in centers where a multidisciplinary team and daily laboratory testing of concerned factors are available. General anaesthesia is often preferred in these patients.No formal recommendations exist, and contraindications are relative for regional anaesthesia.a neurological postoperative surveillance is mandatory due to increased risk of developing epi-medullar haematoma and compression of neurological structures when neuraxial technique be used.

Traumatic oro-tracheal intubation (OTI) should be avoided. In case of difficult intubation, the use of a fiberscope or videolaryngoscope may reduce the risk of bleeding and mucosal lesions. Risk of nasal/ urethral bleeding during Ryle's tube and foley's catherter insertion should be kept in mind. Oral route for Ryle's tube may preferred. Patient with VWD should be advised to avoid intramuscular injection and any type of platelet-inhibiting medications. Low dose heparin prophylaxis should be considered in the peri-operative period of surgeries with a high thrombotic risk, especially when replacement treatment is administered.[5,6]

In summary, though we always don't have access for VWF:Ag, VWF:RCo or Ristocetin co-factor and/ or collagen binding activity (VWF:RCo and/or VWF:CB) for ideal management, patients with vWD do not carry an increased operative risk during elective procedures. The mainstay of success depends on appropriate preoperative evaluation, multidisciplinary team management, judicial administration of prophylactic & corrective therapy.

References

1. Barbara Yawn, William L. Nichols, Margaret E. Rick, Diagnosis and management of von Willebrand disease: guidelines for primary care.Am Fam Physician. 2009 Dec 1; 80(11): 1261–1268.
2. David Lillicrap, Paula James. von Willebrand Disease: An Introduction for the Primary Care Physician.jan 2009 • NO 47
3. Ralston, S., Penman, I., Britton, R., Strachan, M., Hobson, R. and Davidson, S. (2018). *Davidson's principles and practice of medicine*. 23rd ed. Elsevier, pp.974.
4. de Boer, Suzan; Eikenboom, Jeroen.Von Willebrand Disease: From In Vivo to In Vitro Disease Models.HemaSphere: October 2019 - Volume 3 - Issue 5 - p e297
5. Vincent Bonhomme, Aline Defresne,Isablle Maquoi.Anaesthesia recommendations for patients suffering from Von Willebrand disease.orphananesthesia.October,2014
6. Rakesh Garg, Mridu Paban Nath, Sanjay Verma, Ashwani Kumar.Patient with von Willebrand Disease for Gynaecologic Surgery - Perianaesthetic Concerns.Indian Journal of Anaesthesia 2008; 52 (5):573-576