Case report

POSTOPERATIVE HEMORRHAGE CAUSED BY UNDIAGNOSED VON WILLEBRAND DISEASE: A CLINICAL CASE REPORT

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Rezume Postoperative hemorrhage remains one of the most dangerous complications in the immediate postoperative period. Among its causes, von Willebrand disease (VWD) occupies a significant place. Management becomes particularly challenging when surgeons encounter previously undiagnosed coagulopathy during urgent surgery.

Von Willebrand disease is an inherited bleeding disorder characterized by quantitative or qualitative defects of von Willebrand factor (VWF). VWF mediates primary platelet adhesion to damaged endothelium, supports platelet aggregation, initiates thrombus formation, and stabilizes coagulation factor VIII in plasma.

Timely recognition of VWD in surgical practice remains difficult because routine preoperative coagulation tests—including PT, PT%, INR, APTT, TT, and fibrinogen—often remain within normal limits and fail to raise diagnostic suspicion.

We report the case of a 77-year-old male who underwent emergency open mesh hernioplasty for an incarcerated indirect inguinal hernia. During the postoperative period, the patient developed a progressively enlarging subcutaneous hematoma that required surgical re-exploration. Intraoperatively, the surgical team identified no active bleeding source or technical cause. Subsequent hematologic evaluation confirmed Type 1 von Willebrand disease. Targeted conservative hemostatic therapy controlled the bleeding and led to clinical improvement.

Key words: Postoperative hemorrhage, von Willebrand Disease, inguinal hernia

INTRODUCTION

Postoperative hemorrhage occupies a leading position among life-threatening complications encountered in surgical practice [5, 6, 11]. Owing to its aggressive nature, postoperative bleeding can rapidly and dramatically deteriorate a patient's condition, frequently resulting in hypovolemic shock and acute anemia. Such complications often necessitate repeated surgical intervention and transfusion of blood components, impair reparative processes, weaken anti-infective defense mechanisms, worsen overall treatment outcomes, increase rates of disability and mortality, prolong hospitalization, and significantly raise the cost of care.

Despite advances in modern surgical technology, the introduction of high-precision minimally invasive techniques, and the improved effectiveness of contemporary therapeutic measures, postoperative bleeding remains a relevant and challenging clinical problem.

The reported incidence of postoperative hemorrhage varies widely in the international literature, reflecting the influence of numerous contributing factors. These include: the nature of the primary disease, the profile and complexity of the surgical procedure, technical intraoperative difficulties, the trophic condition of tissues in the surgical field, the extent of inflammatory or neoplastic involvement, patient age, severity of comorbidities, the necessity of perioperative anticoagulant or antiplatelet therapy, and the urgency of the operation—which may limit the time available for comprehensive preoperative assessment and optimization.

In addition to the above factors, one should give particular attention to disorders of hemostasis, which significantly increase the risk of postoperative bleeding [5, 6, 11]. Among these, von Willebrand disease (VWD) represents a distinct clinical entity characterized by impaired coagulation and platelet adhesion. It therefore deserves special consideration as a potential underlying cause of unexpected hemorrhagic complications in the postoperative period.

Although VWD is considered relatively common, its true prevalence remains insufficiently defined [7]. According to various literature sources, reported prevalence ranges from 0.6% to 2% in the general population [2, 7, 8]. The distribution of individual VWD subtypes, however, demonstrates even greater variability across statistical reports [1, 2, 8, 9].

The pathophysiology of VWD is complex. Three major types of the disease are recognized [1, 7, 9]:

- Type I — accounting for 70–80% of cases, is inherited in an autosomal dominant pattern and is characterized by a quantitative deficiency of von Willebrand factor (VWF). In this form, the protein's structure and functional integrity are preserved [1, 5].
- Type II — observed in 7–25% of patients, is also typically inherited in an autosomal dominant manner. In this subtype, plasma VWF levels are usually normal or

near normal, but the protein is functionally abnormal. The literature describes more than 20 variants of Type II, the clinically most relevant include Type 2A-AD, 2B-AD, 2M-AD, and 2N-AR. [1, 5, 9].

- Type III — the most severe and least common form, accounting for 5–20% of cases, is inherited in an autosomal recessive pattern and is characterized by a near-complete or complete absence of VWF. This subtype poses the highest bleeding risk, as even minimal trauma or surgical intervention can lead to life-threatening hemorrhage [4].

VWD is primarily a hereditary coagulation disorder, but in rare cases it may develop as an acquired condition secondary to other diseases—most commonly lymphoproliferative, myeloproliferative, or autoimmune disorders. This form, known as acquired von Willebrand syndrome, results from either reduced synthesis of VWF or accelerated clearance of VWF from the circulation [2, 3, 9, 10].

The underlying cause of hereditary VWD is a mutation in the gene that regulates VWF synthesis [2]. Von Willebrand factor is a multimeric glycoprotein essential for primary hemostasis, mediating platelet adhesion to damaged vascular endothelium, promoting subsequent platelet aggregation, and initiating thrombus formation.

Additionally, VWF plays a crucial role in transporting and stabilizing coagulation factor VIII, thereby supporting normal plasma levels of factor VIII [2, 3, 7, 9].

The pathognomonic clinical manifestation of VWD is spontaneous or prolonged bleeding, occurring either without provocation or following physiological and pathological triggers such as trauma, surgery, or dental procedures. Patients frequently exhibit easy bruising (ecchymoses) and have a marked predisposition to recurrent epistaxis, menorrhagia, hematuria, and melena or hematochezia [5, 6].

Notably, a subset of patients may have no prior history of clinically significant bleeding episodes, and therefore may remain undiagnosed with VWD until a hemostatic challenge occurs [2].

Routine coagulation studies—such as prothrombin time (PT), prothrombin activity (PT%), international normalized ratio (INR), activated partial thromboplastin time (APTT), thrombin time (TT), and plasma fibrinogen concentration—are frequently within normal limits, or altered only minimally. As a result, standard coagulation profiles often fail to raise diagnostic suspicion and are generally insufficient for early recognition of VWD.

Accurate laboratory diagnosis relies on specific assays, including:

1. von Willebrand factor antigen (vWF:Ag);
2. Ristocetin cofactor activity of von Willebrand factor (vWF: RCo);
3. Factor VIII procoagulant activity (FVIII: C).

When indicated, additional studies may include:

- von Willebrand factor collagen-binding activity (vWF: CB)
- VWF multimer analysis
- Platelet aggregation studies using major inducers (ristocetin, thrombin, adrenaline, ADP, and collagen).

Because genetic defects cause VWD, it is not a curable disorder. Management is symptom-directed and indicated primarily in situations of active bleeding or when invasive procedures are planned, including surgical or dental interventions [6, 8]. The therapeutic objective is to increase plasma VWF levels using vasopressin analogues (e.g., desmopressin) in cases of quantitative deficiency, or to provide factor replacement therapy when functional defects are present [5, 6, 8]. To prevent premature fibrinolysis and enhance clot stability, antifibrinolytic agents are also employed [5, 6, 8].

CLINICAL CASE PRESENTATION

A 77-year-old male was brought to the Emergency Department of a University Clinic by the national emergency medical response team (service 112) as an urgent case. Upon admission, the patient was conscious, cooperative, fully oriented, and hemodynamically stable, and was complaining of persistent, severe pain in a large mass located in the left inguinal region. The patient also reported intermittent, colicky, moderate abdominal pain, dry mouth, abdominal distension, nausea, multiple episodes of vomiting containing gastric chyme, and generalized weakness.

According to the patient's history, a large left inguinal hernia had developed over the past nine years. The hernia reportedly enlarged with increases in intra-abdominal pressure, frequently descending into the scrotum, and, while supine, could be manually reduced completely back into the abdominal cavity. Approximately four hours prior to hospital admission, following a severe coughing episode caused by accidental aspiration of water, the hernia became irreducibly incarcerated, after which the aforementioned symptoms progressively developed, prompting emergency hospitalization.

Physical Examination:

The patient had a normosthenic body habitus. The tongue was dry, the abdomen was distended, participating minimally in respiration. Auscultation revealed intermittent hyperactive bowel sounds, while percussion demonstrated pronounced tympany, with preserved hepatic dullness. On superficial and deep palpation, the abdomen was soft but diffusely tender, without signs of peritoneal irritation. In the left inguinal region, there was a fixed, firm, markedly tender mass with a smooth surface, measuring 13.0x9.5x7.5 cm, with its distal third extending into the scrotum. could no longer reduce the hernia manually into the abdominal cavity, and the cough impulse was negative. A nasogastric tube drained enteric fluid consistent with small bowel contents. Plain abdominal X-ray demonstrated multiple air-fluid levels consistent with mechanical small bowel obstruction. Ultrasonography of the inguinal region revealed small bowel loops within the hernia sac, along with a small amount of free fluid.

Laboratory Findings:

Complete Blood Count (CBC): WBC: 30.34x10^9/L, NEUT: 92.8%, RBC: 7.90x10^{12}/L, HGB: 14.1 g/dL,

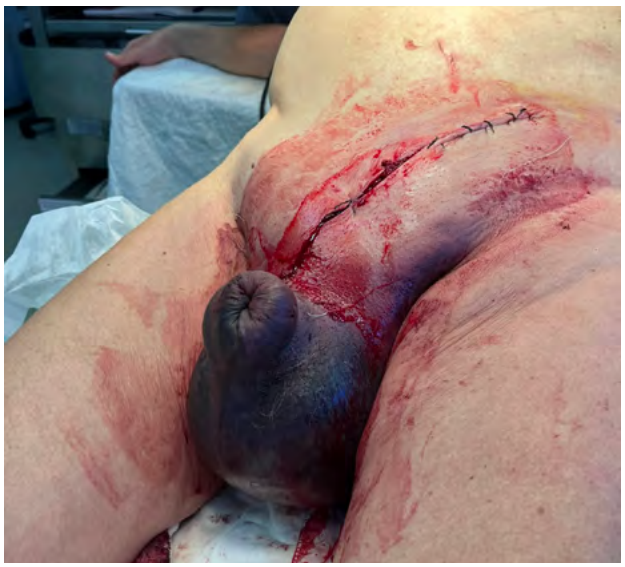

Figure 1. Postoperative hematoma prior to the second surgical intervention.

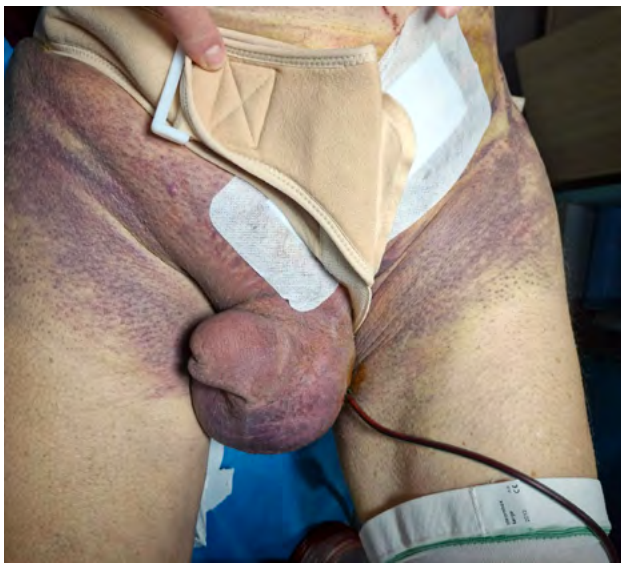

Figure 2. Ten days after the second surgical intervention.

HCT: 48.0%, MCV: 60.80 fL, MCH: 17.90 pg, PLT: 694.00x10^9/L, PCT: 0.67x10^{-2} L/L;

Arterial Blood Gas (ABG): pH: 7.40, pCO_2: 35.40 mmHg, pO_2: 99.10 mmHg, HCO_3 (act): 21.50 mmol/L, HCO_3 (standard): 22.30 mmol/L, BE (B): –2.60 mmol/L, BE (act): –3.30 mmol/L, $ctCO_2$: 22.60 mmol/L, SaO_2: 97.60%, FOHb: 94.70%, FCOHb: 1.90%, FMetHb: 1.10%, Lac: 1.57 mmol/L, Na^+: 136.00 mmol/L, K^+: 3.93 mmol/L, Cl^-: 103.00 mmol/L, Ca^{2+}: 1.08 mmol/L, Glucose: 110 mg/dL;

Coagulation profile: PT = 14.20 sec, PT% = 70.60%, INR = 1.24.

Based on the clinical evaluation, the surgical team diagnosed: unilateral or unspecified inguinal hernia with obstruction, without gangrene (ICD-10 - K40.3). After appropriate preoperative preparation, the team performed an emergency operation under balanced anesthesia (WAA408): reconstructive surgery for inguinal hernia (NCSP - JASB10).

Upon opening the hernia sac, the surgeons identified approximately 150 mL of yellowish, odorless serous fluid and loops of small intestine. They aspirated the fluid. After confirming bowel viability, they freely reduced the intestinal loops into the abdominal cavity. The surgeons then performed hernia repair using the Lichtenstein technique. The operation proceeded without technical difficulties or intraoperative complications, and the surgeons achieved complete hemostasis.

During the postoperative period, the patient developed a progressively enlarging subcutaneous hematoma at the operative site. Despite the immediate application of local hypothermia, the hematoma gradually expanded over several hours, first involving the scrotum and left inguinal region and eventually the entire hypogastrium (Figure 1, Figure 2).

These postoperative changes strongly suggested ongoing vascular bleeding and prompted wide re-exploration of the herniotomy and hernioplasty area. Intraoperatively, the surgeons identified no discrete source of active bleeding other than diffuse, low-intensity oozing from the postoperative tissues. This finding raised suspicion of a possible coagulopathy.

Subsequent additional investigations, performed in collaboration with a hematologist, confirmed the presence of Type 1 von Willebrand Disease (VWD). Administration of antifibrinolytic therapy and agents stimulating von Willebrand Factor (VWF) synthesis successfully achieved hemostasis.

DISCUSSION

Identifying VWD can be particularly challenging in urgent or emergency surgery if clinicians have not previously diagnosed it. In such cases, the limited time available before surgical intervention often makes comprehensive preoperative evaluation difficult, particularly in emergency operations. From a diagnostic and preoperative preparation standpoint, the specificity of VWD lies in the fact that standard coagulation tests (e.g., routine coagulation panel) frequently fail to detect this disorder. reflected This pattern in the presented case: the preoperative coagulation profile did not reveal abnormalities (PT = 14.20 sec, PT% = 70.60%, INR = 1.24), and the marked thrombocytosis (PLT = 694x10^9/L, PCT = 0.67x10^{-2} L/L) instead suggested an increased risk of thrombosis, rather than bleeding.

Therefore, in suspecting VWD, a thorough and precise medical history (anamnesis) becomes critically important. However, this step also has inherent limitations. Prior to surgery, a patient may fail to recall or may underestimate previous episodes of abnormal bleeding, providing incomplete or misleading information. Additionally, a subset of VWD patients may never have experienced clinically significant bleeding or may never have received an official diagnosis, despite having the disease.

In the presented case, as later confirmed through external sources, the patient had experienced prior bleeding episodes but had intentionally concealed this history. He had never sought medical attention and had not under-

gone evaluation, which delayed recognition of the underlying coagulopathy.

In cases of postoperative bleeding at the surgical site, clinicians typically first suspect a procedural technical error. Consequently, this often compels the surgeon to perform a repeat and essentially "unnecessary" reoperation, even though von Willebrand Disease-caused bleeding requires an entirely different, conservative treatment approach. Suspicion of a coagulopathy usually arises only after intraoperative re-exploration fails to reveal a technical cause of bleeding. Must also emphasize that additional surgical trauma inflicted during reoperation may further aggravate the bleeding.

CONCLUSION

- In general surgery, VWD represents a significant risk factor for life-threatening postoperative hemorrhage.
- Normal routine coagulation test results do not exclude the presence of VWD or the possibility of massive postoperative bleeding.
- Since standard coagulation tests cannot reliably detect VWD promptly, obtaining a thorough, detailed bleeding history is paramount.

PRACTICAL RECOMMENDATIONS FOR SURGEONS

- If intraoperative re-exploration fails to reveal an active source of bleeding — Consider von Willebrand Disease!
- Questions aimed at identifying prior bleeding episodes, including their character and circumstances, must be incorporated into every surgical patient's mandatory preoperative assessment questionnaire.
- When VWD is suspected, and before the disease type and specific targeted therapy are clarified, it is advisable to administer cryoprecipitate and antifibrinolytic agents promptly.

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ფონ ვილებრანდის არადიაგნოსტირებული დაავადებით გამოწვეული პოსტოპერაციული სისხლდენა. კლინიკური შემთხვევის აღწერა

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ლალი ახმეტელი, ლიანა საგინაშვილი

თბილისის სახელმწიფო სამედიცინო უნივერსიტეტის ქირურგიის N1 დეპარტამენტი

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რეზიუმე

პოსტოპერაციულ პერიოდში განვითარებულ სიცოცხლისთვის საშიშ გართულებათა შორის განსაკუთრებული ადგილი უკავია ფონ ვილებრანდის დაავადებით (VWD) გამოწვეულ სისხლდენებს. მათი მართვა განსაკუთრებით რთულდება იმ პაციენტებში, რომელთაც ოპერაციამდე საერთოდ არ იცოდნენ თუ იყვნენ დაავადებული ამ კოაგულოპათიით.

ფონ ვილებრანდის დაავადება მემკვიდრეობითი კოაგულოპათიაა, რომელიც ხასიათდება ფონ ვილებრანდის ფაქტორის (VWF) რაოდენობის, სტრუქტურის ან ფუნქციის დეფექტებით.

VWF წარმოადგენს გლიკოპროტეინს, რომელიც უზრუნველყოფს სისხლძარღვის დაზიანებული ენდოთელის ზედაპირზე თრომბოციტების პირველად ადჰეზიას, მათ შემდგომ აგრეგაციას და თრომბის წარმოქმნის პროცესს. ის ასევე უზრუნველყოფს სისხლის შედედების VIII ფაქტორის ტრანსპორტირებას და მისი დონის სტაბილიზაციას პლაზმაში.

დაავადების დროულ გამოვლენას ართულებს ის გარემოება, რომ ოპერაციის წინ ჩატარებული კოაგულოგრამის ისეთი სტანდარტული ტესტების მონაცემები როგორიცაა: პროთრომბინის დრო (PT), პროთრომბინული აქტივობა (PT%), საერთაშორისო ნორმალიზებული ფარდობა (INR), აქტივირებული პარციალური თრომბოპლასტინის დრო (APTT), თრომბინის დრო (TT), ფიბრინოგენის კონცენტრაცია (Fibrinogen) ხშირ შემთხვევაში ნორმის ფარგლებშია ან მისგან იმდენად უმნიშვნელოდ არის გადახრილი, რომ დაავადების დროული ამოცნობისთვის შესაფერ დასკვნის გაკეთების საშუალებას არ იძლევა.

წარმოდგენილი კლინიკური შემთხვევა შეეხება 77 წლის მამაკაცს, რომელსაც საზარდულის ჩაჭედილი ირიბი თიაქრის გამო გაუკეთდა სასწრაფო ღია ჰერნიოპლასტიკა ბადის გამოყენებით.

პოსტოპერაციულ პერიოდში ნაოპერაციებ მიდამოში ჩამოყალიბდა მზარდი კანქვეშა ჰემატომა. რის გამოც განმეორებით ჩატარდა ქირურგიული ჩარევა. ინტრაოპერაციული რევიზიით აქტიური სისხლდენის მიზეზი არ იქნა ნანახი.

ჰემატოლოგის მიერ ჩატარებული დამატებითი კვლევებით პაციენტს აღმოაჩნდა I ტიპის VWD და ჩატარებული ადეკვატური კონსერვატიული მკურნალობით მიღწეული იქნა ჰემოსტაზი.

საკვანძო სიტყვები: პოსტოპერაციული ჰემორაგია, ფონ ვილებრანდის დაავადება, საზარდულის თიაქარი