Case Report

Anesthetic Strategy for a Femur Fracture Surgery in a Patient with Hemophilia B (Factor IX Deficiency): A Case Report

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Abstract

Hemophilia is a hemorrhagic trend affects mostly males (X-related recessive disease). In 85% of cases it is caused by factor VIII deficiency, being called hemophilia A or classic hemophilia. In approximately 15% of cases there is factor IX deficiency (hemophilia B). The objective of this report was to describe the case of a patient with Hemophilia B underwent a femur fracture surgery. Male patient-18 years old, 81.7 kg, 183 cm, carrying hemophilia B, victim of a motorcycle traffic accident, was admitted in emergency room with Glasgow 15, excoriations in arms and thorax, and a femur fracture. He was first subjected to external fixation and posteriorly underwent surgery to correct a distal femur fracture. Hemophiliac patients' administration has made strides. Hemophilia ought not to be a contraindication for an obtrusive strategy; be that as it may, various conditions are required to give effective surgery and an uncomplicated and safe postoperative course.

Key words: Femur Fracture, Factor IX, Hemophilia B.

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Introduction

Hemophilia is a hemorrhagic trend affects mostly males (X-related recessive disease). In 85% of cases it is caused by factor VIII deficiency, being called hemophilia A or classic hemophilia. In approximately 15% of cases there is factor IX deficiency (hemophilia B).[1-3]

Hemophilia B is characterized into severe (<1%), moderate (1%–5%), or mild (5%–40%) phenotypes in light of the plasma component IX action of influenced people.[4] A variety of underlying mutations have been identified and linked with different levels of clinical severity.[4,5] The serious phenotype is portrayed by unconstrained and repetitive draining scenes into joints and muscles, with hemarthroses being the dominating reason for long haul disability.[5] The moderate phenotype is described by incidental unconstrained drains and delayed seeping with minor injury or surgery. At last, patients with the gentle phenotype once in a while exhibit unconstrained draining yet may have critical seeping with significant injury or surgery. Forceful component substitution is required fundamentally for patients with moderate and serious hemophilia B phenotypes.[1,4]

Hemophilia ought not to be a contraindication for an invasive procedure; be that as it may, various conditions are required to give fruitful surgery and an uncomplicated and safe postoperative course.[6]

In any capacity, when these patients are submitted to surgical procedures, it merits uncommon consideration and a multidisciplinary group of wellbeing experts educated about the sickness, including qualified hematologist, surgeon and anesthesiologist.[3,7,8]

The objective of this report was to describe the case of a patient with Hemophilia B underwent a femur fracture surgery.

Case Report

Male patient, 18 years old, weight 81.7 kilograms, 183 cm in height, carrier hemophilia B (factor IX deficiency). He was a victim of a motorcycle traffic accident, was admitted in emergency room with Glasgow 15, normal cardiac and pulmonary auscultation, excoriations in arms and thorax, and a femur fracture. Blood pressure of 157 x 83 mmHg and heart rate of 126 beats per minute. After damage controls, he had an elective surgery purpose to correct a distal femur fracture.

During pre-anesthetic evaluation, presented in good condition, hydrated, no fever and eupneic. Normal psychomotor development and no other comorbidities. Denied allergies, addictions and family and personal history negative for anesthetic complications. Evaluation Mallampati class I with good mouth opening and class II according to American Society of Anesthesiologists (ASA). Cardiac and pulmonary auscultation revealed normal limits.

Initial tests in emergency room showed hematocrit of 42.1%, hemoglobin of 14.2 g.dL-1, leukocytes of 14.900/mm3, 274.000/mm3 platelets, serum sodium of 147.1 mEq.L-1, serum potassium of 3.7 mEq.L-1, fasting blood glucose of 96 mg.dL-1, creatinine of 0.61 mg.dL-1, urea of 16 mg.dL-1, activation time

prothrombin of 100.0% (INR: 1.00), coagulation time 8 minutes, bleeding time 2 minutes 30 seconds and normal electrocardiogram.

On the third day of hospitalization factor IX was administered in a dose of 50 UI.kg-1 IV (4000UI) intravenous until fifth day. On the sixth day it was administered 7500UI (93.75 UI.kg-1) three hours before the surgery.

The patient was placed supine, monitored, and pre medicated with clonidine 75 mcg intravenous (IV). At this moment tranexamic acid 8.0g (97.9 mg.kg-1) was administered preemptively. After pre-oxygenation, anesthesia was induced with fentanyl 350 µg, IV propofol 150 mg IV and atracurium 40 mg IV. The others adjuvants administered were cefazolin 2g, dexametasone 10mg, ondansetron 8mg, haloperidol 5mg, morphine 8mg, dipyrone 2g and lidocaine 100mg.

There was no bradycardia events and low blood pressure were corrected with metaraminol 0,5mg IV as necessary.

Intubation was successful as Cormack-Lehane grade II without difficult. Anesthesia was maintained with sevoflurane and atracurium as an option for muscle relaxation.

The monitoring was maintained sinus rhythm, saturation between 98% and 100%, end tidal CO2 between 32 and 38 and mean arterial pressure between 73 and 90 throughout the period.

Diuresis total was 150 ml (0.93 ml.kg-1.h-1). The intraoperative blood gases and pH showed immediate postoperative between 7.34 and 7.38; PO2 between 98 and 121, bicarbonate between 22.6 and 26.7 and base excess between -3.2 and +2.5. None packed red blood cells and a bag of fresh plasma was necessary.

Total anesthesia time was 2 hours and 45 minutes and course proceeded with two episodes of intraoperative hypotension promptly treated with metaraminol 0,5mg in bolus. The patient was extubated taken at surgery and sent to the Intensive Care Unit.

Post-surgical bleeding was not significant and the patient had discharge from hospital in three days without major complications.

Table 1: Laboratory tests until in hospital days.

Day	Hematocrit %	Hemoglobin $g.dL^{-1}$	Platelets $/mm^3$	PT (seconds)	aPTT (seconds)
Admisson	42.0	14.2	274000	12	32
Third	40.2	13.9	271000	18	79
Fourth	41.1	13.9	266000	14	39.5
Fifth	41.1	13.6	258000	13	37.4
Sixth	42.1	13.8	256000	13	37.1

PT – prothrombin time
aPTT - Activated Partial Thromboplastin Time

Discussion

So far, there are few cases[9] of anesthetic procedures with hemophilia B published in the literature submitted to general anesthesia, since previous studies have described the management hemophilia A and C.[3,6,7,10]

Surgical treatment is a safe and reliable choice for addressing complications including hemophilia-related osteoarthropathy given the implementation of effective measures for treatment during the perioperative period.[7]

The single most important factor which contributes to the severity of hemophilia is the nature of mutations. Even the phenotypically severe hemophilia cases can further be subclassified based on the type of mutations they carry. Even both hemophilia A and B mutation databases show that less severe gene defects are more common in hemophilia B than in A.[11]

It is severe if coagulation factor VIII level is below 1%; it is moderate if it is between1% and 5%; and mild if it is above 5%.[1,3,5] Normal factor VIII plasma levels vary 0.5 U/mL to 1.5 U/mL and each U/mL corresponds to 100% factor VIII found in 1 mL of plasma. Laboratory diagnosis of hemophilia A is based on prolonged TTPa and factor VIII deficiency, normal factor IX and von Willebrand factor levels.[1,3]

The dose of recombinant coagulation factor IX was adjusted to sustain factor IX activity above 80%, as suggested by Makino.[9] The patient showed a favorable course without hemorrhagic tendency. We could safely manage anesthesia without requiring allogeneic blood transfusion. The goal in this stage is to correct factor IX deficiency before the procedure, which is hematologist's responsibility. It must be known that patients need 100% correction of their factor IX before any surgical procedure, and this must be confirmed before surgery.[3,8,10]

In Hemophilia A, checking ought to be accomplished by dosing variable VIII levels here and there a day. To the Hemophilia B, we can think similarly. In spite of the fact that not perfect, it is additionally conceivable to observing with serial TTPa estimation if serial element VIII or IX estimation is not accessible. Patients' coagulation will be viewed as satisfactory when the proportion between patients' TTPa and typical TTPa is equivalent to or underneath 1.2.[8]

Fresh frozen plasma is utilized subsequent to the 1950s to treat hemophilia. It contains 175 to 250 mL of volume for each sack with 70 to 90 U.dL-1 variable VIII, component IX, von Willebrand element and other coagulating elements. It can supplant 15% to 20% component VIII with a volume of 800 to 1000 mL. It might be utilized to

treat known coagulation element insufficiencies in situations when the particular concentrate is not accessible. Prothrombin concentrate complex is comprised of prothrombin concentrate, elements IX and X and variable element VIII sums. Utilized as a part of hemophilia A in patients with component VIII inhibitors, in the measurements of 75 to 100 U.kg-1. It is related to thromboembolic issues. Cryoprecipitate and Factor VIII concentrate are demonstrated to hemophilia A.[3,12,13] Desmopressin increases the level of Factor VIII and Factor von Willebrand. It is therefore useful in Hemophilia patients with mild to moderate (not useful in severe).[3,13] Not affect the level of factor IX , which is contraindicated in hemophilia B.[13]

The level of plasma IX factor increases approximately 1% per unit / kg of Factor IX infused. The recombinant factor IX is less replacement (0.8 adults and 0.7% in children). The half-life of factor IX is 18-24hs.[13,14]

Épsilon aminocaproic acid has a limited use because of low power and high toxicity. (13) Tranexamic acid is an adjuvant, mainly in factor XI deficient patients. Its use is indicated in minor bleeding of mucous during surgical and dental procedures. Use is contraindicated in patients with hematuria, severe chest surgery, or in patients haemophilia inhibitors (neutralizing antibodies factor).[13]

Recent data from a handful of patients who have undergone gene therapy for hemophilia B are very encouraging with a sustained factor IX level of 0.05 IU/mL maintained for over 4 years. Prophylaxis is also associated with high peak levels, which permits patients to maintain an active lifestyle.[15]

Factor replacement therapy may be provided either "on demand" for symptoms related to bleeding or as "prophylaxis" in which scheduled infusions are undertaken in an attempt to prevent hemorrhage. Primary prophylaxis refers to factor replacement that is started to prevent clinical bleeding episodes in the infant or young child, while secondary prophylaxis refers to replacement therapy that is initiated in response to recurrent bleeding symptoms. Prophylaxis has the potential to change the landscape in hemophilia B by reducing debilitating musculoskeletal complications in patients with severe hemophilia and improving quality of life. Current clinical research and development efforts are predominantly aimed at manipulating the pharmacokinetic and physiologic properties of factor IX to prolong the biological half-life and/or enhance in vivo hemostatic function. Alternative approaches seek to "rebalance" the coagulation response via long-acting agents.[1,16]

Hemophilia is associated with an increased risk of blood transfusion after lower extremity total joint arthroplasty. Same care is needed to the femur surgery. Patients and providers should discuss these risks before surgery.[17] Before induction of anesthesia in a patient with hemophilia B should be performed one factor measurement, which is expected to be above 60% (preferably more than 80%) for major surgery and 40% for minor procedures. In postoperative monitoring it´s desirable that factor IX is between 30–50%. In patients with hemophilia A the pre-operatory measurement must be preferably close to 100% (between 80-100%).[13]

Conclusion

Hemophilia ought not to be a contraindication for an obtrusive strategy; be that as it may, various conditions are required to give effective surgery and an uncomplicated and safe postoperative course.

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How to cite this article: Sakae TM. Anesthetic Strategy for a Femur Fracture Surgery in a Patient with Hemophilia B (Factor IX Deficiency): A Case Report. Acad. Anesthesiol. Int. 2016;1(1):6-9.

Source of Support: Nil, **Conflict of Interest:** None declared.