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Perioperative Challenges in a Case of Hemophilia Undergoing Lower Limb Surgery: A Case Report

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Abstract

Classic hemophilia is a rare X-linked recessive hereditary bleeding disorder which usually manifests as spontaneous bleeding or persistent bleeding even after trivial trauma. The risk of perioperative bleeding can be reduced by a thorough knowledge of the disease process and multidisciplinary team approach. We report management of a young male recently diagnosed with hemophilia A posted for surgery for fracture shaft of femur following trauma.

Keywords: Factor VIII, Hemophilia A, Spontaneous bleeding.

Introduction

Hemophilia A, although rare, is the most common inherited bleeding disorder. Historically, these patients were considered high risk and were frequently denied surgical procedures due to life threatening bleeding. Advances in medicine and with the development of Factor VIII (FVIII) concentrates major surgical procedures are now performed. [1] Anesthesia providers should be aware of the current treatments and recommendations in order to provide the best possible evidence based care.

Case report

A 20 year old 60kg male was brought to the emergency after trauma to left leg diagnosed as fracture shaft of femur. He gave history of spontaneous bleeding into joints which was first noticed by his parents at 5 years of age affecting mainly the knee joints following which he was diagnosed with hemophilia A. His treatment consisted of replacing FVIII level whenever he suffered from any episode of spontaneous bleeding. He was electively planned for open reduction and internal fixation with plating.

On preoperative examination his heart rate was 108/min, regular, blood pressure of 118/76 mm of Hg with normal heart sounds on auscultation. Airway examination revealed Mallampatti grade 1, inter-incisor distance more than 4cms, thyromental distance more than 6cms and no adventitious sounds on chest auscultation. His lab reports were

Hemoglobin (Hb) - 6.9 g%

Total Leukocyte Count (TLC) -10,000/cu mm

Platelet Count - 300 × 10^3/microL

Prothrombin Time (PT) - 12.8 s

International Normalized Ratio (INR) - 0.95

Bleeding Time (BT) -2 min 30 s

Clotting Time (CT) -15 min 10 s

Activated Partial thromboplastim Time (aPTT) - 47.2 s

FVIII level < 1%.

A hematological opinion was sought and was advised screening for FVIII inhibitors. After testing negative for FVIII inhibitors, FVIII level was corrected prior to surgery along with Hb level correction and was posted for surgery 3 days after the last transfusion. Hematologist orders included

D_0-D_3, 3000 units of FVIII BD, D_0 being the day of surgery (morning dose to be given before surgery)

D_4-D_6, 2500 units BD

D_7-D_{10}, 1500 units BD

Investigations in the evening prior to surgery were

Hb-14.5 g%

TLC- 5800/cu mm

Platelet count- 180 × 10^3/microL

PT -13.5 s

INR - 1

APTT - 48.80 s

FVIII level = 130%

On the day of surgery in the OR, on examination patient's pulse rate was 112/min, blood pressure was 110/78 mmHg and oxygen saturation of 100%. Monitoring was done using pulse oximetry, electrocardiogram and noninvasive blood pressure. An 18 G peripheral cannula was secured and general anesthesia was administered using intravenous (IV) Fentanyl 2mcg/kg and IV Propofol 2mg/kg. Neuro muscular blockade was done using IV Vecuronium 0.1mg/kg. Airway was secured a-traumatically with a 7.5mm ID cuffed pre lubricated endotracheal tube and intermittent positive pressure ventilation was established. Maintenance of anesthesia was done using 1% Sevoflurane in 50% each mixture of Oxygen and Nitrous Oxide and IV Vecuronium 1mg as needed. Pressure points were padded with cotton to prevent hematomas or hemarthroses. Surgery was performed under lower limb tourniquet to minimize blood loss. Intraoperatively one unit of packed Red Blood Cells (PRBC), two units of Fresh Frozen Plasma (FFP) and a crystalloid volume of 2500 ml were transfused. Analgesia was achieved with IV Paracetamol and IV Fentanyl.

The patient remained hemodynamically stable with no acute events. The total blood loss was estimated to be 550 ml. Normothermia was maintained throughout the surgery.

At the end of surgery that lasted for 2.5 hours neuromuscular blockade was reversed using IV Neostigmine and IV Glycopyrrolate after the standard criteria for extubation was met. Patient was transferred to recovery room after extubation.

Eight hours after the surgery patient received another dose of FVIII and was put on the regime as per hematologist. The post-operative period was uneventful. The patient was discharged on day 10 post operatively with an Hb level of 9.2 g%.

Discussion

Hemophilia A is an X linked recessive hereditary disorder affecting the male progeny in an incidence of 1/5000 male live births. It is characterized by deficiency of clotting FVIII. These patients usually present with spontaneous hemorrhage or uncontrollable bleeding into joints, muscles and vital organs even after a trivial trauma because of delayed clot formation and friable clot unable to maintain vascular integrity. The

severity of bleeding is inversely related to FVIII level in the body. The normal value of FVIII in plasma is 0.5-1.5 IU/ml or 50-150%. Patients with severe hemophilia have FVIII level below 1% of normal. Factor VIII activity in patients with mild, moderate and severe are 5 -<40%, 1-4% and less than 1%. Coagulation studies in these patients reveal a high activated partial thromboplastin time but normal platelet count, bleeding time and prothrombin time. Increased serum bilirubin may also be seen in case of large hematomas. [2] High morbidity and mortality is attributed to the risk of excessive bleeding either spontaneously or after trauma or any surgical procedure. Intracranial hemorrhage is the most feared complication in severe disease. [3]

For these patients undergoing any type of surgical intervention, a thorough pre-anesthetic evaluation is essential as they are at a risk of excessive bleeding regardless of the severity. Management of the factor replacement should ideally be done after consultation with a hematologist, as was done in the presented case. An inhibitor assay should be conducted in these patients. Inhibitors are antibodies that neutralize FVIII derivatives and can render the disease more difficult to manage. [4] As these patients receive frequent transfusions they should be screened for transfusion related infections. World Federation of Hemophilia (WFH) guidelines recommended FVIII level for hemophilia A patient undergoing minor surgery is 50-80% preoperatively and 80-100% preoperatively for major surgery. Perioperative administration of Factor replacement in hemophiliacs is pivotal for hemostasis during surgery. These levels are obtained by the following dose calculation

Required IU = body weight (kg) x desired FVIII rise (%)(IU/dL) x 0.5 (IU/kg per IU/dL). [5]

In the absence of inhibitors, plasma FVIII levels increase approximately 2IU/dL per infused IU/kg body weight. Factor VIII should be administered twice daily due to its half-life of 8-12 hours.

Intraoperative risk of bleeding can be minimized by avoiding the use of Succinylcholine as fasciculation can precipitate bleeding [3], maintaining adequate depth of anesthesia to avoid any kind of airway trauma [6], use of previously greased endotracheal tube to reduce friction with the mucosal surface and employment of a skilled anesthesiologist to perform endotracheal intubation. We employed tourniquet to minimize bleeding in this case. Additionally antifibrinolytics like Tranexamic acid or Epsilon amino-caproic acid can be used as an adjunct therapy to promote clot stability. [7] Post-operative pain modalities include the use of opioids, Acetaminophen or Cox-2 inhibitors, while the use of non-steroidal anti-inflammatory agents is discouraged due to risk of GI bleeding. [2] Prevention of multiple punctures for intravenous or arterial access can be done by early use of an ultrasound. Intramuscular medications should be avoided. A thorough risk benefit analysis should be done before proceeding with regional anesthesia as guidelines do not recommend regional anesthesia. Sub arachnoid block using a small gauge spinal needle may be preferred. [8] In case of anticipated difficult airway neuraxial anesthesia can be considered after adequate FVIII levels in the body has been attained.

Even for minor surgical procedures patient should be taken to a center with adequate blood bank support to provide sufficient quantities of FVIII concentrates and blood testing facilities. [9]

Conclusion

The major goal is to prevent excess bleeding. Anesthetic considerations must be individualized to the

patient. A careful preoperative evaluation, stating the risk of hemorrhage, screening for other comorbidities and a multidisciplinary approach for management of these patients is very beneficial in deciding the postoperative outcome.

References

1. Franco JA, Cabot MH. Haematology and Anesthesia. In: Nagelhout JJ, Plaus KL, eds Nurse Anesthesia. 5th ed. St. Louis, MO: Elsevier Saunders 2014; 880-899.
2. Mallya P, Kaimar P, Jithesh R, Ranjan RK, Ambareesha M. Anaesthetic management of a patient with haemophilia. Indian J Anaesth 2007;51:145-47.
3. Mistry T, Dogra N, Chauhan K, Shahani J. Perioperative considerations in a patient with haemophilia A: A case report and review of literature. Anesth Essays Res 2017;11:243-5.
4. Eckhardt CL, van der Bom JG, van der Naald M, et al. Surgery and inhibitor development in haemophilia A: A systematic review. J Thromb haemost, 2011;9:1948-58.
5. Hopkins J. Anesthetic considerations for the patient with hemophilia A. CRNA Today, 2018;3:3-7
6. Sethi M, Gurha P. Perioperative management of a patient with haemophilia-A for major abdominal surgery. Indian Journal of Anaesthesia, 2017;61:354-55.
7. Madan N, Rathnam A, Bajaj N, Treatment of an intraoral bleeding in haemophilic patient with a thermoplastic palatal stent- A novel approach. Int J Crit Illn Inj Sci 2011;1:79-83.
8. Abramovitz S, Beilin Y Thrombocytopenia, low molecular weight heparin, obstetric anaesthesia. Anesth Clin N Am 2003;21:99-109.
9. Gyanesh P, Dhiraaj S. Anesthetic management of a patient with haemophilia A with spontaneous acute subdural hematoma. J Anesthesiol Clin Pharmacol 2013;29:117-20.