CASE REPORT

Bali Journal of Anesthesiology (*BJOA*) 2017, Volume 1, Number 2: 29-30
E-ISSN: 2549-2276

Published by DiscoverSys

Perioperative management of patient with hemophilia a underwent orthopedic surgery

Tjokorda GA Senapathi,[1] I Made Gede Widnyana,[2]
I Gusti Ngurah Mahaalit Aribawa,[3] Christopher Ryalino[4*]

ABSTRACT

Hemophilia presents challenging consideration for anesthesiologists. In non emergency cases it is essential that factor VIII is raised to its optimal activity prior to surgery. Intra operative bleeding is a fatal complication in hemophilia. Certain measurements must be taken under considerations to manage this case for anesthesia. Peripheral lines should be secured with utmost care. Intramuscular injections and arterial punctures must be avoided. Intubation and airway positioning should be done gently in deep plane anesthesia. Small vessels hemostatis must be taken care of by the surgeon. A multidisciplinary team has to be involved when patients with hemophilia are planned for surgery. The knowledge related to replacement therapy should be mastered not only by hematologist, but also by the whole team involved in patient management.

Keywords: hemophilia, elective surgery, factor VIII, cryoprecipitate
Cite This Article: Senapathi, T.GA., Widnyana, I.M.G., Aribawa, I.G.N.M., Ryalino, C. 2017. Perioperative management of patient with hemophilia a underwent orthopedic surgery. *Bali Journal of Anesthesiology* 1(2): 29-30. DOI:10.15562/bjoa.v1i2.10

[1,2,3]Senior Lecturer, [4]Resident Department of Anesthesiology and Intensive Care, Sanglah General Hospital, Bali

INTRODUCTION

Hemophilia A is a hemorrhagic trend almost exclusively affecting males (X-related recessive disease). In 85% of cases, it is caused by factor VIII deficiency, called hemophilia A or classic hemophilia.[1] Diagnosis of hemophilia is usually suspected when bleeding symptoms occur spontaneously or after trauma.[2] Preoperative preparations, careful intubations and invasive tools administration, and the readiness of blood factor transfusions may play an important role in managing patients with hemophilia in anesthesia.

CASE REPORT

An 11 years old boy, 30 kg, presented with a lump on his left fifth finger since eight months prior to admission. Previously about one month before, he was playing soccer and when he fell to the ground the finger was stepped by his friend. The lump was getting bigger in size, but with no such pain in the area. The finger's joint movement is limited. He was diagnosed with pseudo cyst and planned for open biopsy with frozen section.

During lab exams for surgery, his APTT level was unmeasurable, despite normal PT and bleeding time. Upon consultation to pediatrician, he was found to have history of bruises in multiple occasions, sometimes without any history of trauma. A similar history in the family could not be evaluated since he was adopted. He was diagnosed with Hemophilia A and planned for cryoprecipitate transfusion and Koate-DVI, an intravenous anti-hemophilic factor. Prior to surgery, his hemoglobin level was 11.9 g/dL, and his hemostasis factors were PT 11,9 (13,1), APTT 61,2 (34,1), and INR 1,03.

He was planned for general inhalational anesthesia with ETT insertion. He was pre-medicated with Midazolam 1 mg. Medications used were Propofol 50 mg, Fentanyl 50 mcg, and Atracurium 15 mg. Intubation was conducted carefully and gently, avoiding excessive trauma to the airway. He was maintained under anesthesia by oxygen, nitrous oxide, and sevoflurane. Cryoprecipitate and PRCs were ready to use and available on-site.

Surgery was uneventful, lasted for 120 minutes. Pseudo cyst was removed. Bleeding was 100 mL, no blood components were used during surgery. Post-surgery he was transferred to intermediate ward, and received another dose of Koate-DVI. Continuous drip of fentanyl and IV paracetamol were prescribed for analgetics. He was discharged from hospital 5 days after the surgery.

DISCUSSION

Hemophilia A, a recessive X-linked disorder involving lack of functional clotting factor VIII (FVIII), represents 80% of hemophilia cases. Severe cases (<2% functional factor VIII) have spontaneous bleeding. Moderate (2-10% functional factor VIII) and mild (>10%) lead to excessive bleeding only after trauma or surgery. Patients have high APTT but platelet count, bleeding time and prothrombin time are normal. Factor assay is diagnostic.[3]

A multidisciplinary team has to be involved when patients with hemophilia are planned for surgery.[4] The knowledge related to replacement therapy should be mastered not only by

*Correspondence to: Christopher Ryalino, Department of Anesthesiology and Intensive Care, Sanglah General Hospital, Bali
ryalino@gmail.com

hematologist, but also by the whole team involved in patient management.[5]

In preparing patients with hemophilia A for surgery, factor VIII levels are routinely raised to approach 100% of normal activity. It should be maintained for the first 3 postoperative days, from the day 4 onwards it should be maintained at 80%, from 7th day onwards it is allowed to decline to 40% of normal activity. The formula used to calculate the factor VIII dose is:

$$N = \text{plasma volume (ml/kg)} \times \text{weight (kg)} \times \text{Percent activity increase}$$

where N is the number of units required. Plasma volume is 40 ml/kg for adults. Since half-life of factor VIII is about 12 hours, it must be administered twice daily. Cryoprecipitate is next choice of blood product in the management of hemophilia A, which provides 80 units of factor VIII per bag.[6]

All the patients with hemophilia, regardless of its severity, are at risk of excessive bleeding during and after surgery. Intramuscular premedication should be avoided.[1,6] Vascular access itself does not cause excessive bleeding and should be appropriate for the proposed procedure.[1] Avoid arterial puncture whenever possible.

Extra care should be taken in intubation it can cause submucosal hemorrhages, which can turn the situation to a life-threatening one. Nasal intubation should be avoided as it can prove traumatic and bleeding from the site can lead to aspiration.[1,6] Intubation should be done in deep plane of anesthesia.[7] Care should be taken during positioning of the extremities, and pressure points should be padded to prevent intramuscular hematomas or hemarthrosis.[1,5,6,7]

Hypertension and tachycardia during surgery in hemophiliacs can lead to increased surgical bleeding. Controlled hypotension techniques prevent hemostasis of small vessels but are not recommended. Hemodynamic conditions should be maintained near normal.[1] Suctioning the airway must be gentle to avoid unnecessary trauma to the airway and oral cavity. Use of both NSAID and steroids for should be avoided due to its potential to cause gastrointestinal bleeding.

Surgeon should give special attention to small vessel hemostasis, rather than trusting on hemostatic physiological mechanisms. The patient was taken up for surgery at the earliest after the APTT was normalized or optimized. The availability of sufficient quantities of clotting factor concentrates was ensured before undertaking the surgery.[1]

Postoperative PICU or intermediate room is mandatory to monitor the signs and symptoms of intracranial hemorrhage. Postoperative monitoring for bleeding was done with Hb and AT levels along with factor VIII assay.[1,7]

CONFLICT OF INTEREST

None to declare.

FINANCIAL SUPPORT AND SPONSORSHIP

None

REFERENCES

1. Khokhar RS, Hussain A, Khan MU, Hajnour M, Qureshi S, Aqil M. Anesthetic management of patient with hemophilia a undergoing emergency ventriculoperitoneal shunting: A case report and review of literature. Saudi Journal of Anaesthesia. 2016;10(4):474-476. doi: 10.4103/1658-354X.179120.
2. Shah UJ, Narayana M, Smith JG. Anaesthetic considerations in patients with inherited disorders of coagulation. Continuing Education in Anesthesia, Critical Care & Pain. 2014;5(1):26-31.
3. Gyanesh P, Dhiraaj S. Anesthetic management of a patient with hemophilia A with spontaneous acute subdural hematoma. Journal of Anaesthesiology, Clinical Pharmacology. 2013;29(1):117-120. doi: 10.4103/0970-9185.105819.
4. Flores RP, Bagatini A, Santos AT, Gomes CR, Fernandes MS, Molon RP. Hemophilia and anesthesia. Rev Bras Anestesiol. 2004 Dec; 54(6):865-71.
5. Shrestha GS, Poudyal B, Bhattarai AS, Shrestha PS, Sedain G, Acharya N. Perioperative management of two cases of hemophilia with spontaneous intracerebral hemorrhage undergoing emergency craniotomy in resource constrained setup of Nepal. Indian Journal of Critical Care Medicine: Peer-reviewed, Official Publication of Indian Society of Critical Care Medicine. 2014;18(11):754-756. doi: 10.4103/0972-5229.144023.
6. Mallya P, Kaimar P, Jithesh P, Ranjan RK, Ambareesha M. Anaesthetic Management of a Patient with Haemophilia. Indian Journal of Anaesthesia. 2007; 51(2):145-7.
7. Kumar P, Banerjee A, Bansal T, Pirkad A, Govil V. Hemophilia A: an anesthetic challenge. Anaesth Pain & Intensive Care 2016;20(2):221-223