Perioperative management of patient with hemophilia a underwent orthopedic surgery

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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 hemostasis 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.

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 spontaneous 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 premediated 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
hematologist, but also by the whole team involved in patient management. 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.

All the patients with hemophilia, regardless of its severity, are at risk of excessive bleeding during and after surgery. Intramuscular premedication should be avoided. Vascular access itself does not cause excessive bleeding and should be appropriate for the proposed procedure. Avoid arterial puncture. 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. Intubation should be done in deep plane of anesthesia. Care should be taken during positioning of the extremities, and pressure points should be padded to prevent intramuscular hematomas or hemarthrosis.

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. 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 hematostatic 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.

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.

**CONFLICT OF INTEREST**

None to declare.

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None

**REFERENCES**


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