

Anesthetic Management of a Video-Assisted Thoracoscopy and Pulmonary Biopsy in a Hemophilic Patient. a Case Report

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Abstract

Hemophilia A is an X-linked recessive hereditary bleeding illness that manifests as increased bleeding after moderate trauma and spontaneous bleeding. It is caused by a deficiency of the clotting factor VIII (FVIII). We report a video assisted-thoracoscopy in a 48-year-old patient with Hemophilia A. Hemophilia shouldn't be a barrier to invasive surgery, but there are a few conditions that must be met to ensure a straightforward intraoperative and postoperative course. Understanding the disease, a multidisciplinary team approach, and appropriate anesthetic management can result in a successful surgical outcome.

Key words: Activated factor VIII; anesthesia; bleeding; coagulopathy; hemophilia; thoracic surgical procedure

Introduction

Hemophilia is an inherited X chromosome-linked bleeding disorder, predominantly in males, with an incidence of 5000 to 7000 males. Female are carriers and male-to-male transmission does not occur [1]. Factor VIII deficiency, often known as hemophilia A, accounts for 85 to 90% of all cases of the disease and is marked by bleeding into the joints and soft tissues [1,2]. For patients with hemophilia, intracranial bleeding is the leading cause of death [3].

Factor VIII replacement, preferably using recombinant factor concentrates, is the mainstay of hemophilia A treatment. The severity of the condition determines the dosage, frequency, and length of therapy. Patients typically receive preoperative dosages of factor concentrate to elevate plasma concentrations to hemostatic levels prior to surgical procedures. In order to prevent bleeding, these levels should be kept up in the postoperative phase by repeated dosages [1].

The risk of uncontrollable bleeding due to the defect in the intrinsic pathway of the coagulation system makes perioperative management very

challenging. It is worth noting that even after a period of apparent hemostasis, there is a risk of rebleeding or late postoperative bleeding [1]. The purpose of this report is to highlight the anesthetic management of a hemophilic patient and the prevention of perioperative coagulopathic bleeding.

Case Report

A-48-year-old male patient with hemophilia A and arterial hypertension, diagnosed with interstitial lung disease was electively scheduled for a video thoracoscopy with pulmonary biopsy under general anesthesia. The patient regularly took prednisone for the lung disease and Olmesartan for gastritis.

A perioperative plan was to maintain 100% factor VIII concentration and, for that, 3000 unit of factor VIII concentrate was transfused in bolus an hour before the procedure. At preoperative evaluation, previous laboratory values of prothrombin time, activated thromboplastin time, hemoglobin, liver and kidney values were within of the baseline values.

The administration of Factor VIII was performed according to the table below:

Day	Desired factor elevation	number of units of factor VIII
D1	100% 1h before the surgery	3.000 IU 1h before de surgery and 8h after surgery
D2	50% 8/8h	1.500 IU FVIII 8/8h
D3	50% 12/12h	1.500 IU FVIII 12/12h
D4	50% 12/12h	1.500 IU FVIII 12/12h
D5	50% 24/24h	1.500 IU FVIII 24/24h
D6	50% 24/24h	1.500 IU FVIII 24/24h
D7	50% 24/24h	1.500 IU FVIII 24/24h
D8	50% 24/24h	1.500 IU FVIII 24/24h
D9	50% 24/24h	1.500 IU FVIII 24/24h
D10	50% 24/24h	1.500 IU FVIII 24/24h
D11	Reassessment of hematology regarding the need to maintain factor VIII	

Table 1: Number of factor VIII units to be administered according to the desired elevation

(Prepared by the authors according to the protocol of Hospital das Clínicas UFMG)

The baseline heart was 92bpm, blood pressure 130/90mmHg, satO₂ 94%. In the operating room, a standard monitor was attached including ECG, pulse oximeter and noninvasive blood pressure.

An 18-gauge canula was inserted into a vein on the dorsum of the left hand. Infusion of Ringer lactate solution was initiated and hydrocortisone 100mg was administered. Anesthesia was gently induced with sufentanil 0,3µg/kg, lidocaine 1mg/kg, propofol 2,5mg/kg. After confirmation of loss of consciousness, rocuronium 0,6mg/kg was administered intravenously and oral intubation was performed with a 7.5mm cuffed endotracheal tube. Anesthesia was maintained with sevoflurane and bone pressure points were wrapped with cotton during positioning on the operating table to avoid intramuscular hematoma or hemartosis. Methadone 10mg was given for analgesia.

Hemodynamic and other vital parameters were stable and the duration of the surgery was 120min. Estimated blood loss during the procedure was minimal. The patient was extubated after reversing the neuromuscular block with Sugammadex 4mg/kg, as the use of neostigmine and atropine can cause tachycardia that increases the risk of bleeding at the surgical site. The patient was extubated after recovery of neuromuscular blockade and adequate spontaneous breathing.

The patient was admitted into the recovery room without any complications. Factor VIII concentrate was administered postoperatively to maintain the required concentration of at least 50% of normal.

Discussion

The severity of the clinical manifestation of hemophilia A, one of the most common inherited clotting factor deficiencies, depends heavily on the baseline factor VIII level [4]. Depending on the level of factor VIII activity, hemophilia is classified as mild, moderate, or severe. The severity of the condition is typically correlated with the expression of the phenotype. While moderate forms only experience bleeding in reaction to trauma or surgery, if not adequately treated, severe forms exhibit profuse bleeding that is either spontaneous or caused by trauma [4].

Patients may undergo surgical treatments for various conditions such as hernia repair, cholecystectomy, gastrointestinal and thoracic operations, or for hemophilia-specific conditions including arthropathies [1].

It is necessary to treat the hemostatic deficiency with a specialized replacement therapy. Desmopressin acetate 1- desamino-8-D-arginine vasopressin (DDAVP), with or without adjuvant antifibrinolytic medication, can be used to treat mild forms. [4].

Plasma factor VIII levels will rise by around 2% for each unit of factor VIII clotting activity per kilogram of body weight. Factor VIII can be given twice daily or as a continuous infusion because its half-life is 8–12 hours. The

length of clotting factor replacement after surgery, however, and the precise factor amount needed for hemostasis are uncertain [4]

According to Yehia, one of the ways to calculate factor VIII replacement before surgery is through the following formula: FVIII dose (IU/kg) = desired increase of FVIII /2 to maintain a FVIII level of 70 IU/dl. This level is considered based on the type of the surgery [1]

Antifibrinolytic drugs, such as tranexamic acid, can be administered intravenously prior to inducing anesthesia because they improve hemostasis by enhancing clot stability [4].

Cryoprecipitate can also be used as it contains 80 units of factor VIII activity per bag. Fresh Frozen Plasma contains all plasma proteins including factor VIII and may be used in absence of the specific clotting factor concentrate [5].

Restricted fluid therapy is desirable in order not to dilute the plasma coagulation factor concentration [5].

To deliver the hemostatic drugs, a trustworthy venous access must be established. Central venous access may be required and should be obtained prior to surgery in some cases (small children, problematic venous access). Arterial cannulation should wait until the missing clotting factor is replaced. The use of depolarizing neuromuscular blockers should be avoided because muscle fasciculation can make bleeding worse [4]

Intubation should be performed gently, avoiding mucosal trauma, and insertion of temperature probes and pharyngeal aspiration should be performed carefully because trauma of tongue and airway muscle can rapidly lead to profuse bleeding and airway obstruction. During the procedure, hypertension and tachycardia can result in increased bleeding in the surgical area. Tracheal extubating needs to be performed in deep plane of anesthetic avoiding cough reflex [3].

For postoperative analgesia, non-steroidal anti-inflammatory drugs should be avoided due to their anti-platelet effects, predisposing to a worsening of complications such as gastrointestinal bleeding. Some authors suggest that neuraxial blocks as well as peripheral nerve blocks may be performed safely in a hemophilic patient if the level of factor VIII is maintained throughout the perioperative period [3]

Conclusion

In conclusion, during the perioperative context, patients with inherited bleeding disorders are at higher risk. Surgery in this population of patients is still difficult, but with careful preoperative preparation by a knowledgeable hematology team and adequate anesthesia management, most operations should be successful.

Declaration of interest

The authors declare no conflicts of interest.

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