CASE REPORT

Hemophilia A: an anesthetic challenge

Prashant Kumar[1], Arnab Banerjee[2], Teena Bansal[3], Arjun Pirkad[2], Vasudha Govil[2]

[1]Professor; [2]Postgraduate Resident; [3]Assistant Professor
Department of Anesthesiology & Critical Care, Pandit Bhagwat Dayal Sharma Post Graduate Institute of Medical Sciences (PGIMS), Rohtak, Haryana 124001, (India)

Correspondence: Dr. Arnab Banerjee, Postgraduate Resident, Department of Anesthesiology PT.B.D Sharma PGIMS, Medical Rd, Maharshi Dayanand University, Rohtak, Haryana 124001, (India); Email: dr.arnab77@gmail.com

ABSTRACT

Hemophilia A is a congenital bleeding disorder with low factor VIII levels in the blood. We report the anesthetic management of a hemophilic patient presenting with trauma in the left forearm. Patient received Factor VIII perioperatively with no excessive blood loss intraoperatively. We stress that adequate preoperative preparation and a planned anesthesia leads to a successful management of hemophilic patients.

Key words: Hemophilia A; Hemophilia A, Congenital: Hemophilia, Classic; Factor VIII; Deficiency, Factor VIII

Citation: Kumar P, Banerjee A, Bansal T, Pirkad A, Govil V. Hemophilia A: an anesthetic challenge. Anaesth Pain & Intensive Care 2016;20(2):221-223

Received: 16 January 2016; **Reviewed:** 16 February, 3 March 2016; **Corrected:** 29 February, 3 June 2016; **Accepted:** 10 June 2016

INTRODUCTION

Hemophilia A patients with factor VIII (FVIII) level <1%, may have spontaneous bleeding (in muscles and joints), but patients with level between 1% to < 40% present with massive bleeding only after trauma or surgery.[1] Platelet count, bleeding time and prothrombin time are normal but APTT remains high. FVIII assay is diagnostic. Sometimes bleeding can be life threatening and requires immediate management.[2] We report the anesthetic management of a hemophilic patient presenting with trauma to the left forearm.

CASE REPORT

A 23 year old male weighing 50 kg presented to the emergency department with a history of trauma to his left forearm. Past history revealed acute subdural hemorrhage (SDH) in the left fronto-temporal region with a history of joint swelling 2 years back, for which he was operated under general anesthesia. He had received 10 units of factor VIII perioperatively as he was diagnosed to be suffering from hemophilia A. No other significant positive history. On clinical examination there was a swelling at the left forearm. Ultrasonography of the forearm revealed edema of skin and subcutaneous tissues along with a collection of 4.7×2.8 cm at lateral aspect of left forearm with thickening. As he was a known case of hemophilia A peripheral lines were secured with utmost care and blood samples for routine lab investigations, FVIII assay and blood grouping and cross-matching sent. Two units of fresh frozen plasma (FFP) and one unit of cryoprecipitate, were transfused immediately. Routine lab investigations were normal with APTT 95%, INR 1.17, FVIII assay was 3%, FVIII inhibitor screening was negative. Hematology opinion was done and they advised to keep the FVIII level at 45 to 50% to prevent intraoperative bleeding. Four units of FVIII (4 units = 1000 IU) were transfused over 2 days. A level of 28.80% of FVIII was achieved. Another 4 units (1000 IU) were transfused over next 2 days and a level of 42.50% was achieved. Two units of cryoprecipitate and 2 units of FVIII (500 IU) were transfused for two consecutive days before surgery and FVIII was found to be 44.50% the day before surgery.

He was explained about the procedure and written informed consent was obtained before surgery. Preoperatively 250 IU were infused. Patient was shifted to the operation room with utmost care and paddings were done on the pressure points. Standard monitors were attached. Intravenous line with 18 G cannula was secured and concurrent administration of Ringer's lactate solution was initiated. General anesthesia was induced with glycopyrrolate, thiopentone, fentanyl and atracurium. Trachea was intubated and anesthesia was maintained with isoflurane 0.75% in nitrous oxide and oxygen in ratio of 67:33. FVIII 250 IU and two FFPs

were transfused intraoperatively. Intraoperative blood loss was 250 ml. Patient was reversed successfully and was shifted to the recovery room where 250 IU of FVIII were transfused. It was advised to transfuse 750 IU of FVIII 12 hourly for the next 3 consecutive days (FVIII 40%), then 500 IU of FVIII 12 hourly for the next 5 days (FVIII 20-30%). He was discharged on 8th postop day and followed up after one week with a FVIII level of 10%.

DISCUSSION

Our patient was a known hemophilic who was successfully operated for acute SDH two years back, and now presented to us with fracture of both bones of his forearm. Management of hemophiliacs needs FVIII assays, FVIII, blood products as well as a well-coordinated team effort. The recommended plasma factor level and duration of administration for major surgery are shown in Table 1.[3]

Each FVIII unit per kilogram of body weight raises the plasma FVIII level by approximately 2%, (half-life of 8-12 hours) and should be transfused slowly (<3 ml/min).[4] Cryoprecipitate (30-40 ml) provide 80 units FVIII, while one ml of FFP contains one unit of FVIII activity.[5] Desmopressin and tranexamic acid are useful as adjunctive therapy in these patients. In hemophiliac patients with inhibitors use of Factor VIIa has been documented.[6]

When our patient presented to the emergency department, peripheral line was secured with utmost care and blood products (FFP and cryoprecipitate) were transfused immediately to prevent further bleeding in forearm. Initially FVIII level was 3% for which patient received

Table 1: Suggested plasma factor peak level and duration of administration

Type of hemorrhage		No significant resource constraint		Resource constraint	
		Desired level (IU/dl)	Duration (days)	Desired level (IU/dl)	Duration (days)
Joint		40-60	1–2, may be longer if response is inadequate	10-20	1–2, may be longer if response is inadequate
Superficial muscle/no NV compromise (except iliopsoas)		40-60	2–3, sometimes longer if response is inadequate	10-20	2–3, sometimes longer if response is inadequate
Iliopsoas & deep muscle with NV injury /substantial blood loss	Initial	80-100	1-2	20-40	
	Maintenance	30-60	3–5, sometimes longer as secondary prophylaxis during physiotherapy	10-20	3–5, sometimes longer as secondary prophylaxis during physiotherapy
CNS/head	Initial	80-100	1-7	50-80	1-3
	Maintenance	50	8-21	30-40 20-40	4-7 8-14
Throat and neck	Initial	80-100	1-7	30-50	1-3
	Maintenance	50	8-14	10-20	4-7
Gastrointestinal	Initial	80-100	7-14	30-50	1-3
	Maintenance	50		10-20	4-7
Renal		50	3-5	20-40	3-5
Deep laceration		50	5-7	20-40	5-7
Surgery (major)	Pre-op	80-100		60-80	
	Post-op	60-80 40-60 30-50	1-3 4-6 7-14	30-40 20-30 10-20	1-3 4-6 7-14
Surgery (minor)	Pre-op	50-80		40-80	
	Post-op	30–80	1-5, depending on type of procedure	20-50	1-5, depending on type of procedure

eight units of FVIII over 4 days and FVIII level increased to 42.50%. Target FVIII level was 45 to 50% before surgery. Patient received one unit of cryoprecipitate and two units of FVIII (500 IU) for two consecutive days before surgery and FVIII level assay achieved was 44.50% preoperatively. In our patient FVIII inhibitor assay was also done which was found to be negative.[5]

In our case following precautions were taken perioperatively:

1. Peripheral lines secured with utmost care.
2. Intramuscular injections and arterial punctures were avoided.
3. Smooth induction under deep plane of anesthesia.
4. To prevent muscle fasciculation (aggravate muscle and joint hematomas), use of succinylcholine was avoided.
5. Tracheal intubation was atraumatic.
6. Gentle suctioning of oropharynx was done.
7. Extremities and pressure points were padded.
8. Non-steroidal analgesics were not used as it causes gastrointestinal hemorrhage.
9. Hypothermia was prevented.
10. Hemodynamic conditions were maintained near normal.
11. Small vessel hemostasis was achieved with minimal intraoperative blood loss
12. FVIII concentrates were continued both intraoperatively as well as in the postoperative period.
13. Availability of blood products and FVIII concentrates were ensured before surgery.
14. FVIII assays were done postoperatively to achieve the desired level of FVIII.[7,8]

To conclude, our patient having hemophilia A, presenting to us for orthopedic surgery, with trauma in the forearm, was managed successfully. This case emphasizes the importance of elaborate history taking, careful physical examination, coordination between an anesthesiologist, surgeon and hematologist for planning bleeding prophylaxis, and a planned anesthesia (induction-maintenance-extubation sequence), for a successful management of hemophilic patients.

Conflict of interest: Nil

Authors' contribution: All the authors took part in the conduct of case and manuscript preparation.

REFERENCES

1. White GC 2nd, Rosendaal F, Aledort LM, Lusher JM, Rothschild C, Ingerslev J. Definitions in hemophilia. Recommendation of the scientific subcommittee on factor VIII and factor IX of the scientific and standardization committee of the International Society on Thrombosis and Haemostasis. Thromb Haemost 2001;85(3):560. [PubMed]
2. Aronstam A, Rainsford SG, Painter MJ. Patterns of bleeding in adolescents with severe haemophilia A. Br Med J 1979;1(6161):469-70. [PubMed] [Free full text]
3. Rickard KA. Guidelines for therapy and optimal dosages of coagulation factors for treatment of bleeding and surgery in haemophilia. Haemophilia 1995;1(S1):8–13. doi: 10.1111/j.1365-2516.1995.tb00104.x. [PubMed]
4. Björkman S, Berntorp E. Pharmacokinetics of coagulation factors: clinical relevance for patients with haemophilia. Clin Pharmacokinet 2001;40(11):815-32. [PubMed]
5. Stanworth SJ. The evidence-based use of FFP and cryoprecipitate for abnormalities of coagulation tests and clinical coagulopathy. Hematology Am Soc Hematol Educ Program 2007:179-86. [PubMed]
6. Roberts HR, Monroe DM, White GC. The use of recombinant factor VIIa in the treatment of bleeding disorders. Blood 2004;104:3858-64. [PubMed] [Free full text]
7. Mallya P, Kaimar P, Jithesh R, Ranjan RK, Ambareesha M. Anesthetic management of a patient with haemophilia. Indian J Anesth. 2007;51:145-7.
8. Flores RP, Bagatini A, dos Santos AT, Gomes CR, Fernandes MS, Molon RP. Hemophilia and Anesthesia. Rev Bras Anestesiol 2004;54:865-71. [PubMed] [Free full text]

★★★★★