International Journal of Medical Research Professionals
P-ISSN: 2454-6356; E-ISSN: 2454-6364
DOI: 10.21276/ijmrp

IJMRP

Case Report .

Anaesthetic Management of a Case of Hemophilia: A Case Report and Literature Review

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ABSTRACT

Hemophilia A also known as classic hemophilia is a congenital hereditary bleeding disorder. It can present with variable symptoms along a spectrum between spontaneous bleeding and persistent bleeding after minor trauma or surgery. It is necessary to have understanding about the disease pathophysiology and perioperative risk involved for the safe management in perioperative period. We report a case of an adult with hemophilia A presented with fracture shaft of femur after trauma for femur nailing.

Keywords: Hemophilia, Bleeding Disorder.

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Article History:

Received: 01-06-2018, **Revised:** 27-06-2018, **Accepted:** 24-07-2018

Access this article online	
Website: www.ijmrp.com	Quick Response code
DOI: 10.21276/ijmrp.2018.4.4.047	

INTRODUCTION

Hemophilia A is also known as classic hemophilia. It is a congenital bleeding disorder which usually affects males. Its hereditary transmission is through X linked recessive trait and is characterized as deficient or defected clotting factor 8.[1] The incidence is approximately 1 per 5000 males.[2] Individual may present with internal or external bleeding episodes. Symptoms range from mild to severe forms. Individuals with severe form suffer from more severe and frequent bleeding episodes while mild hemophiliacs typically suffer more minor symptoms except after surgery or serious trauma. Bleeding may occur anywhere in the body, superficial bleeding such as those caused by abrasions, may be prolonged. Due to the lack of fibrin, scab may easily be broken up, which may cause re-bleeding. Joints, muscles, brain are some of the more serious sites of bleeding. A minor trauma can lead to a life threatening hemorrhage in hemophilia patients. These patients need a multidisciplinary team approach and systemic as well as local trauma site care for a favorable outcome. Early intervention and timely factor VIII replacement can save from life threatening complications.

CASE PRESENTATION

A 22 yr male brought in emergency with history of fall from stairs. A fracture involving SOF was diagnosed. The patient was a diagnosed case of hemophilia A since childhood. His Factor 8 essay came to be 19.1% with aPTT of 68s vWF antigen assay was 223.76 and Ristocetin cofactor activity assay was 199.5. His inhibitor titre was found to be negative. He had no previous history of bleeding joints and other comorbidities. On examination blood pressure was 110/86 with pulse rate of 94. Hematologist prescribed factor VIII concentrate 2250 IU, 12hrly as per the weight of patient. Femur nailing was planned for fixing the fracture under general anaesthesia. Preoperative aPTT 33 and platelet count of 5 lac observed. In the operative room iv line secured with 18 G cannula and monitor attached. Patient was induced with 2mg/kg propofol and 2mcg/kg of fentanyl. Airway secured with ETT no. 8 cuffed using videolaryngoscope, minimizing airway trauma, with vecuronium. Intraoperatively, maintained with O2 and N2O and desfluorane. Infusion of tranaxemic acid started. Vital parameters were maintained throughout procedure. Surgery lasted for one and half hours. Intraoperative blood loss was minimal. After surgery patient was extubated and shifted to PACU. Factor 8 continued for 3 days post op. Post op period was uneventful.

DISCUSSION

Hemophilia A is caused by deficiency of coagulation factor VIII while hemophilia B is caused by deficiency of coagulation factor IX. The severity of bleeding is related to level of clotting factor.[3] Normal levels of factor VIII are 0.5 to 1.5 IU/ml or 50-150%[4] (1 IU/ml = 100% of F8 in 1 ml of normal plasma). Level of clotting factor and degree of severity is shown in table 1.

Most spontaneous bleeds occurs internally, into the joints or muscles. However, major cause of mortality is intracranial hemorrhage.[5]

Table 1: Level of clotting factor and degree of severity

Severity	Clotting factor level	Bleeding episode
Mild	**5- <40% of normal or 5-40 IU/dl**	Spontaneous bleeding- rare Severe bleed- only after major surgery or trauma
Moderate	**1-5% of normal or 1-5 IU/dl**	spontaneous bleeding-occasional; whereas prolonged bleeding- with minor surgery or trauma
Severe	**<1% of normal or <1 IU/dl**	Spontaneous bleeding into joints or muscles, predominantly in the absence of identifiable hemostatic challenge

Table 2: Suggested Plasma Factor Peak Level and Duration of Administration.[10]

Surgery		No significant resource constraint		significant resource constraint	
		Desired level (IU/dl)	Duration (Days)	Desired level (IU/dl)	Duration (Days)
Major	**Pre-op**	80-100	-	60-80	-
	Post-op	60-80	1-3	30-40	1-3
		40-60	4-6	20-30	4-6
		30-50	7-14	10-20	7-14
Minor	**Pre-op**	50-80	-	40-80	-
	Post-op	30-80	1-5	20-50	1-5

Trauma poses a significant risk even in mild form of the disease. Immediate intervention is required in these patients to ensure hemostasis. Reliable clotting factor monitoring and inhibitor assay should be done. Factor VIII deficiency is recommended to be replaced with recombinant or viral inactivated plasma-derived concentrates.[6,7] The dose of factor VIII is calculated by 0.5 × weight (kg) × desired factor level in IU/dl, according to the world federation of hemophilia 2012 (WHF) guidelines.[3] Duration and desired factor level of clotting factor depends on the type of surgery. Maximum rate of transfusion should not be more than 3ml/min in adults.[8] Plasma FVIII level will raise by approximately 2 IU/dl per unit of transfused FVIII/kg of body weight, in the absence of an inhibitor.[9]

Other pharmacological agents that can be of great value include desmopressin, tranexamic acid and epsilon aminocaproic acid.

Desmopressin (1-deamino-8-D-arginine vasopressin, DDAVP) is a synthetic vasopressin analogue. DDAVP has been used in patients with measureable baseline levels of plasma FV111 and low inhibitor levels with favourable outcome.[11] Loomes et al reported that DDAVP administrations in moderate hemophilia A, to be adequate for treatment in case of minor bleeding or trauma.[12]

Tranexamic acid and ε-aminocaproic acid are useful as an adjunct therapy in hemophiliacs as it promotes clot stability by their antifibrinolytic activity.[13] Perioperative management of the patient with hemophilia should be carefully planned. Intramuscular injections should be avoided. Other measures like, gentle manipulation of the airway to prevent submucosal haemorrhages and padding of the pressure points to prevent intramuscular haematomas or haemarthrosis, should be taken care of.

To conclude we successfully managed a patient with hemophilia with fracture femur for nailing according to WHF guidelines.

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Source of Support: Nil.

Conflict of Interest: None Declared.

Cite this article as: Sumit Soni, Balbir, Anudeep. Anaesthetic Management of a Case of Hemophilia: A Case Report and Literature Review. Int J Med Res Prof. 2018 July; 4(4):209-10. DOI:10.21276/ijmrp.2018.4.4.047