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Case Report: Management of Hemophilia-A for B/L TKR

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Abstract

This report describes successful management of 36 yrs old patient with hemophilia-A, who received general anaesthesia for B/L TKR. He was given Anti-Haemophilic Factor (AHF)-VIII starting before the surgery and continued for till his full recovery.

Haemophilia A is an X-linked recessive hereditary disorder characterized by a deficient or defective factor VIII. Haemophilia-A is more likely to occur in males than in females as males have only one X-chromosome. Thus females are almost exclusively asymptomatic carriers of the disorder. Before the commercial availability of Factor VIII, surgery was impossible in haemophilia patients and even tooth extraction could be life threatening.

Keywords: *Haemophilia, AHF VIII, General Anaesthesia.*

Introduction

Hemophilia is the inherited x-linked bleeding disorder. This is because of low or deficient Anti-hemophilic Factor VIII, popularly known as hemophilia-A or Classic hemophilia and factor IX deficiency known as hemophilia-B or Christmas disease. Factor VIII level in normal individuals range from .6 to 1.5 IU/ml but in hemophilic patients these levels are less than .5 IU/ml or absent. Haemophilia is characterized by spontaneous bleeding into the joints. This is also a clinical in type-3 Von Willebr and disease and in the severe forms of other coagulation factor deficiencies. The joints most frequently involved are knees, ankles and elbows. Recurrent bleeding into the joints causes chronic synovitis and

destruction of articular cartilage and subchondrial bone. This condition is called as chronic hemophilic arthropathy, causes pain, stiffness and deformity leading to severe impairment of function. Now as the clotting factor concentrates are available, major surgical procedure like bilateral total knee replacement can be performed safely for correction of the severely involved joints. This is a case presentation of bilateral total knee replacement done in the patient of severe hemophilia-A at Indian Spinal Injuries Centre, New Delhi.

Case Report

A 36yrs old male, wt.-80kg, complaining of pain B/L knee for last 22 yrs. Pt. was a known case of severe Hemophilia-A and was admitted for B/L total knee replacement in our institute.

Since childhood patient had frequent episodes of gum bleeding, red & blue patches on body (which were painful on touch). He had frequent episodes of pain in both knees which get relieved by analgesics & anti-inflammatory and it used to take about 7-10 days to resolve. At the age of 14 years, his left knee developed unbearable pain and was very swollen. Patient was shown in a tertiary centre where he was diagnosed as a case of severe Hemophilia-A.

Soon his knees movements become restricted and over next few years both knees became fixed. Though patient's other joints were normal except complain of ankle pain. Patient had history of infrequent episodes of malena. He used to take AHF VIII 1000-1500 IU himself after bleeding. Patient had no any other medical problem except Hemophilia.

Patient has two brothers and two sisters. They do not have such problem. Patient's maternal uncle died of uncontrolled bleeding in an accident- ? Hemophilia. Patient's two maternal cousins are diagnosed patients of Hemophilia-A.

Plan of Anaesthesia and Surgery

Presence of antibodies to AHF VIII was ruled out. Consultation from Hematologist was sought. Assurance from laboratory was taken for perioperative AHF VIII assay. Patient was admitted two days before scheduled date of surgery. Patient was counselled before surgery and consent for surgery, anaesthesia and post-operative analgesia was taken. Adequate blood and blood products were arranged. A chart was made for peri-operative AHF VIII infusion. APTT was 98. Hb-14.4, PCV-44.1 rest routine investigations were within normal limit. HIV and Hepatitis-C ruled out. Night before surgery AHF VIII 4000 IU infused and APTT came to 37.2. On the day of surgery AHF VIII 4000 IU infused and AHF VIII assay done. AHF VIII was 120%, APTT 28/30, PT 13/13, INR 1.0 and vitals were WNL. Two wide bore cannula were inserted. General Anaesthesia was administered as per standard protocol. Surgery was done under tourniquet application. During surgery (8 hours after previous dose) AHF VIII 4000 IU was infused. Induction, maintenance and recovery from anaesthesia were uneventful.

Chart for AHF VIII Infusion.

Date	Time	Dose	Expected AHF VIII Assay
Night before surgery	9.00PM	4000 IU	100%
Day of surgery	9.00 AM 9.00 PM	4000 IU 4000 IU	100% 100%
Next 2 days post op.	9.00 AM 9.00 PM	4000 IU 4000 IU	100% 100%
Next 3 days post op.	9.00 AM 9.00 PM	2500 IU 2500 IU	62.5% 62.5%
Next 3 days post op.	9.00 AM 9.00 PM	1500 IU 1500 IU	37.5% 37.5%
Next 4-6 wks post op.	For Physio.	750 IU	19.0%

Total blood loss till 2 hrs post surgery was about 750 ml. One unit blood was transfused. Vitals were stable and patient was shifted to ICU for overnight observation. For post-operative analgesia injection fentanyl 75 mcg/hr was started in ICU. Patient was comfortable and vitals remained stable post-operatively.

Post Operatively

On night of POD -0: Hb 11.9, PCV 36.4, APTT 38/32, PT12.6 and INR 1.0

POD -1: Rt.drain-10ml, Lt. drain-100ml. Hb 11.2, PCV 33.8, APTT 37/32, PT 12.7 INR 1.02.

POD -2: Rt. drain-50ml, Lt. drain-150ml. APTT, PT, and INR were WNL. AHF VIII assay-102%.

For DVT prophylaxis, no medication was given. However IPC (Intermittent Pneumatic Compression) device was applied.

On 3rd POD, drains were nil and APTT, PT and INR were WNL.

On 4th POD, drains were removed. From 5th POD, physiotherapy was started. Patient was discharged on 7th POD with proper advice.

Two weeks after surgery patient himself reduced the dose and took only 250 IU. But during physiotherapy he developed pain and swelling in left knee, so he increased the dose to 750 IU. Now patient is comfortable and has resumed his routine work.

Discussion

Management of haemophilic arthropathy is difficult and needs multi-speciality approach. For good outcome patient should ideally be seen by orthopaedic surgeon, haematologist, physiotherapist and anaesthetist before scheduled

surgery. Educating the pt. is crucial to a successful outcome. Once the joint replacement surgery has been decided, the following points should be addressed:

- Counselling of patient regarding surgery, hemophilia treatment, possible complications like post operative bleeding and management should be done.
- Possible duration of hospital stay (usually 4-14 days) and expected cost involved should be discussed. Frequent factor replacement is essential to minimize the bleeding and to promote the healing following TKR. Adequate factor supply must be secured before surgery.
- Pt. should be willing to receive whole blood, blood products and concentrates required for haemostasis.
- Patient should be motivated to complete the required physiotherapy following surgery and outcome should be discussed.
- Lab. testing including AHF VIII inhibitor, APTT, PT, INR, HBsAg, HCB, HIV status should be before surgery. If pt. is HIV positive, the CD4 count should be >200.
- Pain management should be discussed in detail before surgery.
- Discharge planning should be made prior to surgery.

Medical Consideration

Having been exposed to contaminated blood products many people with hemophilia have chronic HIV or hepatitis-C infections. Pre-operative careening should be done to determine the current immune status in HIV positive cases.

If pt. is on interferon and ribavirin therapy for hepatitis-C, the surgery should be delayed until the side effects of the treatment disappeared.

Besides routine investigations the following tests are done-

- AHF VIII inhibitor status
- HIV antibody, viral load and CD4count.
- HCV and viral load.
- Fibrinogen, APTT, PT INR & Platelet count.
- Cardiopulmonary status.

Successful joint replacement surgery can be done with plasma derived or recombinant AHF VIII and possibly with cryoprecipitate/fresh frozen plasma for pt. with hemophilia-A.

Laboratory capable of performing factor activity levels should be easily accessible for post-op factor VIII assay. (In our case we tied-up with such laboratory near to our institute. Only after taking assurance from that lab for timely factor VIII assay, we took the case).

If factor activity levels are not available within a reasonable turnaround time (<24hrs), then the APTT should be measured immediately after an infusion of factor. APTT gives rough guide to adequate treatment. Factor activity levels should be above 80% in post-operatively.

Anesthetic Considerations

Considering the risk of epidural bleed and haematoma formation the SA/CSE is contraindicated. General Anaesthesia is the choice of anaesthesia in hemophilic pts.

DVT Prophylaxis

There are varying opinions over DVT prophylaxis in the patients with the hemophilia undergoing joint replacement surgeries. Some doctors prefer injectable DVT prophylaxis, others prefer stockings or intermittent pneumatic compression device. According to some people, there is no need of DVT prophylaxis.

Anti-Hemophilic Factor VIII

AHF VIII is available in two forms:

- Purified Freeze-Dried Human Coag. Factor VIII.
- Recombinant Factor rVIII

Purified Freeze-Dried Human Coag. Factor VIII is cheaper than rVIII but there is rare risk of transmission of disease.

AHF VIII is indicated for Hemophillia A.

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