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Anaesthetic management of a patient with Haemophilia-A posted for cystolithotomy under general anaesthesia

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Conflicts of Interest: Nil

Abstract: Haemophilia A is an X-linked recessive hemorrhagic disorder characterized by the deficiency of factor VIII or antihaemophilic factor. We report 19 year old male patient with haemophila A, who received general anesthesia for exploration of multiple bladder calculi. In this case report we briefly reviewed the literature on haemophilia and challenges faced in the management of haemophilia.

Keywords: Haemophilia A, bleeding disorder, general anaesthesia

Introduction

Haemophilia A is an X-linked recessive hemorrhagic disorder characterized by the deficiency of factor VIII or antihaemophilic factor. Haemophilia A has a wide spectrum of manifestations ranging from persistent bleeding after minor trauma to spontaneous haemorrhage [1]. Prior to the advances in various fields of medicine and blood banking surgery was almost impossible in haemophiliac patients, even simple procedure like tooth extraction was life threatening due to uncontrolled bleeding. But recently development of factor VIII has reduced the morbidity and mortality amongst the haemophilia patients. Successful anesthetic management depends on the special care and a multidisciplinary team of health professionals informed about the disease, including qualified hematologist, surgeon, and anesthesiologist.

Case report

We report a 19 year old male patient with haemophila A, who received general anaesthesia for exploration of multiple bladder calculi. Patient complaints of lower abdominal pain since 4-5 years investigated and diagnosed as multiple urinary bladder calculi on USG abdomen and IVP. Past history revealed that patient was known case of hemophilia A since 15 years. At the age of 4 years he had history of fall and trauma to knee followed by uncontrolled bleeding which was not responding to conventional treatment. He was then investigated for haemophilia A and found to have reduced factor VIII levels. Family history of the patient revealed his first degree relative suffering from haemophilia A. There was no other significant history of any medical or surgical disorder or exposure to anaesthesia. On examination patient was moderately built, height 170 cm, weight 50 kg, heart rate was 84/min regular, blood pressure was 120/80 mm of Hg, the findings of the general and systemic examination were normal. Airway examination showed adequate mouth opening with mallampatti grading 1, neck movements were normal. Preoperative investigation revealed haemoglobin of 11 gm%, prothrombin time was 15/12, INR 1.2, APTT 180 seconds. Factor VIII concentration was found to be about 1%. The patient was scheduled for exploration bladder calculi under general anaesthesia. Preoperatively hematologist was consulted for the expert opinion of factor VIII infusion perioperatively. An 18 gauge intravenous cannula was inserted in left upper limb. As per haematologists advice 2500 units of factor VIII were given half an hour prior to surgery intravenously. On table premedication was provided with inj. Glycopyrrolate 0.2

mg IV, inj. Midazolam 1 mg IV, inj. Fentanyl 100mcg IV. Following pre-oxygenation for 3 minutes, anaesthesia was induced with inj. Profolol 2 mg/kg and Suxamethonium 1.5mg/kg was used to facilitate the insertion of 9.0 mm portex cuffed oral endotracheal tube. Anaesthesia was maintained using Sevoflurane as inhalational agent, inj. Vecuronium 6mg as muscle relaxant and 50:50 O_2:N_2O. Reversal of residual neuromuscular blockade was achieved satisfactorily with inj. Glycopyrrolate 0.4 mg and inj. Neostigmine 2.5 mg IV. Patient extubated when spontaneous ventilation established. After extubation of the trachea the patient was given oxygen by facemask for 10 minutes in the operation theatre. The entire procedure lasted 1 hour, all vital parameters measured throuought the procedure, estimated blood loss during surgery was minimal about 30 ml. Postoperatively analgesia was provided by injection Tramadol 100 mg IV twice a day. Postoperatively the patient was given factor VIII 750 IU after 12 hours while on day 2nd 750 IU every 6 hourly, on the third postoperative day the patient's factor VIII activity was 40%. On the seventh day postoperatively the patients Hb was 10.7 gm%. The patient was discharged on the seventh post operative day.

Discussion

Hemophilia is an X-linked recessive hemorrhagic disease due to mutations in the *F8* gene (hemophilia A or classic hemophilia) or *F9* gene (hemophilia B). Hemophilia A is the most common hereditary disorder associated with serious bleeding. Male subjects are clinically affected; women, who carry a single mutated gene, are generally asymptomatic. The intrinsic limb of the coagulation system is impaired hence haemostasis depends upon vascular and extrinsic mechanisms. It has a wide spectrum of manifestations ranging from persistent bleeding after minor trauma to spontaneous haemorrhage [1]. In all symptomatic cases there is a tendency toward easy bruising and massive hemorrhage after trauma or operative procedures. In addition, "spontaneous" hemorrhages are frequently encountered in regions of the body that are normally subject to trauma, particularly the joints, where recurrent bleeds into the joints (*hemarthroses*) lead to progressive deformities that can be crippling. Clinically hemophilia A and hemophilia B are indistinguishable, the diagnosis is made after specific determination of FVIII or FIX clotting activity. The disease phenotype correlates with the residual activity of FVIII or FIX and can be classified as severe (< 1%), moderate (1–5%), or mild (6–30%). Typically, the global tests of coagulation show only an isolated prolongation of the aPTT assay. Patients with hemophilia have normal bleeding times and platelet counts. Generally mild disease has normal life expectancy without any therapy, while without treatment patients with severe hemophilia have a limited life expectancy. Factor replacement therapy for hemophilia can be provided either in response to a bleeding episode or as a prophylactic treatment. Primary prophylaxis is defined as a strategy for maintaining the missing clotting factor at levels ~1% or higher on a regular basis in order to prevent bleeds especially the onset of hemarthroses. One unit of FVIII per kilogram of body weight increases the plasma FVIII level by 2%. The FVIII half-life of 8–12 h requires injections twice a day to maintain therapeutic levels. One can calculate the dose needed to increase FVIII levels using the simple formula below.

FVIII dose (IU) = Target FVIII levels – baseline FVIII levels x body weight (kg) x 0.5 unit/kg.

Also cryoprecipitate is enriched with FVIII protein (each bag contains ~80 IU of FVIII) and was commonly used for the treatment of hemophilia A decades ago it is still in use in some developing countries but because of the risk of bloodborne diseases, this product should be avoided in

hemophilia patients when factor concentrates are available. Another treatment modalities include DDAVP and antifibrinolytics drugs like EACA, tranexemic acid. DDAVP is a synthetic vasopressin analogue that causes a transient rise in FVIII and von Willebrand factor (vWF), but not Factor IX, through a mechanism involving release from endothelial cells. Patients with moderate or mild hemophilia A should be tested to determine if they respond to DDAVP before a therapeutic application. DDAVP at doses of 0.3 µg/kg body weight infused over a 20-min period is expected to raise FVIII levels by two- to threefold over baseline. DDAVP does not improve FVIII levels in severe hemophilia a patients, as there are no stores to release.

Bleeding in the gums and during oral surgery requires the use of oral antifibrinolytic drugs such as EACA or tranexamic acid to control local hemostasis. Tranexamic acid is given at doses of 25 mg/kg three to four times a day. Intramuscular injection was avoided due to risk of bleeding and hematoma. Care should be taken during positioning of the extremities, and pressure points should be padded to prevent intramuscular hematomas or hemarthrosis [2]. Post operatively analgesics such as aspirin and other NSAIDs were avoided as it can predispose to gastrointestinal hemorrhage. Patient controlled analgesia is also a safe and effective alternative to intramuscular injections [3]. Our decision to proceed with general anesthesia was based on a risk-benefit analysis, weighing the risk of neuraxial bleeding with a regional anesthetic versus benefits of general anaesthesia [4].

Conclusion

Considering the risk of neuraxial bleeding in haemophiliac patients, general anaesthesia can be safely used than regional anaesthesia with close monitoring of factor VIII level perioperatively.

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