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Case Report on Hemophilia

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ABSTRACT

Hemophilia is a bleeding disorder that results from congenital deficiency in a plasma coagulation protein. Hemophilia A is due to deficiency of factor VIII, whereas hemophilia B is due to factor IX deficiency. Hemophilia A & B are recessive X-linked diseases. In general, the disease affects only males, while females are carriers. This is a case of 86 year old male patient admitted in a medical ward with complaints of fever associated with chills and headache, burning micturition, heamaturia since 15-20 days, patient also complaints of vomiting occasionally 4-5 times a day and generalized weakness since 1 month. Patient is a known case of hypertension since 5 years and on medication which is combination of amlodipine and atenolol. Patient is also a known case of haemophilia A diagnosed in childhood. Patient has a history of fall 10 years back since then he can't walk. Patient was recently diagnosed with type 2 diabetes, perisplenic abscess, urinary tract infection, bilateral knee effusion from respective lab data. During the course of treatment, physicians and other health care professionals were advised to avoid use of NSAIDs, aspirin, IM injections, cannulas and suggested the physician regarding the use of cryoprecipitates when necessary and to monitor factors level. During the course of treatment, physicians and other health care professionals were also informed about medication error in the prescription and advised to use suitable drugs for the management of anemia. Patient was referred to other hospital for further evaluation. The main goal in the treatment of hemophilia is to control and prevent bleeding episodes. Treatment response can be monitored through clinical parameters, such as cessation of bleeding and resolution of symptoms.

Key Words: Hemophilia, Bleeding, Cryoprecipitate, congenital deficiency, Factor VIII, Factor IX deficiency.

INTRODUCTION

Hemophilia is a bleeding disorder that results from congenital deficiency in a plasma coagulation protein. Hemophilia A is due to deficiency of factor VIII, whereas hemophilia B is due to factor IX deficiency. Hemophilia A & B is recessive X-linked diseases. Hemophilia A (acquired haemophilia) is a severe bleeding diathesis due to impairment in the function of coagulation factor VIII caused by autoantibodies. These autoantibodies are defined as factor VIII inhibitor or circulating anticoagulant. [1] The incidence of haemophilia A is seen in approximately 1in 5000 male births, whereas Hemophilia B occurs less commonly, with only 1/4th the incidence of haemophilia A. There are no significant racial differences in the incidence of hemophilia. Family history of haemophilia is negative in 30% of severe haemophilia patients, representing a spontaneous mutation. Hemophilia A and hemophilia B are recessive X-linked diseases; the defective gene is located on the X chromosome. Therefore, the disease affects only males, while females are carriers. In Affected male's abnormal allele is present on their X chromosome and no matching allele on their Y chromosome. Thus their daughters would be obligatory carriers and their sons would be normal (assuming the mother is not a carrier). Female carriers have 1 normal allele so they do not usually have a bleeding tendency. Sons of female carrier and a normal male

have 50% chance of being haemophiliac, whereas daughters have 50% chance of being carriers. Therefore there is a "skipped generation" mode of inheritance in which the female carriers, who are the children of haemophiliacs do not express the disease but can pass it on to the next male generation. In 1984 researchers cloned and isolated the human factor VIII gene. More than 800 unique mutations in the factor VIII gene were reported, including point mutations, deletions, and insertions, have been reported (http://europium. csc.mrc.ac.uk). Deletions and nonsense mutations are often associated with severe forms of factor VIII deficiency, because no functional factor VIII is produced. In 1993 researchers identified that 45% of severe hemophilia A gene abnormalities was caused by an inversion in the factor VIII gene at intron. This discovery has simplified carrier detection and prenatal diagnosis for families with this gene mutation. The characteristic bleeding manifestations of hemophilia include excessive bleeding after trauma or surgery, hemarthroses, muscle hemorrhages, ecchymosis. The severity of clinical bleeding generally depends on the degree of deficiency of factor VIII or factor IX. [2]

CASE PRESENTATION

This is the case of 86 years old male patient admitted in a south Indian teaching hospital located in Bengaluru, the patient came with the chief complaints of fever on and off associated with chills & headache, burning micturition, generalised weakness since 1 month and complaints of vomiting occasionally that is 4-5 times/day. Patient also complained of haematuria since 15 to 20 days. Patient has a past history of Haemophilia A which was diagnosed in childhood. Patient has a history of fall 10years back since then patient is unable to walk. Patient also presents with history of Hypertension since 5 years and is on regular medication. Patient family history showed that the Male offspring is affected. During general physical examination patient was found to have bilateral knee joint swelling and presented with Pallor positive. Patient was conscious, co-operative, well oriented to time, place and person. Complete blood test, coagulation profile, patient sugar levels and urinary examination was performed. Patients RBC, Haemoglobin, haematocrit, MCV were found to be low and the coagulation profile showed that APTT level was near to the lower limit and the sugar & HbA1c levels were high. Due to the symptoms of hematuria Urine examination was performed and many pus cells and RBCs were seen (TABLE 1). Vitals of the patient are as follows Pulse: 72bpm, BP: 150/90 mmHg, Temperature: febrile, Abdomen pelvic scan report shows that patient has Cholelithiasis, Dilated CBD with IHBR dilation, Multiple hyperechoic lesions in the spleen. Hence From the subjective & objective evidence it was diagnosed as the patient is suffering from "Haemophilia with perisplenic abscess with UTI with Type 2 Diabetes Mellitus".

TABLE 1

TEST	OBSERVED RANGE
RBC	3.5×10^6/mm³
HGB	9.9 gm/dl
HCT	27.2%
MCV	77µm³
COAGULATION PROFILE:	
APTT	26.5 seconds
INR	1.1
PT	12.5 seconds
SUGAR LEVELS:	
FSG	126 mg/dl
RSG	247 mg/dl
PPBG	286 mg/dl
HBA1C	7.7%
URINE EXAMINATION:	
PUS CELLS	4.5/HPF
RBC	POSITIVE
MALARIAL PARASITE: NEGATIVE	

Patient was hospitalised for 8 days. Patient was treated with Regular insulin for first 3 days later changed to oral hypoglycemic agent (Glimepiride 1mg+ metformin 500mg) combination orally twice daily for type 2 Diabetic mellitus. Inj. tranexamic acid 5ml (100mg/ml) was administered SOS to reduce hemorrhage. Amlodipine & atenolol combination (50mg+5mg) orally once daily was given for treatment of hypertension. Nitrofurantoin (100mg) orally

twice daily and syrup Di sodium hydrogen citrate was given to treat UTI and anti emetic Inj. Ondansetron (4mg) was administered SOS to stop vomiting. Antibiotics such as cefoperazone (1gm) + sulbactam (500mg) were given to treat perisplenic abscess. Paracetamol was given SOS to treat fever. Patient was on IV fluids throughout the hospitalised days. (TABLE 2)

TABLE 2

Drugs	Dose	Route	frequency	1	2	3	4	5	6	7	8
IVF Normal saline Ringer lactate	1 pint 1pint	IV	@ 75ml/hr	+	+	+	+	+	+	+	+ +
Ranitidine	50mg	IV	1-0-1	+	+	+	+	+	+	+	+
Paracetamol	650mg	PO	SOS	+	+	+	+	+	+	+	+
Inj. H.Actrapid	Acc to sliding scale	SC		+	+	+	-	-	-	-	-
Inj. Tranexamic acid	1amp	IV	SOS	+	+	+	+	+	+	+	+
Tab Amlodipine + Atenolol	50mg+5mg	PO	1-0-0	-	+	+	+	+	+	+	+
Tab Nitrofurantoin	100mg	PO	1-0-1	-	-	+	+	+	+	+	-
Syp. Di sodium hydrogen citrate	2tsp	PO	1-1-1	-	-	+	+	+	-	-	-
Tab Glimepiride + Metformin	1mg+500mg	PO	1-0-1	-	-	+	+	+	+	+	+
Tab Voglibose	0.2mg	PO	1-0-0	-	-	-	+	+	+	+	+
Syp. Potassium Chloride	10ml	PO	OD	-	-	-	-	+	+	+	+
Inj ondansetron	4mg	IV	SOS	-	-	-	-	-	+	+	-
Inj. Cefoperazone+ sulbactam	1gm+500mg	IV	1-0-1	-	-	-	-	-	+	+	+
Tab Domperidone	10mg	PO	1-0-1	-	-	-	-	-	+	+	-
Tab Artesunate	100mg	PO	1-0-1	-	-	-	-	-	-	+	+
Tab ondansetron	4mg	PO	1-1-1	-	-	-	-	-	-	-	+

INTERVENTION

During the course of treatment, physicians and other health care professionals were advised to avoid use of NSAIDs and aspirin as it interferes with the stickiness of blood platelets and adds to the problem of bleeding. Physicians were advised to avoid IM injections, cannulas because of possibility of causing muscle bleed and suggested the physician regarding the use of cryoprecipitates when necessary and to monitor factors level and advised about the dosage to be administered which must be based on patients body weight (Factor VIII 1IU/kg= 2% rise in factor VIII activity, Example: 50IU= 100% correction). [3] During the course of treatment, physicians and other health care professionals were also informed about medication error that is Tab. Artesunate was prescribed without indication and physicians were advised to use fresh frozen plasma for the management of anemia.

CONCLUSION

Patient was referred to other hospital for further evaluation. The main goal in the treatment of hemophilia is to control and prevent bleeding episodes. Treatment response can be monitored through clinical parameters, such as cessation of bleeding and resolution of symptoms. Use of cryoprecipitates plays an important role in treatment of haemophilia. Clinical trials have demonstrated that recombinant factor VIII products are comparable in effectiveness to the plasma-derived products. The risk of developing an inhibitory antibody to factor VIII in patients with severe haemophilia A with the use of recombinant factor VIII is 28% to 33%. This risk is higher than that reported with plasma-derived products. [2] As this Prescription had medication error as a clinical pharmacist we recommend Medication reconciliation including an indication review for each prescription as it plays an important role in patient safety. Medication review, Patient education and poor communication between health care providers are the main factors responsible for medication error. [4]

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