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Case Report

Anaesthetic Management of a Patient with Congenital Factor VII Deficiency Undergoing Spine Surgery for a Tethered Cord: A Case Report

Faizah Mufti*, Bashir Ahmad Dar, Majid Khan, Shehla Bashir

Department of Anaesthesiology, Sher E Kashmir Institute of Medical Sciences, Jammu and Kashmir, India

ABSTRACT

Background: Congenital factor VII deficiency is a rare bleeding disorder that can lead to excessive bleeding during the intra-operative period and sometimes also during the postoperative period, which can lead to overall increase in morbidity and mortality. These patients may present with different clinical scenarios which may vary from being asymptomatic to bleeding that can be mild to life threatening. The recombinant form of activated factor VII (rFVIIa, NovoSeven®) has been used in the management of hemorrhage in the patients with factor VII deficiency.

Case: Patient's factor VII levels were determined. We administered recombinant factor VIIa, two hours prior to surgery. Repeat sample for coagulation profile was taken 1 hour after administration. Post operatively dose was to be repeated 4 hourly until adequate hemostasis was achieved. Throughout the procedure patient was thoroughly monitored and surgery was completed uneventfully in 2 hours and 15 mins.

Results: Our patient's factor assay showed a severe factor VII deficiency (0.3% of normal). However, the surgery was performed successfully without any bleeding or thrombogenic complications.

Conclusion: Patients with congenital FVII deficiency who require surgical intervention can be effectively managed with rFVIIa. Our patient received the dose pre operatively prior to intervention and was well tolerated and helped in achieving effective hemostasis both during surgery and in the post-operative period.

Keywords: Congenital FVII deficiency; Bleeding disorders; Recombinant activated FVII

INTRODUCTION

Congenital factor VII deficiency is a coagulation disorder with autosomal recessive inheritance and prevalence rate of about 1:5,00,000. Factor VII (FVII), which is a vitamin K-dependent coagulation factor, plays a role in initiation of the extrinsic coagulation pathway with tissue factor [1]. The severity of clinical manifestations in congenital FVII deficiency patients may vary from asymptomatic to mild bleeding as in bruising and epistaxis, to severe life-threatening bleeding such as intracranial hemorrhage [2]. Factor VII deficiency can be suspected when a coagulation screening test reveals an isolated prolongation of Prothrombin Time (PT) with a normal activated Partial Thromboplastin Time (aPTT) [1]. Deficiency of factor VII results in isolated prolongation of the Prothrombin Time (PT) or increased International Normalized Ratio (INR). The activated Partial Thromboplastin Time (aPTT) usually remains unaltered as well as thrombo-elastography [3]. Diagnosis is usually made incidentally where patient usually show decrease in factor VII levels without having any liver disease or without any history of intake of drugs or substances inhibiting factor VII [1].

Managements of patient with factor VII deficiency varies according to the clinical presentation of the patient which can vary from no intervention to administration of recombinant activated factor VII. The most commonly used treatment in these patients for prevention of spontaneous or surgical bleeding is administration of recombinant factor VII. However, there is still no optimal therapy for the patients with congenital factor VII deficiency and only few reports that have been reported could guide the peri-operative use of recombinant factor VII [4,5].

Correspondence to: Dr Faizah Mufti. Sher E, Department of Anaesthesiology, Kashmir Institute of Medical Sciences, Jammu and Kashmir, India, E-mail: arshiamufti@gmail.com

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CASE PRESENTATION

A 9 month old male child, weighing 8 kgs, 2nd in birth order, full term baby born *via* caesarean section with normal perinatal and developmental history and fully immunized was admitted in neurosurgery department of our hospital as a case of filum terminal lipoma with tethered cord. There was not any significant past medical history or family history of the child and no historical evidence of any bleeding disorder. Preoperative laboratory tests showed all the base line investigations within normal limits (Hb-11.7, TLC-9.8, platelet-268; urea-26, creatinine-0.14, Bilirubin-0.43, ALT 28, ALP-421, Albumin-4.6), however the coagulation profile was altered, showing PT/INR-deranged, aPTT-32. Abdominal ultrasound showed unremarkable study. His liver function tests were all normal and patient had no documented history of any liver disorder. Thinking of some lab error, repeat coagulation profile was sent which again showed deranged coagulation profile. Another sample was sent to outside laboratory for confirmation which showed PT-39, INR-2.8, aPTT-33.

Following this, a suspicion of coagulation disorder was made and case was discussed with pediatricand hematology departments which advised for assessment of factor assays. Factor assay showed a severe factor VII deficiency (0.3% of normal).After ruling out other possibilities and that the patient was not on any medication that would inhibit factor VII activity, and decreased factor VII assay, he was diagnosed as a case of congenital factor VII deficiency. After discussing the case with hematology department, it was advised for administration of recombinant factor VIIa (NovoSeven), 2 hours prior to surgery 720 μg (in a dose of 90 μg/kg), and a repeat sample for coagulation profile to be taken 1 hour after administration. Post operatively dose should be repeated 4 hourly until adequate hemostasis is achieved.

We planned anesthetic management by general anesthesia for his surgery. Blood and blood products including Fresh Frozen Plasma (FFP) and cryoprecipitate were made available preoperatively. In the operating room, an ECG along with noninvasive blood pressure, saturation with pulse oximeter and temperature monitoring was done. Two intravenous lines were secured with 24 G cannula and fluid was connected *via* a micro drip set. General anesthesia was induced with propofol-2.5 mg/kg, fentanyl l-2 μg/kg along with sevoflurane and then, the trachea was intubated following administration of rocuronium -0.9 mg/kg and confirmed by auscultation and end tidal carbon dioxide. Anesthesia was maintained with isoflurane (<1%), oxygen and nitrous oxide gas mixture and rocuronium (0.45 mg/kg). Patient had already received recombinant factor VII (Novo Seven) 2 hours prior to surgery. Throughout the procedure patient was thoroughly monitored and surgery was completed uneventfully in 2 hours and 15 mins. The patient was reversed back with neostigmine-60 μg/kg and glycopyrolate-10 μg/kg and extubated. There were no signs of any bleeding or hemorrhage/oozing and no additional rFVIIa or FFP was transfused perioperativey.

DISCUSSION

Although congenital deficiency of FVII increases the risk of posttraumatic and postoperative bleeding, it is difficult to predict the perioperative risk of bleeding due to a poor correlation between FVII activity and severity of bleeding in patients with congenital FVII deficiency [1].

Factor VII is a protein/coagulation factor required for normal hemostasis. Deficiency of factor VII can cause bleeding, particularly if factor VII is extremely low, but many cases lacking factor VII function either entirely or sub totally may not present with any history/ episodes of bleeding and can remain asymptomatic [6].

Inherited factor VII is an autosomal recessive disorder equally affecting males and females. Bleeding tendency in factor VII deficiency correlates with the level of factor VII in blood, and indeed several cases with factor VII level below 1% or 2% of normal have reportedly been asymptomatic, which shows no firm relation between actual level of factor VII and bleeding tendency [6].

The diagnosis of factor VII deficiency is usually suspected by any history of bleeding tendency, bruising, recurrent epistaxis, musculoskeletal bleeding and prolonged bleeding after dental extraction or due to accidental discovery of an abnormal PT/INR with normal aPTT [6]. The diagnosis of FVII deficiency relies on the discordance between the prolonged PT and the normality of APTT. Prolongation of INR may be from moderate (1.5-1.8) to high (>2.0), depending on plasma FVII coagulant levels and is confirmed by factor VII assays [7]. Usually when factor VII levels are below 10% and assays of other factors-II-X, fibrinogen are normal, it is clear that patient is suffering from factor VII deficiency [6].

This patient of ours presented with the similar features, with no history or evidence of bleeding or any bruising and was diagnosed incidentally during his pre-operative assessment where his coagulation profile was found to be deranged, which led to his further evaluation and diagnosis of severe factor VII deficiency which was 0.3% of the normal.

Yoshida et al., in their study suggested that factor VII levels of less than 10% is a risk factor for bleeding complications associated with surgical procedure [1]. Similarly, a case reported by Sun et al, had similar observations in their study [4]. However, Sheth et al., considered that there was a weak correlation between plasma FVII levels and the risk of bleeding; instead, the patient's personal and family history were much more important [7,8]. Sevenet, et al. also found that the occurrence and severity of bleeding were not necessarily associated with plasma FVII levels and the risk of bleeding could not be accurately predicted and that the history of bleeding was a more reliable predictor for perioperative bleeding risk [9,10].

Similarly, the patient in our case had severe factor VII deficiency 0.3%, but we did not encounter any bleeding complications, the part of which may be contributed to the fact that a good/ moderate amount of dose was administered to the patient two hours prior to the surgery and no additional measures like FFP or cryoprecipitate were required during the procedure (Table 1).

Analysis of deficiency	FFP Level	FBP Level
Sensitivity	100%	50%
Specificity	94.44%	77.78%
Positive predictive value	66.67%	20%
Negative predicted value	100%	93.33%
Accuracy	95%	75%

Table 1: The complication percentage of the Sensitivity and predictive value and accuracy of percentage calculated by first and second half's.

Many things have been used to increase plasma factor VII levels (Prothrombin complex concentrates, FFP's) but rFVIIa is the most widely accepted therapeutic option for congenital factor VII deficiency in surgical settings [1]. Although, there are no formal guidelines established for treatment of factor VII deficiency, substitution therapy is preferred for the management of bleeding episode(s) and prevention of bleeding complications during surgery [6].

Many studies have recommended it in small doses of 15-30 μg/kg(1), but we in our case after consulting the hematology used a slightly higher dose of 90 μg/kg as the patient had a severe factor VII deficiency (0.3%), while most of the other reported cases had factor VII levels of more than 1%. A bolus dose of rFVIIa was given two hours prior to surgery and was sufficient enough to achieve adequate hemostasis. There was no reported complication of thrombosis in the post-operative period.

CONCLUSION

In conclusion, patients with congenital FVII deficiency who require surgical procedure can be managed effectively and safely using rFVIIa and anti-fibronolytic agents as was seen in our patient in whom the said treatment was well tolerated and maintained effective hemostasis with good clinical outcome. Knowledge of previous history of bleeding and the patient's response to rFVIIa should be helpful in managing bleeding during surgical procedure.

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