

Successful Management of a Pregnant Lady with Hereditary Factor VII Deficiency Accidentally Discovered at Time of Delivery Without Any Need of Factor Replacement

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Abstract

Background: Factor VII deficiency is an autosomal recessive bleeding disorder with an incidence of one in 500,000. Few cases have been reported in pregnancy, and only two.

Patients were treated with recombinant factor VIIa. In the past, fresh frozen plasma or factor VII concentrate has been the conventional treatment for this disorder, with different modalities of management has been done in our presenting case.

Case Report: We report a case of factor VII deficiency diagnosed during pregnancy with a factor VII level of 30%. After prophylactic treatment apart from factor VIIa replacement therapy, the patient did not manifest any signs or symptoms of excessive bleeding during labor or postpartum.

Conclusion: Given the inherent risks of transmission of human immunodeficiency virus and hepatitis with blood products, usually the recombinant factor VIIa, as an alternative safe and effective medication, has been usually used during labor, delivery, and the puerperium. We present our case of a pregnant woman with incidentally discovered of mild factor VII deficiency who did not received recombinant factor VIIa intrapartum and successfully delivered.

Keywords: Factor VII Deficiency; Pregnancy

Introduction

Congenital Factor VII deficiency is an autosomal recessive bleeding disorder. Homozygotes usually have factor VII levels less than 10% and the severity of bleeding episodes does not correspond to the level of the factor. Neonates with factor VII deficiency are at an increased risk for developing intracranial hemorrhage at the time of delivery. FVII-deficient patient management during surgery or for long-term prophylaxis remains challenging [1].

It has been reported that prothrombin complex concentration, fresh frozen plasma (FPP), or recombinant activated factor VII could be used to prevent bleeding in patients with factor VII deficiency during pregnancy. However, there is a scarcity of data about the best method to manage patients with this rare condition [2].

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Case Report

Our patient is a 35-year-old lady. She is G1P0 with no personal or family history of bleeding disorders. Her pregnancy had been uneventful. The patient was admitted in active labor at 38+4 weeks of gestation and was found to have complete normal blood count, isolated prolonged Prothrombin Time (PT) 23.5 seconds (Normal 12.1 - 15.7), International Normalized Ratio (INR) of 1.98 (Normal 0.8 - 1.3), normal activated Partial Thromboplastin Time (aPTT) 30.7s (normal values 23 - 33s). Fibrinogen- 5.0 mg/dl. Further tests revealed a factor VII level of 30% (normal range 60 - 150%). Mixing studies suggested the absence of an inhibitor. Screening for lupus anticoagulant and anticardiolipin antibodies was negative.

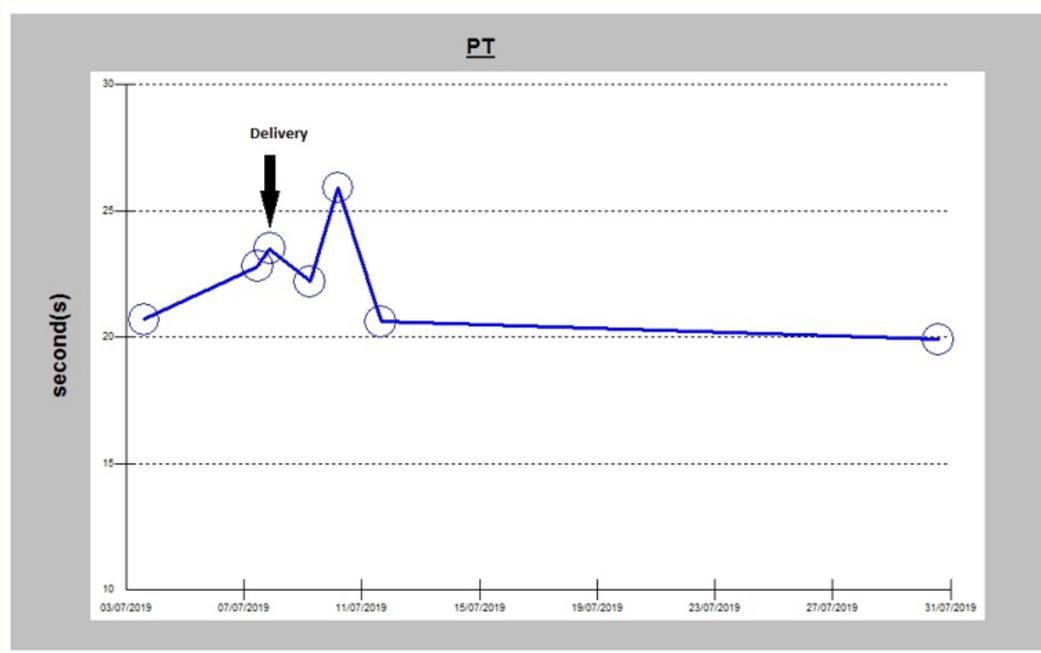


Figure 1: Prothrombin time of the patient.

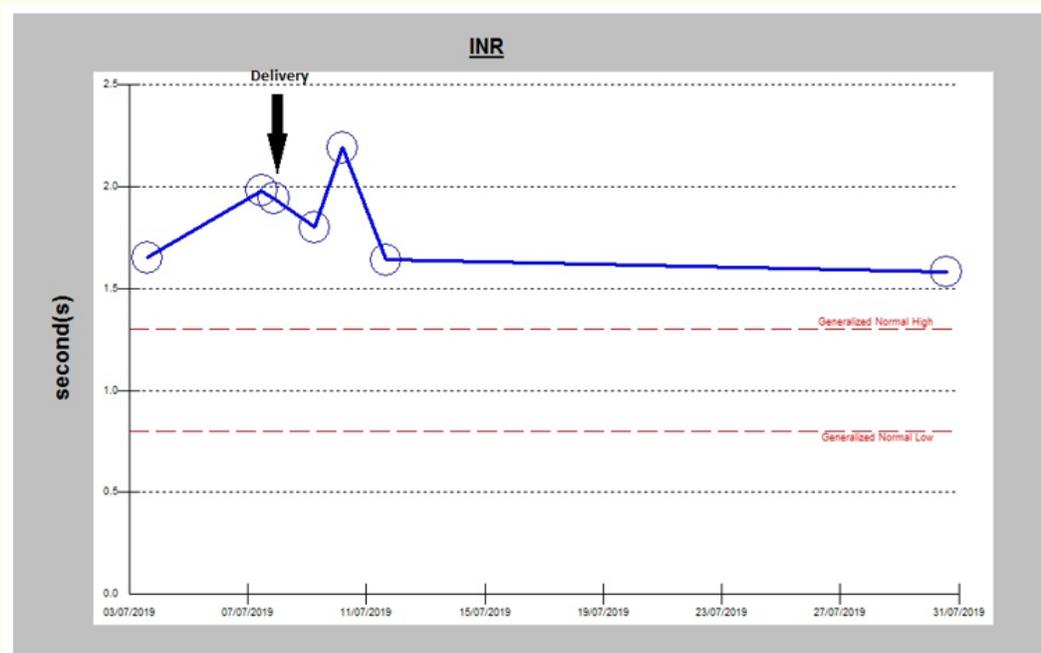


Figure 2: INR of the patient.

A multidisciplinary team, including Hematology, Obstetrics, and Maternal-Fetal Medicine, proposed a management plan for the patient. It was agreed to administer FFP 6 units once prelabour, intravenous Tranexamic acid (1 gm TID for 5 days), along with intravenous Immunoglobulin (IVIG) 1 gm/kg IVIG four hours prior to delivery, without the rush of introducing recombinant factor VIIa. She was taken for emergency cesarean section due to labor dystocia. There was no complication during the surgery with an estimated blood loss of 600 ml. with special precautions to avoid any fetal trauma. She delivered a completely normal and healthy child.

During the postpartum period, the patient was on pneumatic compression stocking as a venous thromboembolism preventive measure. On the 1st day after the surgery, the patient complained of left leg swelling. It was deemed important to do a Doppler study to rule out deep venous thrombosis (DVT). The patient received heparin infusion for five hours until the results of the study came back negative for DVT.

Otherwise, the rest of the postpartum period was uneventful.

The infant was not tested for FVII deficiency because he did not show any clinical evidence of bleeding. No other family members were tested.

Discussion

We report the first case of FVII deficiency with inhibitor in pregnancy in our Hematology center at King Saud University Hospitals, Riyadh, KSA.

There have been at least 16 reported cases of factor VII deficiency in pregnancy based on a MEDLINE search (during the years 1966 - 2000) using the key words “pregnancy” and “factor VII.” According to Our Pubmed research, only two of these patients has been treated with recombinant factor VIIa, keeping in mind that both underwent cesarean delivery [3]. Of the other remaining 14 patients, four did not receive any prophylactic treatment and needed blood transfusion, five received no prophylaxis four received prophylactic factor VII concentrate, and one received prophylactic fresh frozen plasma and all showed a good outcome [4]. Plasma-derived FVII concentrate has also been used [5] but this would have been ineffective in the presence of an inhibitor. In cases proved to have acquired factor VII inhibitors, Intravenous immunoglobulin and immunosuppression with corticosteroids and cyclophosphamide have been used in non-pregnant patients [6]. In pregnancy, high doses of glucocorticoids can lead to fetal adrenal suppression and significant maternal side effects, while use of cyclophosphamide should be limited to life-threatening situations when safer options are not available [7].

We present the first case of a pregnant woman with factor VII deficiency who did not received recombinant factor VIIa intrapartum and successfully delivered a complete healthy newly born boy.

Multidisciplinary team, including Hematology, Obstetrics, and Maternal-Fetal Medicine, proposed a management plan for the patient. It was agreed to administer FFP 6 units once prelabour, intravenous Tranexamic acid (1 gm TID for 5 days), along with intravenous Immunoglobulin (IVIG) 1 gm/kg IVIG four hours prior to delivery, without the rush of introducing recombinant factor VIIa. We believe that rFVIIa was not highly indicated in our case because the level of her factor VII was not that severe deficiency and no evidence of evident bleeding manifestations, besides people who was put on unneeded factor VIIa replacement therapy must be monitored for thromboembolism; however, this risk in otherwise healthy patients is probably low. Our case was taken for emergency cesarean section due to labor dystocia. There was no complication during the surgery with an estimated blood loss of 600 ml. with special precautions to avoid any fetal trauma.

Conclusion

We present our case of a pregnant woman with incidentally discovered of mild factor VII deficiency who did not received recombinant factor VIIa intrapartum and successfully delivered. Close monitor with multidisciplinary team may be a successful approach without the risk of neither bleeding nor thrombotic events of using unneeded factor VII replacement.

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