Management of Labour and Delivery in a Patient With Acquired Factor VII Deficiency With Inhibitor: A Case Report

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Abstract

Background: Acquired factor VII (FVII) deficiency with inhibitor increases the risk of hemorrhage during pregnancy. However, there are no published reports guiding its management in the peripartum period.

Case: A 24-year-old woman with inhibitory antibodies to FVII delivered at 34 weeks of gestation. The patient was administered recombinant factor VIIa (rFVIIa) and tranexamic acid. There were no bleeding-related complications; however, the FVII level was supratherapeutic. The patient returned during a second pregnancy. A reduced dose of rFVIIa was administered. The delivery was complicated by postpartum hemorrhage, which resolved with the addition of uterotonic agents.

Conclusion: Recombinant FVIIa and tranexamic acid offer an effective peripartum treatment in women with inhibitory antibody to FVII. Further research should delineate the optimal time of administration.

Résumé

Contexte : Le déficit acquis en facteur VII (FVII) chez les patientes qui présentent des anticorps inhibiteurs entraîne une hausse du risque d'hémorragie pendant la grossesse. Toutefois, aucun rapport publié n'en guide la prise en charge pendant la période péripartum.

Cas : Une femme de 24 ans présentant des anticorps inhibiteurs en ce qui concerne le FVII a accouché à 34 semaines de gestation. Nous lui avons administré du facteur VIIa recombinant (rFVIIa) et de l'acide tranexamique. Aucune complication associée aux saignements n'a été constatée; toutefois, le niveau de FVII était suprathérapeutique. La patiente nous a de nouveau consultés dans le cadre d'une deuxième grossesse. Une dose moindre de rFVIIa lui alors été administrée. L'accouchement a été compliqué par une hémorragie postpartum, laquelle s'est résorbée à la suite de l'ajout d'agents utérotoniques.

Conclusion : L'administration de facteur VIIa recombinant et d'acide tranexamique constitue un traitement péripartum efficace pour les femmes qui présentent des anticorps inhibiteurs en ce qui concerne le FVII. La tenue d'autres recherches s'avère requise pour la détermination du moment optimal pour l'administration de ce traitement.

Key Words: Factor VII deficiency, recombinant factor VIIa, pregnancy, postpartum hemorrhage, coagulopathy

Competing Interests: None declared.

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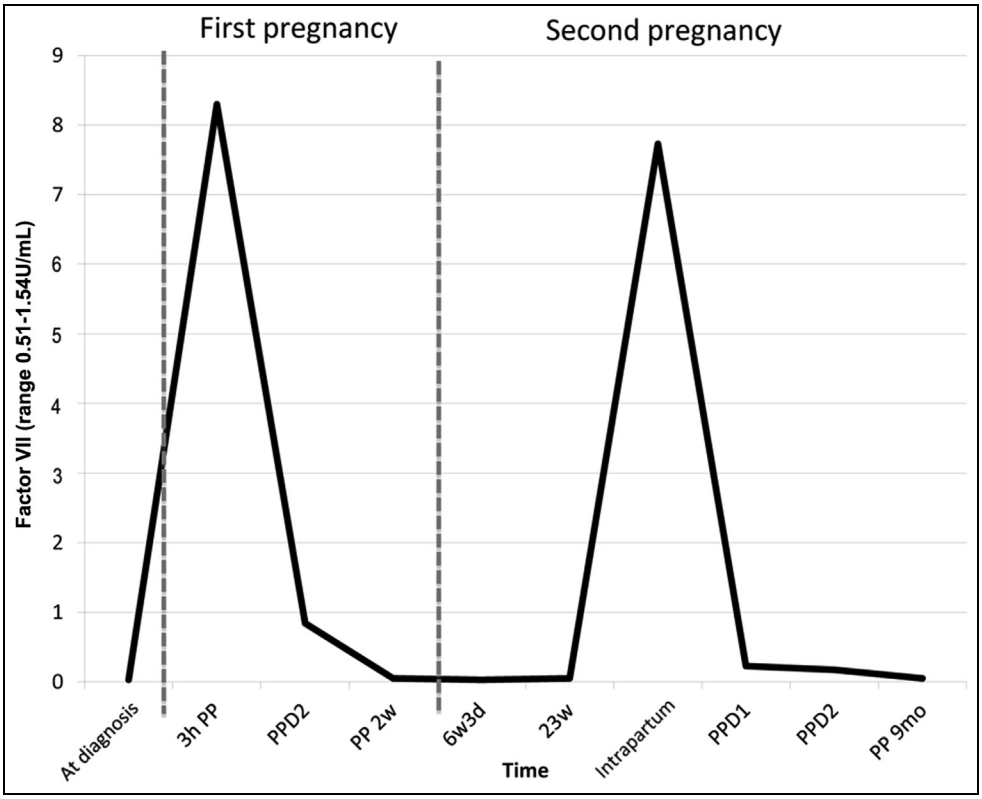
INTRODUCTION

Acquired factor VII (FVII) deficiency with an inhibitory antibody has been described in only a few case reports.[1–5] Management principles reported include FVII replacement and elimination of the inhibitor by apharesis or immunosuppression. The hemostatic level of FVII in the presence of an inhibitor is unknown. In congenital FVII deficiency, a level of 10 to 15 units per decilitre has been proposed,[3] but FVII levels have been poorly correlated with clinical manifestations of bleeding.[6]

Pregnancy poses challenges for management in patients with coagulopathies. In pregnancy, FVII activity increases fourfold in the third trimester.[6] With congenital FVII deficiency this response is impaired, but the risk of obstetric hemorrhage does not parallel plasma markers of disease.[7] Whether these findings are applicable to patients with the acquired factor deficiency is unknown.

We describe here the peripartum management of acquired FVII deficiency due to a known inhibitor antibody to FVII during two consecutive pregnancies in the same patient.

Figure. Factor VII levels at diagnosis and over the course of the first and second pregnancies (normal range 0.51 to 1.54 U/mL). EGA, estimated gestational age; d, days; PP, postpartum; PPD, postpartum day; w, weeks; mo, months



THE CASE

A 24-year-old woman, gravida 3 para 0, presented with threatened preterm labour at 34 weeks of gestation. Her obstetric history included an uncomplicated therapeutic abortion and an early miscarriage. The latter was managed expectantly, and the patient had significant prolonged vaginal bleeding and ecchymoses. Prothrombin time (PT) was prolonged at 49.5 seconds (normal 10.6 to 13.2), PT INR was 4.2 (normal 0.8 to 1.2), and activated partial thromboplastin time was normal. Further evaluation revealed an isolated FVII level of 0.03 U/mL (range 0.51 to 1.54). Mixing studies suggested the presence of an inhibitor. A Bethesda assay also demonstrated the presence of an inhibitor to FVII measuring 5 Bethesda units (normal 0). Screening for lupus anticoagulant and anticardiolipin antibodies was negative. Family and personal histories were noncontributory. It was hypothesized that the patient acquired the inhibitor during a previous pregnancy. Her current pregnancy had been uneventful.

In the management plan created by a multidisciplinary team including representatives of obstetrics, maternal-fetal medicine, and hematology, the patient would be administered recombinant factor VIIa (rFVIIa) at the onset of labour and for at least 24 hours postpartum. Because of the potential risk of fetal bleeding due to transplacental transfer of the inhibitor to the fetus, it was deemed important to avoid fetal scalp sampling, fetal scalp electrodes, and vacuum or forceps delivery. The patient's coagulation parameters were abnormal, and the behaviour of this rare coagulopathy in labour was unknown; both of these indicated a need to avoid neuraxial anaesthesia.[8] An antepartum anaesthesia consultation emphasized concerns about spinal or epidural hematoma, which could cause spinal compression, and the consultant advised against the use of neuraxial analgesia.

Following a period of relative uterine quiescence, the patient rapidly progressed through labour. She had vaginal delivery of a healthy infant weighing 2380 g. The patient received rFVIIa, 90 μg/kg, at the onset of the third stage of labour. The placenta was retained, and after 30 minutes of expectant management it was removed manually. No excessive maternal or fetal bleeding was noted.

Recombinant FVIIa dosage was repeated every two hours for 24 hours. The patient also received oral