

Successful Management of A Vaginal Delivery and Postpartum Hemorrhage in A Patient with Von Willebrand Disease Type: Case Report

Von Willebrand Hastalığı Tip 3 Tanılı Hastada Başarılı Doğum ve Postpartum Hemoraji Yönetimi

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ABSTRACT Von Willebrand disease, which has severe bleeding manifestations like intrapartum and postpartum hemorrhage, was first described by Erik von Willebrand in 1926. The replacement of plasma concentrates containing functional von Willebrand factor and factor VIII is the keystone of the treatment for the disorders due to the disease. A 21-year old nulliparous woman with von Willebrand's disease type 3 was referred to our clinic for perinatal management. Her routine pregnancy follow-up was problemless. At 38 weeks' gestational age, she delivered a 3140 g baby by vaginal route. She was given factor VIII at the labour and for the postpartum 3 days. One week later she suffered from heavy vaginal bleeding. At that time she was also given factor VIII with von Willebrand's factor, ristocetin co-factor and tranexamic acid for another 5 days. Because of the tendency of the hemorrhage at the postpartum first week period, vaginal delivery of a patient with von Willebrand's disease should be monitored very closely.

Key Words: Von Willebrand disease; delivery, obstetric; postpartum hemorrhage

ÖZET İntrapartum ve postpartum hemoraji gibi ciddi kanama sorunları yaratabilecek bir durum olan Von Willebrand hastalığı, ilk olarak 1926 yılında Erik von Willebrand tarafından tanımlanmıştır. Bu hastalığa bağlı bozuklukların tedavisinde von Willebrand faktörü ve faktör VIII içeren plazma konsantrasyonlarının replasmanı ana rolü oynamaktadır. Von Willebrand hastalığı Tip 3 tanılı 21 yaşında nullipar bir gebe perinatal takip için kliniğimize yönlendirildi. Rutin takiplerinde bir sorun saptanmadı ve 38. gebelik haftasında vajinal yolla doğum yaptı. Doğum anında ve takip eden 3 gün boyunca faktör VIII tedavisi verildi. Doğumun 1. haftasında şiddetli vajinal kanama yakınmasıyla başvurdu. Faktör VIII içeren von Willebrand faktör, ristocetin ko-faktör ve traneksamik süresince verildi. İlk haftalardaki kanama yatkınlığı nedeniyle von Willebrand hastalığı olan lohusalar yakından takip edilmelidirler.

Anahtar Kelimeler: Von Willebrand hastalığı; doğum, obstetrik; postpartum kanama

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Von Willebrand disease (vWD) is an autosomally inherited bleeding disorder caused by abnormalities of von Willebrand factor (vWF) which is a multimeric glycoprotein essential for platelet plug formation at the site of vessel injury.¹ vWF is decreased in type 1 and functionally defective in type 2.² Type 3 is the least common of all forms and is characterised by very low (or even undetectable) levels of plasma vWF and factor VIII with severe bleeding disorders.³ Epistaxes, mucosal bleedings, hemorrhage after surgery, and excessive menstrual blood loss are the common manifestations of vWD.⁴ Although levels of vWF increase during preg-

nancy, rapid postpartum decrease may lead to bleeding problems.⁵

Here, we report a case of successful management of a vaginal delivery and postpartum bleeding in a patient with von Willebrand disease type 3.

CASE REPORT

A 21-year-old gravida 1 patient who had a diagnosis of vWD type 3 delivered vaginally. She did not experience any complications or bleeding during her pregnancy. She had been diagnosed to have vWD by being evaluated for severe hemarthrosis after a knee trauma when she was 5-years-old. She was given fresh frozen plasma support at that time. Her following past medical history was unremarkable and she reported normal menses.

On the 38th week of gestation, she had severe painful contractions. On vaginal examination she had 4 cm dilatation of cervix. The level of vWF was 1.7%, von Willebrand factor antigen was < 10%, ristocetin cofactor activity (RICOFA) was 10.8%, activated partial thromboplastin time (aPTT) was 53 seconds. She was given an intravenous bolus of 2000 units of factor VIII (IMMUNATE, BAXTER). 4 hours later, she delivered a 3140 g male baby with a right mediolateral episiotomy. The amount of postpartum bleeding was acceptable and episiotomy incision was repaired. On postpartum 12th hour, another 2000 units of factor VIII (IMMUNATE, BAXTER) was given. Postdelivery period was uneventful. She was also treated with 2 x 2000 units of factor VIII (IMMUNATE, BAXTER) for the following three days. She was discharged from hospital on the 4th postpartum day.

Seven days after the delivery, she was admitted to our service with the complaint of heavy vaginal bleeding. No acute abdomen was detected, and ultrasonographic evaluation did not show signs of retained products in uterine cavity or fluid collection consistent with intraabdominal bleeding. There were normal findings in the examination of episiotomy incision, vagina and cervix. Her hemoglobin level and aPTT were 7.1 g/dL and 43.8 seconds, respectively. She was transfused 2 units of red blood cell and was started a factor VIII replacement

of 3 x 500 units/day with 3 x 1200 IU vWF and ristocetin cofactor. (HAEMATE P 500, BEHRING). Also, she was given intravenous tranexamic acid 4 x 500 mg daily. Her medication continued for 5 days. Finally, her vaginal bleeding decreased and she was discharged. The patient is free of any symptoms nearly 3 months after the delivery.

DISCUSSION

The most common of the hereditary bleeding dyscrasias, vWD affects 1% of the general population.^{6,7} Major symptoms are bruising, nosebleeds, bleeding after injury, surgery or tooth extraction; postpartum bleeding; and menorrhagia.⁸

Today, pregnancy is not contraindicated in patients with coagulation disorders but requires a multidisciplinary approach to management. According to some authors, bleeding due to the coagulation disorder itself is rare during pregnancy because the level of vWF increase in gestational period. If an invasive diagnostic (e.g., amniocentesis) or therapeutic procedure is planned, factor levels - appropriate for the disorder- should be measured prior to the procedure and be replaced when needed.^{9,10} Our patient did not take any medications during her routine pregnancy follow up, but she was given factor VIII concentrate just before the vaginal delivery. Despite such a treatment, she experienced late postpartum bleeding and needed replacement of higher doses. It is obvious that women with vWD are more likely to have certain medical conditions at the time of delivery than women without vWD. FVIII and von Willebrand Factor:Ristocetin co-factor (vWf:RCo) levels along with an aPTT, complete blood count, type and crossmatch should be obtained on admission to the hospital for delivery.¹¹ FVIII and vWF:RCo levels below 50 IU/dL should be corrected before delivery or regional anesthesia. An inherited bleeding disorder is not an indication per se for delivery by Caesarean section; a decision to proceed with a caesarean section should be based on obstetric indications and the delivery should be as atraumatic as possible, minimizing maternal genital tract and (or) perineal lacerations. We chose vaginal route for labour and did not use any instruments. Desmo-

pressin acetate (DDAVP), when administered as prophylaxis to prevent postpartum hemorrhage, is usually given at the time of cord clamping, but, because the peak effect is 40 to 60 minutes after administration,* it may be more beneficial if administered during the second stage of labour or immediately before cesarean delivery.¹² Vacuum, forceps, fetal scalp samplers or electrodes should not be used.¹³ In a survey of 102 women with vWD published by the United States Centers for Disease Control and Prevention, 59% reported a history of postpartum hemorrhage compared with 21% of controls and in another survey, 31% of women with vWD reported a history of increased postpartum bleeding compared with 10% of controls.^{14,15} Also, the risk of late postpartum hemorrhage is increased to 11% to 24% in women with bleeding disorders compared to less than 1% in the general

population.^{16,17} In a review of the literature, it was determined that the average time of presentation of postpartum hemorrhage in women with vWD is 15.7 ± 5.2 days after delivery.¹⁸ Because of the increased complication chance in postpartum period, women with vWD should be evaluated more frequently. Thus, weekly contact is recommended during the postpartum period.¹⁹ Our patient had severe hemorrhage at the first week of delivery just before her control. She was not using any medicine. According to review by James et al., prophylaxis for 2 or more weeks postpartum may be required.²⁰ Prophylaxis should be considered if FVIII or VWF:RCo levels fall below 50 IU/dL. In responders receiving DDAVP, however, more than 300 mg/day of stimate intranasal DDAVP for more than 3 consecutive days may increase the risk of hyponatremia. Because nonsteroidal anti-inflammatory drugs may affect platelet function and systemic hemostasis alternative analgesics should be prescribed.²¹

* Stimate (Desmopressin acetate) Nasal Spray, 1.5 mg.mL, Prescribing Information. Aventis Behring, 2002. p.1-2.

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