SPINAL ANESTHESIA FOR EMERGENCY CESAREAN DELIVERY IN VON WILLEBRAND DISEASE

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ABSTRACT

Von Willebrand disease(vWD) is a common hemostatic disorder encountered inobstetric anesthesia practice. This needs appropriate pre-operative evaluation to choose the best technique of anesthetic management. Evaluation needs to be individualized to consider the risk and benefits of each technique, either general or neuraxial anaesthesia. There are only a few published cases on use of spinal anesthesia in vWD patients for caesarean delivery. We present a case of vWD patient who was given spinal anesthesia for emergency caserean delivery. We were not able to determine the type of the disease, but based on clinical features it was assumed to be of type I or II. Intranasal desmopressin was given as bleeding prophylaxis. Anaesthetic considerations and peri-operative management are discussed.

Keywords: *Von Willebrand's disease(vWD); Caesarean section; Anesthesia.*

INTRODUCTION

Von Willebrand disease (vWD) is the commonest inherited hemostatic disorder with a prevalence of 1%- 2% in general population. It is named after Erik Adolf Von Willebrand, a Finnish pediatrician in 1926. It is associated with quantitative or qualitative deficiencies of von Willebrand factor (vWF) and factor VIII, which result in mild to moderate bleeding episodes (1).

Spinal anesthesia may be considered for patients with vWD since both vWF and factor VIII levels increase in pregnancy with a peak in the third trimester (2,3)There are only a few case reports describing the use of spinal anesthesia for labor or caesarean section in patients with vWD (4-7). We present the anesthetic considerations of a patient with vWD for emergency caesarean delivery.

CASE REPORT

35year old primigravida, diagnosed with von Willebrand disease (vWD) at the age of 18, was admitted at 36weeks of gestation for safe confinement. On admission, her weight was 88 kg with a height of 157 cm. Laboratory evaluation after admission showed platelet count - 1.5 lakh /mm^3, hematocrit - 30%, Prothombin Time (PT) - raised by 0.3 Sec and activated Partial Thromboplastin Time (aPTT) by 19 sec, INR- 1.02, vWF:antigen (vWF:Ag) - 26 IU/dl (50-200 IU/dl), Factor VIII plasma assay- 60 IU/dl (60-150 IU/dl).

vWF:Rco levels (Ristocetin cofactor activity) could not be obtained due to unavailability of this investigation in the institute's laboratory. Hence, it was difficult to differentiate between type 1 and type 2 vWD as vWF:Rco/vWF:Ag ratio could not be determined.

Fetal heart rate was reassuring and patient was continuously monitored in our antepartum unit. Obstetric plan was to continue expectant line of management till 38 weeks of gestation. Patient was explained the risks and benefits of neuraxial anaesthesia for vaginal or caesarean delivery, which would be depending on her coagulation status at that time.

At 37 weeks gestation, decision for emergency caesarean section was taken following spontaneous rupture of membranes with non-reassuring fetal heart rate pattern. Desmopressin acetate (DDAVP) 300mcg nasal spray was given as bleeding prophylaxis 90 minutes prior to surgery.

The patient received metoclopramide 10 mg IV, ranitidine 50 mg IV and was coloaded with 500ml of Ringer lactate solution. Tranexamic acid was used as a hemostatic adjunct .With the patient placed in left lateral position, lumbar subarachnoid block was performed at the L3-L4 interspace with 25G Quincke Babcock spinal needle in a single attempt. 1.8 mL of 0.5% hyperbaric bupivacaine with 10 mcg fentanyl, was injected intrathecally. The patient was positioned with left lateral tilt. A sensory level to pinprick and light touch at T4 was recorded. Surgery proceeded uneventfully. Estimated surgical blood loss was 1000 mL. An oxytocin infusion (10 IU in 500 mL Lactated Ringer's solution) was commenced after delivery of the placenta.

The patient's postpartum course was uneventful, and before discharge her Hct was 30% and her factor VIII plasma assay was 75 IU/dl.

DISCUSSION

vWD is an autosomal dominant disease characterized by quantitative or qualitative defect of

von Willebrand's factor (vWF), a key component of primary and secondary haemostatic process. vWF also binds to factor VIII protecting it against degradation. vWD is classified into three types.

Type 1 (60%- 80% of cases) with quantitative vWF deficiency, with a 5- 30% reduction in vWF and factor VIII: C.

Type 2 vWD with qualitative defects in vWF and is divided into four subtypes (2A, 2B, 2M, and 2N). Type 2B subtype can present as thrombocytopenia in pregnancy (8).

Type 3 (1% of patients) is characterized by negligible or absent levels of vWF and significantly reduced levels of factor VIII coagulant. It is the most severe type.

It is important to determine the type of vWD because it allows the anaesthesiologist to plan the management accordingly. It was presumed that the patient had type 1 or type 2 vWD based on history of mild to moderate bleeding episodes and laboratory values(9,10).

Laboratory Values for VWD

Condition	Description	VWF:RCo (IU/dL)	VWF:Ag (IU/dL)	FVIII	VWF:RCo/ VWF:Ag
Type 1	Partial quantitative VWF deficiency (75% of symptomatic VWD patients)	<30*	<30*	↓ or Normal	>0.5–0.7
Type 2A	↓ VWF-dependent platelet adhesion with selective deficiency of high-molecular-weight multimers	<30*	<30–200*†	↓ or Normal	<0.5–0.7
Type 2B	Increased affinity for platelet GPIb	<30*	<30–200*†	↓ or Normal	Usually <0.5–0.7
Type 2M	↓ VWF-dependent platelet adhesion without selective deficiency of high-molecular-weight multimers	<30*	<30–200*†	↓ or Normal	<0.5–0.7
Type 2N	Markedly decreased binding affinity for FVIII	30–200	30–200	↓↓	>0.5–0.7
Type 3	Virtually complete deficiency of VWF (Severe, rare)	<3	<3	↓↓↓ (<10 IU/dL)	Not applicable
"Low VWF"**		30–50	30–50	Normal	>0.5–0.7
Normal		50–200	50–200	Normal	>0.5–0.7

(Adapted from:- Reference No. 9)

vWF and factor VIII levels were found to be substantially increased(375% and 200% respectively) by term in healthy pregnant patients (2). In contrast, Sanchez-Luceros et al (3) found much smaller increases in vWF and factor VIII levels (60% and 6% respectively). Factor VIII levels should be monitored because of the variable increases seen in pregnancy (11). Clear guidelines on laboratory monitoring or indications for regional or general anaesthesia are not available. General anesthesia is best avoided due to anticipated difficult airway and risk of aspiration, unless required in documented coagulopathy, severe fetal distress, maternal refusal of regional anaesthesia or failed regional attempts. We have followed the Clinical Practice Guideline (2012) of American Society of Hematology on the Evaluation and Management of von Willebrand Disease(17).

Cohen et al (4,7) reported two patients with vWD who received epidural analgesia in labor without any complications; however the vWD subtypes were not described. Milaskiewicz et al (5) described cesarean delivery in a patient with type 1 vWD with epidural anesthesia. Jones et al(6) performed combined spinal-epidural labor analgesia in a patient with type 2A vWD with severe preeclampsia. Kadir et al (11) produced a case series of patients with vWD including the use of regional anesthesia in 8 of these patients. However, no information regarding the anesthetic technique used or the vWD subtypes was provided. The risk of spinal hematoma is rare after spinal anesthesia in obstetric patients (1:200 000), and this complication is usually reported in the presence of severe coagulopathy (16).

Recommended laboratory monitoring before planning for regional techniques include complete blood count, platelet count, PT and aPTT, Factor VIII activity, vWF antigen (vWF:Ag) and vWF ristocetin cofactor activity (vWF:Rco).

VWF:RCo and Factor VIII levels of at least 50 IU/dL should be acheived before delivery and these levels should be maintained for 3-5 days for normal labor and upto 7days after caesarean section in view of continiuing risk of postpartum haemorraghe (17). In such cases regional anesthesia

may be considered if no other coagulation defects are present (9).

Desmopressin acetate(DDAVP) is a synthetic analogue of vasopressin that increases plasma vWF and factor VIII levels three to five times higher than baseline levels within 30 to 60 minutes (12). Administration of DDAVP may be IV (0.3 mcg/kg) or intranasal 150 mcg or 1 spray for persons <50 kg and 300 mcg or 2 sprays for persons weighing ≥50 kg (18). Patients with type 1 vWD are likely to show a good response to DDAVP(10). Cohen et al described the use of DDAVP in a patient with vWD receiving epidural analgesia for labor(4). Patients should limit fluid intake for 24 hours following DDAVP. Serum electrolytes should be monitored after surgery or after multiple doses of DDAVP. (13)

vWF concentrate and factor VIII concentrate are more commonly used in patients who show inadequate response to DDAVP, such as those with types 2 and 3 vWD [14]. 40 to 60 Ul/kg daily is the recommended dose. For surgical patients, factor VIII should be dosed every 12 hours on day of surgery and every 24 hours postoperatively.

Postoperative deep venous thrombosis has been reported in nonobstetric patients with vWD receiving repeated infusions of factorVIII concentrates [14,15,16). Antifibrinolytic amino acids(EACA) or tranexamic acid are also useful adjuncts for surgical hemostasis.

Fresh frozen plasma may be used to correct coagulation defects. Cryoprecipitate has 5 to 10 times more factor VIII and vWF as compared to fresh plasma (13). Cryoprecipitate every 12 or 24 hours normalizes factor VIII levels. In patients with fibrinogen levels less than 0.8 g/l, 10 to 15 units of cryoprecipitate are needed.

The best anesthetic option for patients with vWD should be decided on a case-by-case basis as demonstrated by Butwick et al who successfully performed neuroaxial anesthesia in type 1 vWF(18). The coagulation status of our patient was carefully monitored to ensure that hematologic goals were achieved. The decision to proceed with regional versus general anaesthesia was made after risk-benefit consideration. In addition, our patient maintained a strong desire to avoid general anesthesia for her cesarean delivery. We couldn't use a pencil point needle because it was unavailable at that time and we were not able to arrange one due to the emergency nature of the procedure.

Close monitoring of factor VIII and vWF is advised for all patients with type 1 vWD because of the potential variability during pregnancy. The decision to use regional anesthesia should be individualized. Further studies on neuraxial anesthesia in obstetric population is necessary to establish the consensus for routine practice of this technique

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