ECTOPIC PREGNANCY IN A CASE OF VON WILLEBRAND DISEASE

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ABSTRACT

BACKGROUND
Von Willebrand disease is a most common inherited bleeding disorder. It is caused by either a qualitative/quantitative defect in Von Willebrand factor. Patients may have extensive mucosal bleeding & prolonged bleeding after surgery. Bleeding complications in pregnancy are more common when the Ristocetin cofactor assay & factor VIII levels are <50 IU/dL.\(^1\) In this article, we discuss about a known case of vWD presented with ruptured ectopic pregnancy following previous LSCS. Such case when presenting in emergency, expertise in obstetric management and intensive care together saved the patient.

KEYWORDS
Von Willebrand Disease, Ectopic Pregnancy, Haemoperitoneum, Tranexamic Acid, DDAVP, Desmopressin.


BACKGROUND
A 26-year-old G2, P1, L1, previous LSCS presented with 35 days of amenorrhoea & lower abdominal pain since 2 days. History revealed that the patient was a known case of Von Willebrand disease since 3 years of age. She had repeated blood transfusions. In 1991, she was admitted with buccal haematoma. In 2004, she landed in dental op with bleeding gums & received 3 units of packed cell transfusion. In 2005, she was admitted with puberty menorrhagia in our hospital & transfused with 2 units of blood. In 2011 underwent laparoscopic ovarian cystectomy with 2 units of blood transfusion.

Previous Investigations
Factor VIII - 1.4%, Vwf Rco- 10.7%, Vwf Ag - 10.4 u/dl, RIPA – Normal response, Vwf (CBA) 4.9%. VWF binding assay – 100.2%. Platelet count & morphology normal. Normal PT & Fibrinogen level, Prolonged bleeding time. Prolonged APTT with normal plasma & factor IX deficient plasma but not with factor VIII deficient plasma. Decreased levels of F VIII, Vwf, Rco, Vwf Ag. Decreased levels of collagen binding assay. CBA/VWF 0.4%. Normal Vwf: factor VIII binding assay & platelet function. Patient is diagnosed to have Von Willebrand Disease Type II A.

Previous Obstetric History
On 8.6.2013, patient underwent LSCS & delivered an alive term female child. DDAVP Inj. 0.3 microgram given as IV 1 hr. before surgery over 30 min. & postoperatively 12th hourly up to 48 hours. Desmopressin nasal spray 10 microgram was given 1 hour before surgery & one amp. of Tranexamic acid was given. 4 units of FFP & 2 units of whole blood given.

Patient was discharged on 16th POD & readmitted on 26.6 2013 with bleeding. Desmopressin nasal spray continued 12th hourly. Factor VIII with Vwf given at a rate of 20 U/kg 12th hrly. Patient was discharged & advised regular followup.

Patient now presented with 35 days of amenorrhoea, lower abdominal pain & spotting p/v. Patient mildly anaemic, PR- 82/min., BP- 110/80 mmHg, P/A- mikiad abdominal distension. tenderness in left iliac fossa, P/V- uterus normal size, antverted, left fornical tenderness +, right fornix fullness +.

Inquiries
UPT positive, BT- 3 min., CT- 6 min., RBS- 106 mg%, B. urea – 28 mg%, S. creatinine – 0.9 mg%, Platelet – 2.6 lakhs, PT – 11.8 sec, INR – 0.92 sec. Emergency USG revealed complex bilateral adnexal masses with moderate haemoperitoneum. Culdocentesis – Positive.

With 4 FFP, 2 cryoprecipitates, 2 WB, 1 unit of packed cell transfusion & with Inj. Tranexamic acid 500 mg IV 6th hrly Emergency Laparotomy & Bilateral Salpingo-Oophorectomy done. 5 x 7 cm right haemorrhagic cyst ruptured left ampullary tubal ectopic with 5 x 4 cm Left ovarian cyst. Both ovaries unhealthy. 1000 mL of haemoperitoneum, 200 g of blood clots.

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Management


Histopathology

Microscopy

Ruptured lumen of fallopian tube showing chorionic villi with haemorrhage. Picture is consistent with products of conception. Section from other tube – normal. Section studied from one ovary- shows picture s/o haemorrhagic cyst. Section studied from other ovary shows haemorrhagic cyst with haemosiderin-laden macrophages- s/o endometriotic cyst.

DISCUSSION

Von Willebrand disease is the most commonly inherited bleeding disorder. Prevalence – 1 to 2 %. Heterogenous group of approx. 20 functional disorders involving aberrations of factor VIII complex & platelet dysfunction.

Type I- Most common; Autosomal dominant. Deficiency of structurally normal Vwf.

Type II- Less common; autosomal recessive. Qualitative defects in Vwf.

Type II A- Deficiency of normal high MW multimers of VWF.

Type II B- Due to increased affinity for platelets.

Type III- Least common; Autosomal recessive. Most severe form- severe deficiency, which causes factor VIII deficiency.

Pregnancy and VWD

Pregnancy increases the level of Vwf & factor VIII & patients are less likely to need therapy. Levels of Vwf & factor VIII should be assessed in third trimester. Levels of 50 IU/dL - should be reached before delivery & maintained for 3 days following vaginal delivery & at least 5 days following caesarean section.

Primary Treatment

1-deamino 8-D arginine Vasopressin (DDAVP), which increases plasma factor VIII & Vwf levels. Dose: 0.3 microgram/kg to a max. of 20 microgram given sc/ diluted in 50 to 100 mL of normal saline & given IV for 30 mins. Response is gauged by assessing change in platelet count & Vwf: Ristocetin [RCoF] activity at 90 min. after administration of DDAVP.

Adverse Effects

Headache, flushing, BP changes, fluid retention & hyponatraemia. Women who don’t respond to DDAVP – treated with factor VIII, Vwf concentrates, cryoprecipitate. Prenatal diagnosis is possible. Genetic counselling should be offered to affected families.

Labour and Delivery

Elective caesarean section- not routinely advised. Traumatic delivery – avoided. Ensure adequate uterine contractions in third stage of labour. Careful & prompt repair of episiotomies & perineal tears should be undertaken. Regional anaesthesia – safe if vWF:RCoF Levels > 50 IU/dL. Postnatally close observation for PPH. DDAVP- treatment of choice to prevent & treat mild-to-moderate postpartum bleeding. Tranexamic acid may be used as an adjunct. NSAIDs avoided.

CONCLUSION

Antepartum period is the ideal time to characterise the type of VWD & the response to DDAVP. Multidisciplinary team including haematologist, obstetrician & anaesthesiologist should coordinate & form a management plan.

REFERENCES