VON WILLEBRAND'S DISEASE COMPLICATING PREGNANCY AND LABOUR

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SUMMARY

Von Willebrand disease is an inherited disorder of the Von Willebrand factor portion of the Factor VIII complex. Though this is the second commonest inherited bleeding disorder found in childhood the publications on its association with pregnancy and management are not very common in Indian literature, though there are reports in the English literature. These patients give a history of increased bleeding from minor trauma from childhood and menorrhagia. During pregnancy, postpartum haemorrhage, and bleeding from surgical wounds are common. The diagnosis is established on the basis of a prolonged bleeding time with a normal platelet count, clotting time and a lot Factor VIII level. Cryoprecipitate is the treatment of choice and it has to be given before the patient goes into labour or being taken up for any surgery, and the replacement should be continued for one week to guard against delayed haemorrhage.

Introduction

Von Willebrand's disease is an inherited disorder of haemostasis affecting both sexes, that is characterised by mucocutaneous, post traumatic, post operative and postpartum bleeding. (Lipton R.A. 1982). It is a disorder of the von Willebrand factor portion of the Factor VIII: complex. The publications on the management and outcome of pregnancies in patients with von Willebrand's disease (vWD) is scarce in Indian literature. In view of this we are reporting these two cases with vWD managed during delivery in this hospital.

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Case I : A.M, a twenty one year old primigravida was admitted to the hospital at thirty seven weeks gestation in August 1983. She had been diagnosed to have von Willebrand's disease at the age of six years when she had presented with excessive bleeding from minor trauma, easy bruisability and bleeding gums. She attained menarche at sixteen years. Her periods for the first two years were 4-5/30. Later she developed menorrhagia 12-15/30. Periods were controlled with oral contraceptives.

Her pregnancy was supervised by her family physician and was uneventful.

TABLE - I
HAEMATOLOGY DATA OF THE TWO PATIENTS

		Case I	*Case II*
1.	Haemoglobin	12.6 gm%	6.1 gm%
2.	Bleeding time (Normal 2-6 mts)	>15 minutes	>15 minutes
3.	Clotting time (Normal 10-20 mts)	10 minutes	14 minutes
4.	Prothrombin time	Control 11" Pt. 12"	Control 11.5" Pt. 12"
5.	Partial Thromboplastin time	Control 87" Patient 168"	A.P.T.T. Control 32" Patient 57" (Partial clot)
6.	Correction studies	Corrected by adsorbed plasma	Corrected by adsorbed plasma
7.	Factor assay	Not done	Factor VIII:C activity of 2.6%

She was referred to hospital for delivery. Baseline haematologic data are shown in Table I.

Although it was planned to induce labour at 38 weeks after infusion of fresh plasma, the patient went into spontaneous labour earlier and was delivered of a healthy male infant by outlet forceps. She was given four units for fresh plasma and one pint of AB positive fresh blood during labour, as cryoprecipitate/Factor VIII concentrates were not available at that time. She started to bleed PV and from the episiotomy site, one hour after delivery. She was transfused twelve units of fresh plasma and four pints of fresh blood over 48 hours. The bleeding was less eight hours after delivery and was controlled in 48 hours. Amicar (EACA), 1g Q8H was given after checking the urine microscopy for haematuria to prevent excessive fibrinolysis and was gradually stopped on the eighth day.

Case II : V.M, a twenty seven year old G2 P1 L1 was seen at the antenatal clinic at 35 weeks gestation in June 1988. Her first pregnancy was a caesarean section done six years ago for foetal distress at another hospital. She had post partum haemorrhage and required three transfusions.

She gave a history of easy bruisability, bleeding gums and excessive bleeding following trauma from childhood. She had five blood transfusions for excessive bleeding. Since menarche she had menorrhagia. An older sister had died of similar problems.

She was admitted at 37 weeks gestation. Baseline haematologic data are shown in Table I: Her Factor VIII:C activity was 2.6%. She was transfused 3 units of bank blood for anaemia.

She went into spontaneous labour at 38 weeks. She was given eight bottles of lyophilised cryoprecipitate and her post transfusion F.VIII:C level was 35%. As she developed suprapubic tenderness and foetal tachycardia she was delivered by emergency LSCS under general anaesthesia. She was given eight more bottles of cryoprecipitate on the first day and the F. VIII: C level was 84%. Haemostasis during surgery was good and there was no PPH. She was given 4 bottles of cryoprecipitate on alternate days till the sutures were removed on the 9th day.

Discussion

Von Willebrand's disease occurs from a deficiency or abnormality of a high molecular plasma protein (von Willebrand factor) that circulates as a complex series of multimers of a single subunit. This is required for a normal platelet function and to carry the Factor VIII: C in circulation Lipton R.A. 1982). The clinical features of this disease are extremely variable. During pregnancy there is an increased risk of postpartum haemorrhage (Punnonen R, 1981). The chief immediate cause of excessive bleeding during and after labour in women with vWD is lack of Factor VIII: C. A prolonged bleeding time is characteristically found these patients, while the clotting time is variable.

The literature on the management and outcome of pregnancy in patients with von Willebrand's disease is not very extensive. This disease is the second most common cause of inherited bleeding disorders in our hospital but the association of this disease with pregnancy is not very common. There are few reported cases in English literature (Walker E.H. 1968, Evans P.C. 1971, Noller K.L. 1973, Krishnamurthi 1977, Punnonen R. 1981).

Cryoprecipitate is the treatment of choice for von Willebrand's disease because it contains all forms of Factor VIII macromolecular complex (Cal dwell D.C. 1985). Patients with this disease should be investigated at regular intervals during pregnancy and if Factor VIII: C levels remain low and infusion of cryoprecipitate or fresh frozen plasma should be given at the onset of labour and continued for 4-5 days postpartum to maintain the Factor VIII: C level in the blood at 50% or above. Trauma during delivery should be minimised. Caesarean section can be undertaken if indicated, provided the Factor VIII: C level is normal. Replacement of this factor is advisable to cover surgery and until healing of the scar is established. (Krishnamurthi 1977).

In the two cases that we are reporting the management was different. Cryoprecipitate was not available for the first patient. Fresh blood and plasma infusions given during delivery were not sufficient to bring up the F VIII: C to haemostatic levels and she had PPH and bleeding from the episiotomy, thereby increasing the morbidity.

In the second patient, infusion of adequate amounts of lyophilised cryoprecipitate prior to surgery ensured adequate haemostasis during caesarean section. Continued infusion for the next nine days prevente PPH and secondary haemorrhage from the suture line. Antiplatelets drugs like aspirin and intramuscular injections were also avoided.

Pregnant women with vWD would be managed in a tertiary care hospital with access to adequate laboratory facilities. Although the bleeding time is prolonged in the presence of a normal platelet count the clotting time is an insensitive test and may be normal as in these two cases. It is important to have an activated partial thromboplastin time test available to make the diagnosis. Cryoprecipitate or Factor VIII: C concentrates should be used as a matter of choice. Much larger volumes of fresh frozen plasma have to be infused to attain equivalent levels of F. VIII: C activity.

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