• CASE REPORT •

Fetal Surface Placental Hematomas: the Clue in the Diagnosis of a Bleeding Dyscrasia

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Von Willebrand disease (vWD) is the most common inherited bleeding disorder in humans, occurring in about 1% of women and caused by a deficiency or abnormality in the von Willebrand factor (vWF). There are different types of vWD. Type I comprises approximately 80% of the cases, its inheritance is autosomal dominant. Women with vWD have a 10-fold risk of having antepartum bleeding when compared to the general population. We report a case of von Willebrand disease diagnosed due to findings on a routine ultrasound evaluation. [*P R Health Sci J 2013;3:154-156*]

Key words: von Willebrand Disease, Placental hematomas

Von Willebrand disease (vWD) is the most common inherited bleeding disorder in humans, occurring in about 1% of women and caused by a deficiency or abnormality in the von Willebrand factor (vWF) (1). von Willebrand factor is a protein required for the normal adhesion of platelets to the site of injured endothelium and for preservation of factor VIII in the circulation. There are different types of vWD. Type I comprises approximately 80% of the cases, its inheritance is autosomal dominant. Type II is the second more common form and is inherited in an autosomal recessive fashion. Type III is the least common form of vWD and it is also autosomal recessive (1, 2). Women with vWD have a 10-fold risk of having antepartum bleeding when compared to the general population. Although there is an increased risk of preterm delivery and stillbirth due to antepartum hemorrhage, the risk for premature labor and intrauterine fetal demise are not increased (3). Women develop normal levels of factor VIII coagulant activity and vWF antigen during pregnancy although bleeding time may remain prolonged. Treatment is recommended if factor VIII activity is very low or in the presence of active bleeding (2, 4). We report a case of vWD diagnosed due to findings on a routine ultrasound evaluation. The report was approved by the ethics committee of our center.

Case report

A 24 year-old patient (G2P0010) came to our perinatology practice at 33 2/7 weeks for a follow up ultrasound for the evaluation of the fetal growth. A previous ultrasound performed at 20 weeks for anatomical survey was unremarkable. In general, the ultrasound at 33 2/7 weeks showed a normally developed fetus with an estimated fetal weight (EFW) at the 66th percentile and the amniotic fluid volume expressed as an amniotic fluid index (AFI) was normal with a value of 15 cm (normal values are between 9 and 25 cm). An irregularly shaped structure within the placental tissue measuring approximately 11 x 9 x 7 cm and which contained echogenic fluid was noticed (Figure 1). No blood flow could be identified within the mass using color power angio. It was suspected to be blood. The liquid nature of this mass suggested it to be in part a venous lake. A concern was that the nature of this lesion could have represented acute bleeding.

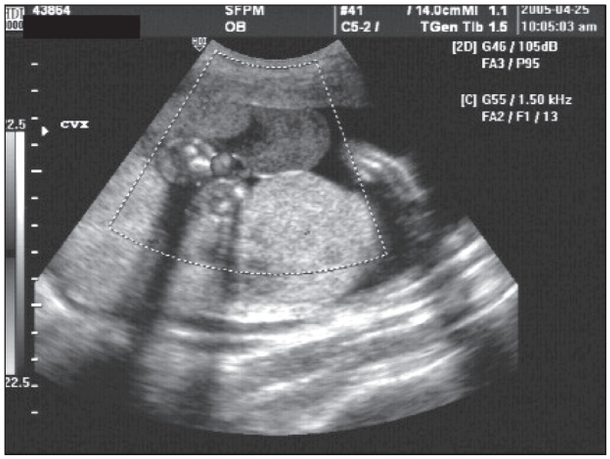

Figure 1. Large hematomas in the anterior and posterior margins of the fundal placenta. Evidence of absence of blood flow within the placental masses.

The patient was admitted to labor and delivery for observation and a complete blood count (CBC). An ultrasound follow up the next day showed that the lesion on the fetal surface of the placenta was stable in size measuring 11 x 10 x 9 cm. The appearance was almost the same, this time showing fluid-fluid level (Figure 2) which confirmed the diagnosis of a hematoma.

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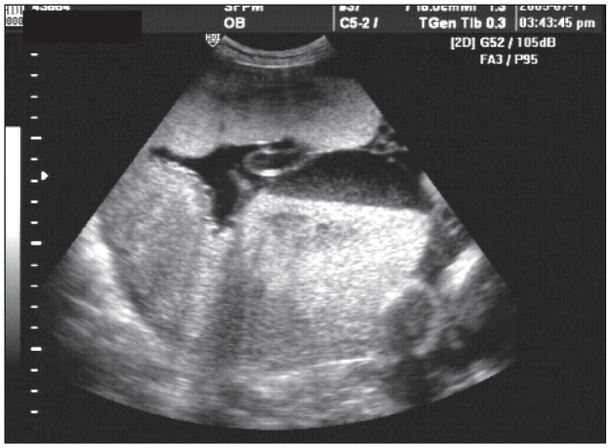

Figure 2. Fluid-fluid level in a large placental hematoma

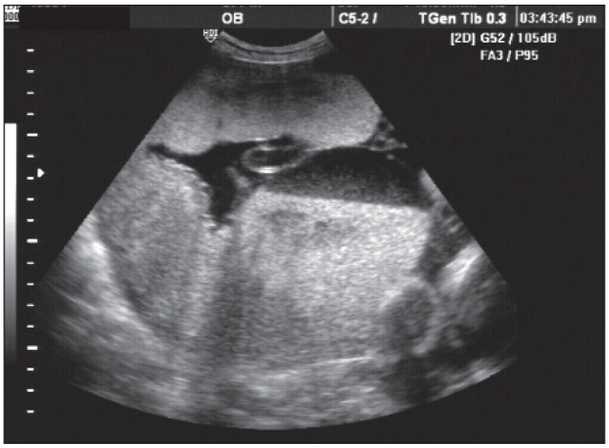

Figure 3. Large intra-placental hematoma with fluid-fluid level.

The peak systolic velocity of the middle cerebral artery was within the normal range consistent with the absence of fetal anemia. The biophysical profile (BPP) was 8/8 and the amniotic fluid volume (AFV) was still normal (AFI=12 cm). The coagulation studies were reported as normal (PT, PTT, fibrinogen, CBC with platelets).

The lesion remained stable the day after so the patient was discharged with plans for twice weekly BPP tests with and reevaluate the timing of delivery as pregnancy progressed.

At 34 6/7 weeks the previously described lesion remained stable but a new hemorrhagic lesion was detected measuring 8.1 x 4.9 x 3.7 cm (Figure 3). In view of the new lesion and the gestational age, the risks and benefits of delivery were discussed. Delivery was recommended. In view of her prior history of bleeding and need for blood transfusion after a termination of pregnancy (TOP), the risk of bleeding during and after cesarean section were discussed. The patient was readmitted to the labor and delivery unit for surveillance. Before the cesarean section she underwent a complete coagulation study which included platelet function which was found to be abnormal. The coagulation panel included: Normal PT (11 sec) and PTT (28 sec), normal platelet count (180,000), abnormal platelet function tests (PFA-100®), prolonged bleeding time (10 min), low vWF:RCo (26 IU/dl), low vWF:Ag (24 IU/dl), and normal Factor VIII (112.3 IU/dl). Hematology consultation was obtained for the administration of desmopressin acetate (DDAVP) for correction of her platelet function after which she underwent an uncomplicated primary cesarean section. A baby boy was delivered in frank breech presentation with Apgar score of 9 and 10. Postoperatively, the hematologist recommended a blood transfusion as her hemoglobin and hematocrit was 21.9 % and 7.2 g/dl, respectively.

At the time of her discharge she was in stable condition without evidence of bleeding. She continued to be followed as an outpatient by hematology.

Discussion

Women with known Von Willebrand disease during pregnancy should be informed of the risks of bleeding complications. They should have a close follow up by a hematologist and a high risk pregnancy specialist during and after pregnancy. Because of the risk of inheritance of vWD, they should meet with a genetic counselor too.

Clotting factors and levels of vWF usually stabilize during pregnancy, so most of the time no hemostatic therapy is needed during pregnancy and delivery. Levels of clotting factor VIII and vWF should be monitored close to delivery to prevent bleeding. Most of the bleeding complications occur in the postpartum period.

Our case was very uncommon in its presentation with marginal sinus and intraplacental hematomas. Intraplacental and fetal surface hematomas are rare occurrences and there are just a few reported in the literature (5). Most of the solid placental masses described in the literature (6) are chorioangiomas and the main difference that those masses have with the placental hematomas is that they show abundant blood circulation inside the mass and the hematomas do not.

Despite normal vWF levels her platelet function was abnormal and most probably related to the intraplacental bleeding. The Platelet Function Assay (PFA) was elevated for both the collagen and epinephrine (Col/Epi) and the collagen and adenosindiphosphate (Col/ADP). Once anemia and thrombocytopenia have been excluded, further evaluation to exclude von Willebrand disease and inherited/acquired platelet dysfunction such as renal failure storage pool disease, release defect, Bernard-Soulier disease, and Glanzmann thromboasthenia should be considered (7). Patients with vWD do not have a higher incidence of placental abruption than the general population. Most of the statistics regarding the effects of the disease during pregnancy refer to patients with a known diagnosis of vWD that have been under specialist supervision

which could be underestimating the risks. In our case, other than a story of an important bleeding after a previous miscarriage, the patient never had any other bleeding episode or investigation of bleeding disorders. She did require transfusions after her c-section due to decreased hematocrit despite normal clotting testing as well as DDAVP administration to correct her platelet function but the bleeding during the procedure was not more than normal.

The patient was further investigated after delivery and the diagnosis of von Willebrand's disease type I was confirmed.

Conclusion

Placental changes on ultrasound compatible with fetal surface hematomas, in the absence of signs of abruption, can be the first sign to suggest a bleeding dyscrasia. A massive hematoma with and otherwise stable fetus can only be of maternal origin.

Resumen

La enfermedad de von Willebrand es el desorden hematológico hereditario más común en los humanos, ocurriendo en aproximadamente 1% de las mujeres y es causado por una deficiencia o anormalidad en el factor von Willebrand. Hay varios tipos de enfermedad de von Willebrand. El tipo I comprende aproximadamente el 80% de los casos y su patrón hereditario es autosómico dominante. Las mujeres con enfermedad de von Willebrand tienen un riesgo 10 veces mayor de tener sangrados anteparto en comparación con la población en general. Nosotros presentamos un caso de enfermedad de von Willebrand diagnosticado por los hallazgos en un examen de ultrasonido de rutina.

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