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POSTER ABSTRACTS

311.DISORDERS OF PLATELET NUMBER OR FUNCTION: CLINICAL AND EPIDEMIOLOGICAL

Pregnancy and Delivery Outcome Among Ladies with Glanzmann Thrombasthenia: A Report from Glanzmann Thrombasthenia Registry of Al-Madinah, Saudi Arabia

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Introduction

Glanzmann's thrombasthenia (GT) is an autosomal recessive bleeding disorder due to functional platelet defects impairing platelet aggregation. AlMadinah has reported the highest prevalence of GT globally, with a prevalence of 1:10,000. Pregnancy had been reported among ladies with GT with different outcomes. As a bleeding disorder, Pregnancy among ladies with GT carries a higher risk for bleeding either during pregnancy or intra and postpartum. Neonates have an increased risk for hemorrhage, either due to alloimmunization or they may carry the gene of GT. Many GT ladies got pregnant earlier when GT diagnosis was difficult, and they did not know their diagnosis; even after the diagnosis was possible, ceasing the menstrual cycle was not culturally acceptable. In this study, we are describing the pregnancies among GT ladies and their outcomes. Methods

This is a report from the AlMadinah GT registry in the Madinah Hemophilia Comprehensive Care Center. The registry covers a period of 20 years and includes all patients with a confirmed diagnosis of GT. 136 GT patients were identified. 11 patients had no data available. A spreadsheet designed to collect detailed data about pregnancy, delivery, and neonates. Electronic and hard copy medical records have been accessed to collect data. Data was analyzed by the statistical package of social sciences (SPSS) version 23. The research and ethical committee approved the study. Results

Twenty-eight pregnancies have been identified. The median age of mothers at the time of pregnancy was 30 ± 5.7 years (20 - 42 years). Mothers' Gravidity was 1-7 (median 2.5 \pm 1.6), and parity was 1-6 (median 2 \pm 1.4). Five miscarriages (18%) identified occurred at the 17th, 11th, 13th and 15th gestational weeks. In contrast, 5th one is not on record. Three miscarriages took place after moderate to severe vaginal bleeding. Mild to severe post-abortion bleeding was controlled with platelets transfusion, activated human recombinant factor 7 (rFVIIa), and tranexamic acid. Thirty-two bleeding episodes occurred during 16 pregnancies, with 1-5 bleeding episodes per pregnancy (Median of 2 episodes). Seven (22%) bleeding episodes were vaginal bleeds, while 25 (78%) were GT-related bleeds such as gum bleeding or epistaxis. Two patients must undergo plasma exchange and be on steroids due to alloimmunization development during pregnancy. Delivery occurred at the gestational age of 34-40 weeks (median 38 ± 2 weeks). Sixteen (70%) deliveries were vaginal, and 7(30%) were cesarean sections. The indications for cesarean sections were family request in 4 cases, fetal distress in two instances, and precautional section in one case. Prophylactic platelets transfusion had given pre-delivery on four occasions, rFVIIa on 4, and a combination of rFVIIa and platelets transfusion on six occasions. Tranexamic acid was used in all cases. Ten deliveries were conducted with no prophylaxis. Post-partum hemorrhage (PPH) occurred in 18 (64%) cases, 13 (72%) cases of primary PPH, and 5 (28%) cases of secondary PPH. We could not find a correlation between pre-delivery prophylaxes and PPH (P=0.2). PPH had controlled successfully with platelets transfusion and rFVIIa though red blood cell transfusion was needed on eight occasions. Twentyeight pregnancies produced 24 babies (13 girls and 11 boys). Four babies had bleeding (2 intracranial hemorrhages, one subdural hematoma, and one gum oozing), and 9 had alloimmunization with mild to severe thrombocytopenia. Of all babies tested for GT, 6 of them were GT.

Conclusions

Pregnancy among ladies with GT carries a higher risk of bleeding. Management of pregnancy in women with GT has to be in a Multidisciplinary approach. The high rate of PPH among GT ladies makes it essential to treat it promptly. Pregnancy among ladies with GT should be studied in multi-centers sitting to learn more about clinical courses and management.

Disclosures Tarawah: Novonordisk, Pfizer, Novartis, Takeda: Consultancy, Honoraria, Membership on an entity's Board of Directors or advisory committees, Speakers Bureau.

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