CASE REPORT

Ovarian Hematoma and Hemoperitoneum in Glanzmann's Thrombasthenia

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Abstract

Aim: This case report is aimed to study the treatment approach in a case of Glanzmann's thrombasthenia (GT) with ovarian hematoma and hemoperitoneum.

Background: Glanzmann's thrombasthenia is an extremely rare life-threatening bleeding disorder that often presents with heavy menstrual bleeding in reproductive-age females.

Case description: A 25-year-old patient presented to emergency with abdominal pain and heavy menstrual bleeding and ultrasonography suggestive of ovarian hematoma and hemoperitoneum. The patient was managed with blood and platelet transfusion and a conservative approach was adopted. Combined oral contraceptive pills were started for heavy menstrual bleeding.

Conclusion: Management of GT is a challenge. Being an autosomal recessive disorder besides menstrual blood loss control, partner testing, and preconceptional and premarital counseling is critical.

Clinical significance: A rarity of diseases poses a problem with no standardized approach and treatment options are mostly reliant on case studies or expert opinions.

Keywords: Autosomal recessive, Bleeding disorder, Glanzmann's thrombasthenia, Hemoperitoneum, Life-threatening, Ovarian hematoma, Rare.

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Introduction

Glanzmann's thrombasthenia is a genetic bleeding disorder with an autosomal recessive pattern of inheritance. The incidence of GT is reported to be 1 in 10,00,000.[1] It is caused by a deficiency of the platelet integrin αIIbβ3 (the platelet fibrinogen receptor). Glanzmann's thrombasthenia is found to be more prevalent in ethnicities where consanguinity is frequent, as seen in Southern Indian/Jewish communities.[1] Reproductive age-group females with GT often deal with heavy menstrual bleeding, which can be life-threatening requiring multiple hospital admissions and has been linked to poor quality of life. It is challenging to manage due to the rarity of the disease with no standard treatment protocols. The treatment options are largely established on available case reports or literature or expert opinion to date.

Case Description

A 25-year-old young unmarried female, with a diagnosed case of GT, presented to an emergency department with complaints of pain in the abdomen for 2 weeks and heavy menstrual bleeding from a remote district. She came to our institute as her hometown has few superspecialty clinics and she was treated here in the past for menorrhagia. The pain was more in the lower abdomen, with continuous dull aching in nature. The patient was diagnosed to have GT at 7 years of age, with a history of occasional epistaxis and bruising. After menarche at 12 years, she had severe polymenorrhagia and was referred to this institute. She had been started on tranexamic acid and subsequently discharged on cyclical progesterone. She was then lost to follow-up but had multiple hospital admissions in her hometown. Her elder sister was a known case of GT and had succumbed to excessive bleeding.

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On examination, the patient had a pulse of 110 per minute, and a blood pressure of 110/70 mm Hg. On abdominal palpation, there was gross distension and tenderness. The internal examination was deferred. Her Hb was 7.4 gm%, White blood count (WBC): 8800/cmm, platelets: 2.8 lakhs, Prothrombin time-International Normalized ratio (PT/INR): 20.3/1.7. Contrast-enhanced CT scan done before admission reported a solid cystic mass, suspicious of ovarian malignancy. Both ovaries and adnexa were seen separately. Ultrasonography repeated in our institute was suggestive of left ovarian hematoma. Magnetic resonance imaging (MRI) abdomen was suggestive of hematometra and hematosalpinx with large periuterine hematoma and hemoperitoneum. Tumor markers were as follows: *S. AFP (serum α-fetoprotein): 1.49, *Sr. CEA (serum carcinoembryonic antigen): <1.0, *Sr BHCG (serum β-human chorionic gonadotropin): <0.13, *Sr. CA 19.9 (serum cancer antigen 19-9): <2.0, *S.LDH (serum lactate dehydrogenase): 602, *S. CA 125 (serum cancer antigen 125): 179.2.

The patient has transfused 8 units of platelets and 2 units of packed cells after a hematology opinion. With MRI findings of hematoma, conservative management was chosen over surgical intervention. The patient was started on continuous combined oral contraceptive (COC) pills and tranexamic acid for her heavy menstrual bleeding to which the patient responded well and improved symptomatically and was discharged on COCs with emphasis on the need for follow-up and premarital counseling. A follow-up scan after 2 months showed lysing hematoma, and the patient was amenorrheic on COCs and improved symptomatically. A further plan of action is to continue oral contraceptive pills with substitution to hormonal intrauterine device (IUD) in near future to prevent further bleeding.

Discussion

In women with GT, heavy bleeding could be a serious problem with an increased risk of acute and protracted bleeding requiring multiple transfusions. Heavy menstrual bleeding along with anemia, the need for frequent hospitalizations, also affect the quality of life and psychological well-being of the woman.[2] Most patients miss work, school, and social activities because of intractable bleeding.[2] Risk of intra- and postpartum hemorrhage is increased during pregnancy.[3] In women with GT, the prevalence of postpartum hemorrhage (PPH) is reported to be 34%. Due to the intraplacental transfer of maternal antiplatelet antibodies against fetal thrombocytes, neonates have more risk of bleeding. There is high maternal and neonatal morbidity and mortality.[3] The standard therapy for nonsurgical episodes of bleeding is platelet transfusion.[1] Other treatment modalities are tranexamic acid, hormonal-based treatment like COC pills or progesterone—only pills and Levonorgestrel-Intrauterine device (LNG-IUD), recombinant FVIIa. Patients receiving platelet transfusions can develop platelet alloimmunization or/and refractoriness.[1] Approximately 17% of GT patients have been known to develop antibodies against human leukocyte antigens (HLAs) after transfusions. This creates a challenge as most patients will need multiple transfusions in their lifetime. In our patient, hemoperitoneum generally a surgical emergency and requires emergency exploration was a concern. A participatory decision of conservative management was taken with close monitoring. A combination of tranexamic acid, blood, and platelet transfusion and COCs after stabilization worked well.[4,5]

Conclusion

Management of GT in reproductive age poses a challenge. Being an autosomal recessive disorder, besides menstrual blood loss control, premarital and preconceptional counseling and partner testing are critical steps in such patients. Due to the rareness of the disorder, there is no standardized approach toward the management of heavy menstrual bleeding among adolescents with GT.

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References

1. King LJ, Huff J, Heber D, et al. Management of refractory menstrual bleeding in an adolescent with Glanzmann thrombasthenia: A case report and review. Case Rep Obstet Gynecol 2020;2020:8848763. DOI: 10.1155/2020/8848763.
2. Rajpurkar M, O'Brien SH, Haamid FW, et al. Heavy menstrual bleeding as a common presenting symptom of rare platelet disorders: Illustrative case examples. J Pediatr Adolesc Gynecol 2016;29(6): 537–541. DOI: 10.1016/j.jpag.2016.02.002.
3. Punt MC, Schuitema PCE, Bloemenkamp KWM, et al. Menstrual and obstetrical bleeding in women with inherited platelet receptor defects—A systematic review. Haemophilia 2020;26:216–227. DOI: https://doi.org/10.1111/hae.13927.
4. Dhar H, Santosh A. Glanzmann's thrombasthenia: A review of literature. J South Asian Feder Obs Gynae 2019;11(2):134–137. DOI: 10.5005/jp-journals-10006-1665.
5. Shrivastava D, Joshi S, Chella H. Study of puberty menorrhagia in adolescent girl in a rural set-up. J South Asian Feder Obs Gynae 2012;4(2):110–112. DOI: 10.5005/jp-journals-10006-1187.