WORLD JOURNAL OF PHARMACY AND PHARMACEUTICAL SCIENCES
SJIF Impact Factor 7.421
Volume 7, Issue 11, 1381-1384 Case Report ISSN 2278 – 4357

TYPE II GLANZMANN THROMBASTHENIA: CASE REPORT

Elizabeth Wilson Baby*[1], Aleena Prakash[1], Akbar Sharooque V.[1], Shalin Elsy Varghese[1], Apollo James[2] and T. Sivakumar[3]

[1]Pharm D. Interns, Department of Pharmacy Practice, Nandha College of Pharmacy, Erode, Tamilnadu.

[2]Asst. Professor, Department of Pharmacy Practice, Nandha College of Pharmacy, Erode, Tamilnadu.

[3]Principal, Nandha College of Pharmacy Practice, Erode, Tamilnadu.

Article Received on
07 Sept. 2018,

Revised on 27 Sept. 2018,
Accepted on 17 October 2018

DOI: 10.20959/wjpps201811-12637

***Corresponding Author**
Elizabeth Wilson Baby
Pharm D. Interns,
Department of Pharmacy Practice, Nandha College of Pharmacy, Erode, Tamilnadu.

ABSTRACT

Glanzmann thrombasthenia is a rare autosomal recessive bleeding disorder due to the absence of glycoprotein IIb/IIIa. Prolonged untreated or unsuccessfully treated hemorrhages are life threatening. It is estimated to affect one in one million individuals in worldwide. The symptoms of Glanzmann thrombasthenia usually begin at birth or shortly thereafter. Purpura, epistaxis, gingival hemorrhage, and menorrhagia are main features and gastrointestinal bleeding and hematuria are less common. Women with GT often also have unusually heavy menstrual bleeding, irregular uterine bleeding, and excess bleeding in childbirth. We present a case of 17 year old female patient was presented with the heavy menstrual bleeding for ten days. The patient had abdominal pain, history of giddiness easy fatigability and loss of weight. Based on bleeding time and flow cytometry. Patient was diagnosed as Type II Glanzmann thrombasthenia. The management done for the patient was blood transfusion and antifibrinolytic. Immediate control of bleeding and found good response.

KEYWORDS: Glanzmann thrombasthenia, Platelet function analyzer, Light transmission aggregometry.

INTRODUCTION

Glanzmann thrombasthenia (GT) is a rare autosomal recessive bleeding syndrome and it first identified by, Dr.Eduard glanzmann's in 1918, from a village children in the Swiss Alps.[1,2]

It affect the platelet function disorder that mainly caused by abnormality in the genes for glycoproteins IIb/IIIa. It affect both males and females. The autosomal recessive means that both parents must carry an abnormal gene (even though the parents don't have the disease) and pass that abnormal gene on to their child. Type 1 disease were patient with absent in platelet aggregation and absent clot retraction but Type 2 were those with absent aggregation and excess clot retraction.[3] GT is more common in marriage between blood relatives. The epidemiologic data shows it more common in Jordan, India and Saudi Arabia and among the Iraqi-Jews and Israel, as compared to other parts of the world.[4]

CASE REPORT

A 17 year old female patient was admitted to the hospital with the complaints of heavy menstrual bleeding since ten days, not associated with abdominal pain but the patient have use 5-6pads/day. The patient has history of giddiness, easy fatiguability, loss of weight. Menstrual bleeding in each episode was 100-150ml. From the general examination pulse is 96bpm, blood pressure is 100/70mmHg, and through systemic examination CVS-S1S2(+), RS-BAE(+), CNS-normal, abdomen inspection found that not distended, umbilicus inverted. The laboratory parameters revealed that hemoglobin count (3.48g/dl), red blood cell ($2.09*10^{12}$) platelet count ($350*10^{9}$/L), Mean cell Hemoglobin(16.2pg), MCHC(26.7g/dl) are lower and Bleeding time of patient is longer than normal. The patient was prescribed with injection vitamin k IV twice days, injection tranexamic acid 500mg IV twice. Blood transfusion is also done. syrup. zincofer, capsule. Livogen XT was prescribed since the patient having anemia. From the fourth day onwards patient started improving and on the sixth day patient was discharged.

DISCUSSION

Glanzmann thrombasthenia is rare disease it affects equally in both male and female. The symptoms of this disorder are usually detectable at the birth or during infancy. The more frequently affected condition of this population is intermarriage within a group (consanguinity). GT is the abnormality in the genes of glycoprotein IIb/IIIa carried on chromosome 17 of DNA.[7] A normal number of platelets but have a prolonged bleeding time, is mostly occur in GT affected individuals. The GT is diagnosed by, highly sensitive Platelet function analyzer (PFA), Light transmission aggregometry (LTA) which is the golden standard diagnostic tool for assessing platelet function, Flow cytometry, which is beneficial for Glycoprotein receptor deficiency and/or dysfunction and Mutation analysis.[8]

Management for patient with GT don't need therapy on regular basis, but during the surgical procedure require treatment for controlling bleeding after injury, and during spontaneous bleeding episodes. Blood platelet transfusions are effective in some patient with GT. In affected individuals, if necessary transfusion can be continued throughout their life.[5] Novo Seven RT, a recombinant factor VIIa product, was approved in 2014. This medication is mainly indicated to treat bleeding episodes and peri operative management, when platelet transfusions are not effective. Nasal packing or application of gel foam soaked in thrombin was the management of epistaxis and gingival hemorrhage. Regular dental care is essential for prevent bleeding from the gums. Hormonal therapy can be done to suppress the menstrual periods. Antifibrinolytic agents are useful for the symptomatic and supportive treatment of GT. In this case, the patient with glanzmann thrombasthenia is diagnosed by blood test and increased bleeding time, prothrombin time. In the initial therapy patient was managed with blood transfusion, which was very effective for the patient. And later started with antifibrinolytic inj. tranexamic acid 500mg which competitively inhibit the action of plasminogen to plasmin. Vitamin k that is a lipid co factor has been prescribed for getting normal blood clotting.

CONCLUSION

For a successful treatment a thorough medical history and hematological consult are compulsory along with adequate precautions. To prevent further complications follow post treatment instructions and undergo frequent patient education.

ACKNOWLEDGEMENT

We express heartfelt gratitude to the faculties and our friends for providing their immense support.

REFERENCES

1. Bellucci S, Damaj G, Boval B. Bone marrow transplantation in severe Glanzmann's thrombasthenia with antiplatelet alloimmunization. Bone Marrow Transplantation, 2000; 25: 32730.
2. Ajit D Dinkar, Sujata K Satoskar, Amit Gothwal Glanzmann's Thrombasthenia: A Case Report and Review July 2011:10.5005/jp-journals-10011-1179.
3. Alan T Nurden, Glanzmann thrombasthenia: Orphanet Journal of Rare Diseases, 2006; 1: 10.

4. Sahida K., Al-Barghouthi, MD, Abdullah Al-Orthman, JMC(Ortho), Ameer Lardhi, Glanzmann's thrombasthenia-spectrum of clinical presentation on Saudi patients in the eastern province J Family community Med., 1997 Jan- Jun; 4(1): 57-61.
5. Meganathan Kannan, PhD, and Renu Saxena, MD, Glanzmann's Thrombasthenia: An Overview: Clinical and Applied Thrombosis/Hemostasis, April 2009; 15(2): 152-165.
6. R. E. Scharf, M. M. Rahman, H. Seidel, The impact and management of acquired platelet dysfunction: Hämostaseologie, 2011; 31: 28–40.
7. Srivastava A, Usher S, Nelson EJ, Jayandharan G, Shaji RV, Chandy M, Seligsohn U, Peretz H,; Prenatal diagnosis of Glanzmann thrombasthenia, 2003 Jul-Aug; 16(4): 207-8.
8. Tia Solh, Ashley Botsford, Melhem Solh; Glanzmann's thrombasthenia: pathogenesis, diagnosis, and current and emerging treatment options: journal of blood medicine, July 2015.
9. Meganathan Kannan, MSc,1 Firdos Ahmad, MSc,1 Birendra Kumar Yadav, MSc, Comparison of Flow Cytometry and Western Blot With Respect to DNA Mutation; Am J Clin Pathol, 2008; 130: 93-98.