Available online on 15.06.2024 at http://jddtonline.info

Journal of Drug Delivery and Therapeutics

Open Access to Pharmaceutical and Medical Research

Open Access Full Text Article

Check for updates

Case Report

Glanzmann's Thrombocytopenia: A Case Report

*[1] Preeti Tiwari, [1] Manav Singh, [2] Dr. S.P. Nayak, [3] Dr. G S Chakraborthy

[1] *PharmD scholar, Department of Pharmacy Practice, Parul Institute of Pharmacy & Research, Parul University, Vadodara, Gujarat.*

[2] *Assistant Professor, Department of Pharmacy Practice, Parul Institute of Pharmacy & Research, Parul University, Vadodara, Gujarat.*

[3] *Principal, Parul Institute of Pharmacy & Research, Parul University, Vadodara, Gujarat.*

Article Info:

Article History:

Received 03 April 2024
Reviewed 09 May 2024
Accepted 28 May 2024
Published 15 June 2024

Cite this article as:

Tiwari P, Sing Mh, Nayak SP, Chakraborthy GS, Glanzmann's Thrombocytopenia: A Case Report, Journal of Drug Delivery and Therapeutics. 2024; 14(6):1-2

DOI: http://dx.doi.org/10.22270/jddt.v14i6.6654

*Address for Correspondence:

Preeti Tiwari, PharmD scholar, Department of Pharmacy Practice, Parul Institute of Pharmacy & Research, Parul University, Vadodara, Gujarat.

Abstract

Glanzmann's thrombocytopenia is a platelet disorder in which platelets have qualitative or quantitative deficiencies of the fibrinogen receptor α2bβ3. Most cases are hereditary but can be acquired also. This disorder in named after Dr.Eduard Glanzmann, who first described it in 1918. Its global prevalence is estimated to be one in a million with higher prevalence rate in population where consanguinity marriage is common. It has good prognosis with proper supportive care.

Case Summary: A 17-year male patient had come with complaint of severe epistaxis randomly for hours in a known case of Glanzmann's thrombocytopenia which was diagnosed 10 years ago. With epistaxis he also had complaints of weakness in upper limbs and gastrocnemius muscle

Diagnosis: Glanzmann's thrombocytopenia

Treatment: Inj. Etamsylate 500mg BD, Inj. Hemocoagulase 1IU OD, Inj. Tranexamic acid 500mg OD, Tab. Vitamin C BD, Tab. Calcium gluconate 500mg BD, Syp. Vitamin D3 and was transfused with PCV (A+) total 12 pint (200ml) and platelets 2 pint (200ml)

Keywords: Glanzmann's thrombocytopenia, Epistaxis, Tranexamic acid, Congenital, Platelet Disorder.

INTRODUCTION

Glanzmann's thrombocytopenia is a rare blood disorder that's expressed by long time and unprompted blood loss that starts occurring from the instant person is born. Person with Glanzmann's thrombocytopenia happens to have bruises easily, have expected epistaxis, and also bleeding from the gums. They have frequent violet or red spots on their skin because of bleeding under the skin or swelling that happens due to bleeding within tissues. Glanzmann's thrombocytopenia is also responsible for delayed blood loss due to injury, surgery, or trauma. Female born with this disorder may also have delayed and often abnormally heavy menstrual flow. Suffered female also have high chances of extreme blood loss during pregnancy and accouchement.

Glanzmann's thrombocytopenia was first documented in 1918 by Dr. Eduard Glanzmann, who described it as a novel platelet disorder that have defective clot retractions and atypical appearances on stained film. It has since been more specifically described as an autosomal recessive genetic disorder in which the platelet count is normal or subnormal, the bleeding time is prolonged, and platelet aggregation is deficient or absent. Due to this deficiency of platelet function it manifests as a bleeding disorder characterized by mucocutaneous haemorrhage of varying severity.

Glanzamann's thrombocytopenia is a rare disorder but is more likely to occur in consanguineous population where intermarriage is common like Iraqi Jews, French gypsies and Jordanian nomadic tribes. It is a non-curable disorder and patient suffering from this disorder have to take medication for preventing excessive blood loss from birth till death. Most cases of this disorder are hereditary but acquired Glanzamann's thrombocytopenia also occurs due to an autoantibody against the platelet fibrinogen receptors. Confirmatory diagnostic test for this disorder if flow cytometry and presence of monoclonal antibodies.

CASE REPORT

A 17-year male patient had come with complaint of severe epistaxis randomly for hours in a known case of Glanzmann's thrombocytopenia which was diagnosed 10 years ago. With epistaxis he also had complaints of weakness in upper limbs and gastrocnemius muscle. Earlier when he had nose bleeding for it as a preventing measure, he plugs cotton in his nose but it causes him difficulty in speaking. In past also he had major complaints of epistaxis and it was more frequent in summer as compare to winter for which sometime he was transfused with blood. The patient is also underweighting with BMI of 16.2 and his diet is also imbalanced. His parents were not having a consanguineous marriage which is most in other cases of Glanzmann's thrombocytopenia but his mother is suffering from epilepsy and father is suffering from hemiplegia (Right side) due to polio. Patient vitals were normal but his laboratory findings show that his haemoglobin levels (7.2g/dl), RBC

(4.15), PCV (25.73%), MCV (61.93fl), MCHC (28.93g/dl), MCH (17.92pg) and platelets level (1.31lac) were low whereas his RDW (19.04%) levels was high, activated partial thromboplastin Time – 35.3sec (test) and 32.6sec (control), and prothrombin time – 12.8sec (control) and INR was 0.99. other liver function test, cardiovascular and respiratory system function were normal. For management of the disorder, he was given Inj. Etamsylate 500mg BD, Inj. Hemocoagulase 1IU OD, Inj. Tranexamic acid 500mg OD, Tab. Vitamin C BD, Tab. Calcium gluconate 500mg BD, Syp. Vitamin D3 and was transfused with PCV (A+) total 12 pint (200ml) and platelets 2 pint (200ml) in 5 days of admission. He was advised for bed rest and to not remove cotton plug but it was making difficulty in speaking for him. He was advising to come for follow up after 7 days and in case of any injury or cut on skin contact physician and rush to nearby hospital.

DISCUSSION

Glanzmann's is a rare platelet disorder which is caused by deficiency of the platelet integrin α2b β3. This integrin is the platelet fibrinogen receptor and thus is essential for platelet aggregation and haemostasis. Person suffering from this disorder have lifelong problem of mucosal bleeding, gingival bleeding, menorrhagia and gastrointestinal bleeding. It is quite rare as it occurs one in million globally and is more common in population that have increased consanguinity. It may be present from birth but mostly is diagnosed later in childhood and risk and prevalence of bleeding increases with age.

It can be diagnosed by two methods one is flow cytometry in which blood sample is placed in a suspension and injected into flow cytometer machine in which cells are counted and categorized, it is useful as it help in identification of markedly decreased CD41(GP2b) and CD61(GP2a) expression levels, which are diagnostic for Glanzmann's thrombocytopenia and the another test is monoclonal antibodies in which proteins similar as our body's antibody are inserted in sample to detect interaction with required proteins and help in diagnosis. Thus, these two diagnostic tests are confirmatory test to detect Glanzamann's thrombocytopenia.

Platelet transfusion is the standard treatment for this disorder but it difficult to adjust the dose as the count of platelets is normal so transfusion must be assessed functionally, by monitoring clinical reduction in bleeding. Patient should be also vaccinated against hepatitis due to infection risk associated with multiple transfusions. Tranexamic acid can also be used as it help in stopping as well as preventing bleeding. Women suffering from this disorder has risk of high blood loss during mensuration so they can be suggested to take oral contraceptive as to preventive measure for menorrhagia. Patients suffering from these disorders should be educated not to take anti-coagulants drugs like aspirin, clopidogrel, warfarin, heparin as well as NSAIDS. It being a rare and incurable disorder but has a good prognosis with appropriate supportive care.

CONCLUSION

Glanzmann's thrombocytopenia is congenital bleeding disorder caused by defect in integrin factor of platelet. Patient suffering from this disorder suffer from lifelong unexpected bleeding episodes. For management of this patient had to take platelet transfusion regularly as it has good prognosis when managed with appropriate supportive care and patient should be informed about not to take anti-coagulant drugs as well as before any surgery measures should be taken as having availability of platelet transfusion if uncontrolled bleeding occurs. Patient should avoid working with sharp objects and regularly visit hospital for follow up.

REFRENCES

1. Swathi J, Gowrishankar A, Jayakumar SA, Jain K. A rare case of bleeding disorder: Glanzmann's thrombasthenia. Ann Afr Med. 2017 Oct-Dec;16(4):196-198. https://doi.org/10.4103/aam.aam_59_16 PMid:29063905 PMCid:PMC5676411 https://medlineplus.gov/genetics/condition/glanzmann-thrombasthenia/#description
2. Sebastiano C, Bromberg M, Breen K, Hurford MT. Glanzmann's thrombasthenia: report of a case and review of the literature. Int J Clin Exp Pathol. 2010 Apr 25;3(4):443-7. PMID: 20490335; PMCID: PMC2872751.
3. Krause KA, Graham BC. Glanzmann Thrombasthenia. [Updated 2022 May 9]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022.
4. https://my.clevelandclinic.org/health/diagnostics/22086-flow-cytometry#:~:text=Flow%20cytometry%20is%20a%20laser,other%20fluids%20in%20your%20body.
5. https://emedicine.medscape.com/article/200311-overview?src=mbl_msp_android&ref=share