International Journal of Basic and Applied Medical Sciences ISSN: 2277-2103 (Online)
An Online International Journal Available at http://www.cibtech.org/jms.htm
2012 Vol. 2 (2) May-August pp.117-118/Kande et al.
Case Report

AN EXTREMELY UNUSUAL CASE OF GLANZMANS THROMBASTHENIA - CASE REPORT

Vidyadevi Kande (Kendre), Sunil S Holikar, Surabhi Chopra and *Sachin B Ingle

Department of Paediatrics, ENT and Pathology MIMSR Medical College Latur, Maharashtra
**Author for Correspondence*

A**BSTRACT**
It is an unusual bleeding disorder with Autosomal recessive inheritance pattern. In this condition platelets lack glycoprotein 2b3a .It is first diagnosed by Swiss paediatrician Glanzmann (1887-1959). Bleeding tendency varies from minimal bruising to fatal haemorrhages in Glanzmans thrombasthenia. Treatment includes platelets transfusion, recombinant factor VIIa and Haemopoietic stem cell transplantation

Key Words: *Glanzmann, 2b3a, platelets, Stem cell*

INTRODUCTION

It is an abnormality of platelets. It is very rare bleeding disorder in which platelets lack glycoprotein 2b 3a so that there is no fibrinogen bridging of platelets to other platelets hence bleeding time is increased. It is inherited as autosomal recessive manner (J. Paulscotland and Robert R. Montogmery. Nelson Textbook Of Pediatric,18th ed) Bleeding tendency in glanzmanns thrombasthenia is variable having minimal bruising to fatal haemorrhages .Treatment involves preventive as well as symptomatic bleeding episodes.

CASES

A 3yr old male patient brought by relatives in YCRH Latur on 3/5/12 at 12 noon in paediatric OPD with complaints of bleeding from nose,There was history of fall with swelling over left eyelid due to injury since night.Patient had similar episodes in the past with first episode at 6 months of age. Similar episodes were noted every 3-4 months with history of platelet transfusion each time. There was no history of any bleeding disorder in family. Antenatal history and birth history was uneventful. No history of icterus, cyanosis, fever and convulsions; and the baby was exclusively breast fed and no history of NICU admission in the postnatal period.The child was immunized as per schedule till date.On examination patient was conscious, well oriented with time, place and person, afebrile with vital parameters within normal limits. Swelling with ecchymosis and ptosis was present over left eyelid. Systemic examination was normal.

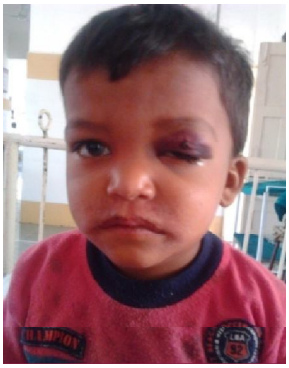

Figure 1: Swelling over left eyelid

Hematology Investigations revealed following findings: Blood group – A+, Hb – 4.5gm%,Platelets – 5,39,000. Reticulocyte count – 1% ,TLC – 14,200,DLC – N – 28%, L – 68%, E – 2%, M – 2%.,RBC

Morphology – moderate anisocytosis, microcytosis, poikilocytosis, moderate hypochromia,APTT – C – 30.4 sec, P – 27.9sec, PT – C- 12.6 sec, P – 13.8 s and Bleeding time - 25 sec which is prolonged. Investigated from national institute of immunohematology for Platelet Glycoprotein Receptor by flowcytometry revealed Gp Ib – 11.4%, Gp IIb IIIa – 0.1, Gp Iy - 89.4% and fibrinogen – 32.2% and finally the case was diagnosed as Glanzmann's Thrombasthenia and treated accordingly. Includes both preventive and supportive management was given to the patient.

Preventive Measures included proper dental hygiene which lessens gingival bleeding,

Avoidance of antiplatelet agents such as aspirin and other anti-inflammatory drugs such as (NSAIDS),

Iron and folic acid supplementations may be necessary if excessive bleedings causes anaemia

And hepatitis B vaccine

For the treatment of bleeding episodes platelet transfusions 4 units with packed cell transfusion 150 ml was given. After platelets transfusion, bleeding stopped & after 3rd to 4th day ecchymosis and swelling decreased.

DISCUSSION

Glanzmanns thrombasthenia is very rare bleeding disorder symptoms occurs in homozygous group (Malik, 1998; Vuckovic, 1996). Study shows it is associated with Hodgkins lymphoma, clinical features varies from mild hemorrhages to massive bleeding (Kitko, Hoffmann, Ghosh, 2009). Study shows formation of antiplatelet antibodies due to repeated transfusions of platelets. Study shows recombinant factor VIIa transfusion is the best way of treatment for qualitative platelet disorder. Some advised stem cell transplantation for severe bleeding.

REFERENCES

Degruchis (1990) Clinical Haematology in medical practice. Congenital qualitative platelet disorder 5th edition chapter **14** 397.

Ghosh K, Kulkarni B, Shetty S and Nair S (2009). Antiplatelet antibodies in cases of Glanzmann's thrombasthenia with and without a history of multiple platelet transfusion. *Indian Journal of Human Genetics* **15** 23-27.

Hoffman R, McGlave P and Shattil SJ, *et al.*, (2000). Hematology Basic Principles and Practice 3rd edition New York.

Paulscotland J, Robert R Montogmery and Churchill Livingstone NY (1999). Congenital Abnormalities of Platelet Function. *Nelson Textbook of Pediatric* 18th edition volume 2 Elsevier chapter 484.13 2088.

Kanman M, Saxena R (2OO9). Glanzmans thrombasthenia an overview clinical and applied thrombosis haemostasis. *Official Journal of International Academy of Clinical and Applied Thrombosis and Hemostasis* **15**(2) 156-165.

Robins and Cortan (2010). Pathologic Basis of Disease. 8th edition Elsevier chapter **14** 670.

Kitko CL, Levine JE, Matthews DC, Carpenter PA (2009). Successful unrelated donor cord blood transplantation for Glanzmann's thrombasthenia. *Pediatric Transplantion* **15** *1-5*

Malik U and Ducher (1998). Acquired Asthenia Glanzmans thrombasthenia associated with Hodgkins lymphoma a case report and review of literature cancer 1764-1768.

Poon MC (2007). The evidence for the use of recombinant human activated factor VII in the treatment of bleeding patients with quantitative and qualitative platelet disorders. *Transfusion Medicine Review* **21**(3) 223-236.

Vuckovic SA (1996). Glanzmann's thrombasthenia revisited. *Journal of Emergency Medicine* **14**(3)299-303.