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Research Paper

Oral Hemorrhage & Glanzmann's Thrombasthenia: A Case Report.

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ABSTRACT: *Glanzmann's thrombasthenia is an abnormality of the platelets. It is an extremely rare coagulopathy (bleeding disorder due to a blood abnormality), in which the platelets contain defective or low levels of glycoprotein IIb/IIIa (GpIIb/IIIa), which is a receptor for fibrinogen. As a result, no fibrinogen bridging of platelets to other platelets can occur, and the bleeding time is significantly prolonged. Here by we present a case report of teenage girl with the same condition presenting with a severe periodontal bleed which was managed medically. She was under medical supervision following a diagnosis of some bleeding disorder since the age of 4 years. With careful early diagnosis and proper supportive care Glanzmann's thrombasthenia has a very good prognosis. This article also provides a literature review on the same.*

KEY WORDS: *Glanzmann's thrombasthenia, Oral Bleeding, Hemorrhage, Periodontal Bleeding.*

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I. INTRODUCTION:

Glanzmann thrombasthenia is a platelet function disorder that is caused by an abnormality in the genes for glycoproteins IIb/IIIa. These genes code for a group of linked proteins normally found on the surface of platelets, the glycoprotein IIb/IIIa receptor (also called the fibrinogen receptor).[1] Because this receptor is absent or is not working properly, platelets do not stick to each other at the site of injury and it is difficult for the normal blood clot to form. Glanzmann thrombasthenia is an autosomal recessive disorder, meaning that both parents must carry an abnormal gene (even though they themselves don't have the disease) and pass that abnormal gene on to their child. Like all autosomal recessive disorders, it is found more frequently in areas of the world where marriage between close relatives is common. Glanzmann thrombasthenia affects both males and females. Glanzmann's thrombasthenia is associated with abnormal integrin αIIbβ3, formerly known as glycoprotein IIb/IIIa (GpIIb/IIIa), which is an integrin aggregation receptor on platelets. This receptor is activated when the platelet is stimulated by ADP, epinephrine, collagen, or thrombin. GpIIb/IIIa is essential to blood coagulation since the activated receptor has the ability to bind fibrinogen (as well as von Willebrand factor, fibronectin, and vitronectin), which is required for fibrinogen-dependent platelet-platelet interaction (aggregation).[2] In contrast, glycoproteinIb receptors are normal with Glanzmann's thrombasthenia. The role of GpIb is to enable platelet activation by contact with the von Willebrand factor-collagen complex that is exposed when the endothelial blood vessel lining is damaged. GpIb receptors are deficient in a disease known as Bernard–Soulier syndrome. Understanding of the role of GpIIb/IIIa in Glanzmann's thrombasthenia led to the development of GpIIb/IIIa inhibitors, a class of powerful antiplatelet agents. Symptoms of Glanzmann thrombasthenia vary quite a bit from one individual to the next, from very mild to potentially life-threatening bleeding.[3] Signs of the disorder are usually first noticed during childhood. People with Glanzmann

thrombasthenia may experience: Easy bruising, Nose bleeds, Bleeding from gums, Heavy or prolonged menstrual bleeding (menorrhagia) or bleeding after childbirth, Abnormal bleeding after surgery, circumcision, or dental work, Rarely, vomiting blood or passing blood in urine or stool due to bleeding in the gut (gastrointestinal hemorrhage) or genitourinary tract (kidneys, ureters, bladder, and urethra). Glanzmann thrombasthenia often causes more problems for women than men because of menstruation and childbirth.[4]The diagnosis of platelet function disorders requires a careful medical history and a series of tests that should be performed by a specialist at a hemophilia treatment center.[5]The bleeding time (a standardized test of the time it takes for a small cut to stop bleeding) is longer than normal. This test may be difficult to perform in young children. The closure time (a test that measures the time it takes for a platelet plug to form in a sample of blood) is longer than normal. Platelets do not clump together the way they should with several different chemicals in a laboratory test (platelet aggregation).GP IIb/IIIa is not detectable in blood samples (using a test called flow cytometry). Most people with platelet function disorders only need treatment during surgical procedures (including dental work) and after injury or accidents.[6] When needed, Glanzmann thrombasthenia may be treated with Antifibrinolytic drugs, Recombinant factor VIIa, Fibrin sealants, Hormonal contraceptives (to control excessive menstrual bleeding), Iron replacement (if necessary to treat anemia caused by excessive or prolonged bleeding), Platelet transfusions (only if bleeding is severe). People with inherited platelet function disorders should not take Aspirin®, nonsteroidal anti-inflammatory drugs (such as ibuprofen and naproxen), and blood thinners, which can make their bleeding symptoms worse.[7]

II. CASE REPORT:

A female patient about 16 years of age reported to the department with a chief complaint of bleeding gums for 4 days. Her medical history showed multiple episodes of bleeding from the gums for 4 days which was spontaneous. There was also prolonged bleeding following minor trauma and hematemesis. She also gives a history of epistaxis and Malena and also told that she had been diagnosed for Glanzmann's thrombasthenia with microcytic hypochromic anemia. Immunization history was uneventful. She was born of non‑consanguineous marriage. There was no significant family history of bleeding disorder. She also gives history of taking livogen and tranexamic acid for the past 3 years, and nine units of blood has been transfused till now. She was ectomorphic and poorly nourished. On examination, the child was conscious, cooperative, and was well‑oriented. Her investigations revealed hemoglobin - 6 g/dl, total leukocyte count - 5800, differential count polymorphs - 63, lymphocytes - 27, eosinophils - 1, monocyte -8, basophil – 1, platelets - 2.48 lakhs, red blood cell (RBC) count - 2.82 lakhs, hematocrit - 19.3, mean corpuscular volume - 68.4, mean corpuscular hemoglobin - 18.4, mean corpuscular hemoglobin concentration - 26.9, erythrocyte sedimentation rate –28 mm 1st hour, peripheral smear-microcytic hypochromic RBCs, white blood cell count normal, no immature cells seen, platelets count adequate, normal size, and morphology. Bleeding time - >15 min, activated partial thromboplastin Time - 27 s (control - 27 s), and prothrombin time/INR - 1. Serum fibrinogen was 200 mg/dL. Vitals were stable. As we were confirmed with the diagnosis we know that the bleeding control will be a difficult task. Oral prophylaxis was done on fast track basis not to cause excessive bleeding. Hydrogen peroxide was used to clean the oral cavity and gums properly. Through chlorhexidine gluconate solution was given for mouth rinsing. Tranexa soaked gauze was applied all over the gums and patient was asked to tightly close the lips to apply pressure on the medicated gauze to control hemorrhage. An intravenous ampule of Revici was also given. After an hour also, there was mile persisting bleed. The Tranexa gauze pack was changed and re applied. Zinc oxide eugenol periodontal pack was applied and a unit of blood as transfused to increase her hemoglobin. The pack was removed after 24 hours. There was no bleed present now. She was advised to take vitamin C supplements daily and to use soft tooth brush with fluoridated tooth paste. She was given proper instructions of maintaining oral hygiene.

III. CONCLUSION:

GT is a rare inherited bleeding disorder. It is primarily found in a limited number of populations, in which consanguineous marriage is common. Patients usually present with easy bruising and bleeding from epistaxis and dental extractions. With proper supportive care, GT has a very good prognosis. GT should always be considered as differential diagnosis while evaluating any case of bleeding disorder. If a thorough medical history is obtained, precautions are taken, and a hematological pretreatment is conducted prior to the referral to dental office, periodontal treatment also can be successfully carried out.

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