Glanzmann’s Thrombasthenia: A Case Report and Review

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CASE REPORT

ABSTRACT

Glanzmann’s thrombasthenia (GT) is a rare autosomal recessive disorder and characterized by a lack of platelet aggregation due to the absence of platelet glycoprotein (GP) IIb and IIIa. Usually, the disease leads to mild hemorrhage but sometimes bleeding is severe enough to be life-threatening. The site of bleeding in GT is clearly defined: purpura, epistaxis, gingival hemorrhage and menorrhagia are nearly constant features; gastrointestinal bleeding and hematuria are less common. In most cases, bleeding symptoms manifest rapidly after birth, even if GT is occasionally only diagnosed in later life. Diagnosis associates mucocutaneous bleeding with absent platelet aggregation in response to all physiologic stimuli, with normal platelet count and morphology. GT is more common in populations where marriage between blood relatives is common. Here, we present a case report of 24-year-old Indian male suffering from GT. A brief review of the relevant literature is also presented.

Keywords: Bernard-Soulier syndrome (BSS), Congenital afibrinogenemia, Epistaxis.

INTRODUCTION

In 1918, Dr Eduard Glanzmann’s first identified, in children from a village in the Swiss Alps.1 In 1956, Braunsteiner and Pakesch reviewed disorders of platelet function and described thrombasthenia as an inherited disease characterized by platelets of normal size that failed to spread onto a surface and did not support clot retraction.2 The diagnostic features of GT including the absence of platelet aggregation as the primary feature were clearly established in 1964 by the classic report on 15 French patients by Caen et al.3 The defect in platelet aggregation observed in GT can be directly related to a defect in fibrinogen binding which is either absent as in type 1, or considerably decreased as in type 2 GT. More recently, variant forms have been reported where the GP IIb/IIIa complex is present but qualitatively abnormal, leading to defective binding of fibrinogen.4 The clinical course can be mild or characterized by repeated hemorrhages whose severity varies from patient to patient. These may be unpredictable in a given patient, sometimes leading to morbidity or to life-threatening hemorrhages independent of the type of defect.5

In addition to compromising platelet-platelet interaction, clot retraction is also affected which contributes to delayed wound healing.6 The signs of GT occur early in life and include easy bruising, epistaxis and prolonged bleeding from relatively minor injuries. Epistaxis, menorrhagia, postpartum bleeding and surgical bleeding can be life-threatening.6 At a molecular level, GT is an extremely complex, heterogeneous condition with multiple deletions and mutations of the genes encoding the αIIbβ3 integrin.

In the review of 177 patients, only 12 were reported from the United States. In contrast, 55 patients were from Israel and Jordan and 42 were from South India.5 In certain ethnic groups, such as South Indian Hindus, Iraqi Jews, French gypsies and Jordanian nomadic tribes, thrombasthenia may actually be a common hereditary hemorrhagic disorder. This has recently been borne out by a report of 382 patients in Iran.8

CASE REPORT

A 24-year-old Indian male, known GT patient, who presented with gingival bleeding, was referred from the General Medicine Department of Goa Medical College and Hospital to the Department of Oral Medicine and Radiology, Goa Dental College, and Hospital Bambolim, Goa, India. There was positive history of repeated episodes of epistaxis and episodes of bleeding from lips, gums and tongue due to minor trauma. Family history revealed that he was born to consanguineous parents. Patient had an elder brother who expired 3 years back and had a history of recurrent episodes of epistaxis; however, the exact cause of the death is unknown. Other clinical manifestations of GT, like hematuria, bleeding of purpuric type, were absent in our patient. On oral examination, his gingival status showed normal appearance of gingiva with areas of bleeding along the marginal and papillary gingiva especially in the mandibular anterior region. Generalized stains were seen on the surfaces of the teeth. A diagnosis of mild localized gingivitis with normal periodontal status was considered (Fig. 1).

Routine laboratory investigations revealed normal hemoglobin (13.4 gm/dl), platelet count (3, 77,000/mm3), PT
(15 seconds), active PTT (29 seconds) and bleeding time (15 seconds). The platelet aggregation test presented no agglutination with epinephrine, ADP and collagen.

Periodontal treatment (scaling and polishing) was advised. The patient was prescribed (Tab Hemsyl 500 mg TDS for 1 day and 0.12% chlorhexidine mouth rinses for 1 week. And advised to use soft bristle tooth brush and maintain oral hygiene. The therapy was successful with decrease in gum bleeding (Fig. 2). Thereafter, he had weekly follow-up for 1 month and monthly recalls for the next 6 months were advised. We also intend to keep a long-term follow-up of this patient to prevent any episodes of bleeding due to negligence in oral health.

**DISCUSSION**

The differential diagnosis in patients with mucocutaneous bleeding, prolonged bleeding times and normal platelets includes von Willebrand’s disease and Bernard-Soulier syndrome (BSS) in addition to Glanzmann’s thrombasthenia, although it may not be easy to diagnose and differentiate among many bleeding disorders in which severe hemorrhage is seen. Normal ristocetin-induced platelet agglutination and normal platelet size clearly rule out the BSS, a disorder of platelet adhesion. Inherited thrombocytopenias are eliminated by a normal platelet count. Normal coagulation parameters rule out clotting disorders that can also affect platelet function such as congenital afibrinogenemia and von Willebrand disease. Acquired thrombasthenia must be eliminated in the absence of a family history of the disease. Platelet $\alpha_{\text{IIb}}\beta_3$ deficiency and abnormal platelet aggregation have been reported in patients with acute promyelocytic leukemia. $^{10}$ The etiology of this acquired disorder is probably a chromosome 15 to 17 translocation. Another problem in diagnosing GT is to eliminate patients with acquired autoantibodies that block aggregation, although these patients would often be thrombocytopenic. $^{11}$ These antibodies can be detected immunologically by their binding to $\alpha_{\text{IIb}}\beta_3$ of control platelets during incubation with the patient’s serum. $^{12}$

**Management of GT Patients in Dental Care Units**

The following methods should be considered to minimize the intraoperative and postoperative bleeding:

- Minimize trauma (e.g. elective sectioning of difficult extractions)
- Avoid flaps, as they provide a much greater bleeding area (harder to control)
- Prefer endodontic treatment of nonrestorable teeth rather than extraction.

Various adjuncts to hemostasis can be employed at the surgical site to enhance hemostasis, aid in vascular closure and prevent clot breakdown.

- Gelfoam is an absorbable gelatin sponge material that holds many times, its weight in blood and provides a stable ‘scaffold’ for clot formation. It is placed in tooth sockets in the form of tapered cones rolled from the sheet material. $^{13}$
- Bleed-X (QAS, Orlando, Fla) is a hemostatic product containing ‘microporous polysaccharide hemispheres’ (potato starch) that dehydrate blood and accelerate clotting. It can be applied to all types of surgical sites, including tooth sockets. It has been used successfully when Gelfoam cones have been rolled in the dry powder and placed in sockets. There are no known contraindications to its use. $^{14}$
- Tisseel (Baxter, Mississauga, Ont) is a fibrin sealant that acts both through its adhesive action and by direct contribution of fibrin to clot formation. Tisseel is technique sensitive and requires special preparation just before application. $^{15}$ It is expensive and is probably best reserved for particularly complicated or difficult dental situations.
- Tranexamic acid has also been used successfully in the form of a mouthwash or tablets after minor surgical procedures to inhibit postoperative bleeding episodes. In addition, the intravenous preparation can be diluted to a 4.8% aqueous solution and used as a mouthwash (4 times daily for 7 days). $^{16}$
- Electrocautery is a useful tool to slow intraoperative bleeding and stem postoperative episodes. However, it must
be used cautiously to avoid excessive tissue necrosis. Not only will the necrosis delay healing but it may also become a source of postoperative bleeding when the necrotic tissue sloughs.\textsuperscript{17}

Use of astringents may be considered, especially on incisions and raw areas. The ‘old tea-bag trick’ refers to the practice of using a tea bag as a pressure pack can be to minimize bleeding.\textsuperscript{18} The use of preformed splints to protect and enhance the placement of pressure on sockets is a valuable adjunct in multiple extraction procedures.\textsuperscript{19} Great care must be taken to ensure that there are no overextended areas on the splint that will traumatize soft tissue. Splints enhance the formation of firm, well-organized clots and prevent them from being dislodged or traumatized.\textsuperscript{19} Avoid prescribing medications that interfere in the normal function of platelets, such as aspirin and other nonsteroidal anti-inflammatory drugs (NSAIDs).

**CONCLUSION**

Although Glanzmann’s thrombasthenia is one of the rare bleeding disorders, one should be aware of this condition while evaluating patients with spontaneous gingival bleeding. Hence, the role of the dentist lies in giving proper periodontal care and encouraging maintenance of optimal oral hygiene, which is essential for both local and systemic health. With proper supportive care Glanzmann’s thrombasthenia has a very good prognosis.

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**REFERENCES**