Management of Dentoalveolar Trauma in a 3-year-old Child with Glanzmann's Thrombasthenia, a Rare Bleeding Disorder: Case Report and Review

Abstract

Glanzmann's thrombasthenia (GT) is a rare inherited bleeding disorder characterized by disturbed ability of the blood platelets to gather around the site of a broken blood vessel and fail to form a plug to stop bleeding due to deficiency of a glycoprotein IIb/IIIa in the process of blood clotting and results in moderate-to-severe bleeding on slightest injury to blood vessels. It is typically diagnosed in infancy or early childhood due to mucocutaneous bleeding tendencies. Treatment goals in GT are aimed at control of bleeding. Dentoalveolar trauma in patient with bleeding disorder poses a challenge in pediatric dentistry. The present article reports the successful management of subluxation dental injury in a 3-year-old child with "Glanzmann's thrombasthenia associated with sickle cell anemia." A narrative review of GT is given highlighting the importance of preventive dentistry and anticipatory guidance to reduce potential complications in such patients.

Keywords: *Bleeding disorders, hemostatic agent, luxation injuries*

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Introduction

Glanzmann's thrombasthenia (GT) is a rare inherited functional disorder of platelets with an incidence of one in 1,000,000 individuals.[1] It is autosomal recessive disorder characterized by normal platelet count, prolonged bleeding time, and abnormal clot retraction secondary to a dysfunction in GPIIb/IIIa complex.[2] It is typically diagnosed in infancy or early childhood due to tendency of mucocutaneous bleeding. The disease has good prognosis with increasing age. GT is more predominant in women as compared to men (58% vs. 42%).[3] It is more common in ethnic groups displaying consanguinity as in Iraqi Jews, selective Arab populations, French Gypsies, and Indians. The bleeding problem is caused by an abnormal gene present on nonsex-linked chromosome thus affecting females as well as males. GT is manifested shortly after birth with evidence of purpura in the neonate, followed by episodes of mucocutaneous bleeding and spontaneous bruising. The majority of patients will be diagnosed before the age of 5 years.[4] Recurrent epistaxis and gingival bleeding are the most common manifestations. Prevalence of severe bleeding decreases with age.[3] Newborns show diffuse petechial hemorrhage or umbilical cord bleeding. Menorrhagia is a critical challenge in female GT patients.[5] Pediatric dentist may encounter such child patients with the complaint of excessive bleeding following tooth eruption, exfoliation, traumatic dental injuries, caries, and other oral pathological lesions such as dentoalveolar abscess and cysts. Harmless events such as sneezing, crying, coughing, or common cold may also provoke bleeding.[6] Light transmission aggregometry, platelet function analyzer, and flow cytometry are diagnostic tests for GT. Western blotting can detect the identity of the affected gene.[7]

Case Report

A 3-year-old female child reported with the chief complaint of bleeding from the oral cavity and injury to the upper front teeth following fall from bed 10 h ago. Clinical examination revealed continuous gingival bleeding along with subluxation of 62 [Figure 1]. Medical records and examination revealed that the child was a prediagnosed case of "Glanzman's

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thrombasthenia" with sickle cell disease "AS" pattern. The patient was diagnosed for sickle cell disease at the age of 1½ years. The patient had a history of blood transfusion at the age of 1 due to decreased level of Hb up to 4.7 g/dl and had a swelling over the forehead. Computed tomography scan was normal. The patient also had spontaneous swelling over the knee that resolved without medication after 5/6 hours. There was a history of prolonged and continued bleeding from the tongue bite while eating and hospitalization for the same at the age of 2. Minor needle pricks at immunization sites also resulted in prolong and excessive bleeding. General physical examination of the patient revealed multiple ecchymotic patches and bruises on skin of the face, trunk, and limbs [Figure 2a and b] that would resolve spontaneously. There was no history of consanguineous marriages in the family. After traumatic dentoalveolar injury, the child received topical application of hemocoagulant ("Botroclot") at home for control of bleeding. However, blood oozing continued for 10 h during previous night before reporting to medical and dental hospital. Thorough oral clinical examination was done, however, radiographic examination was not possible due to severity of bleeding and child's anxious psychological status. Clinically, oozing was aggravated even by normal tongue movements and frequent contacts of tongue with the injured tooth. In view of patient's medical systemic illness and clinical condition, a treatment regimen was determined. A rigid splint was decided to be given in 61, 62, and 63 region using 21-gauge wire and glass ionomer cement (Xtracem Medicept Dental United states) as a luting agent. After splinting the injured tooth, topical hemocoagulant (Botroclot) was applied with soaked sterile gauze for 15–20 min after which the bleeding was controlled [Figure 3a]. The patient was further kept under observation for 1 h to monitor further oozing if any after treatment. There was no evidence of further bleeding after splinting the injured tooth and hence the patient was relieved from the operatory and was advised prophylactic antibiotic and anti-inflammatory drugs. The patient was recalled for check-up after 2 days. Normal healing was evident and also the patient was comfortable and playful. Parents were instructed for home care in terms of soft diet to the child for 15 days, brushing teeth after each meal with a soft brush, and topical application of chlorhexidine gel twice a day for 1 week to prevent infection in the periodontal tissues. The patient was recalled after 10 days and splint was removed [Figure 3b]. While taking photograph postoperatively, again bleeding was started in 73 tooth which was controlled by topical botroclot application. Anticipatory guidance was given to parents about oral hygiene maintenance and prevention of further episodes of orofacial trauma.

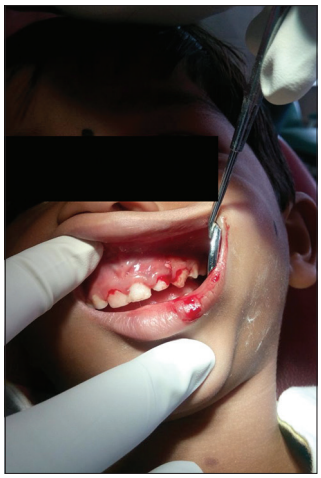

Figure 1: Preoperative clinical photograph showing subluxation injury and bleeding with 62

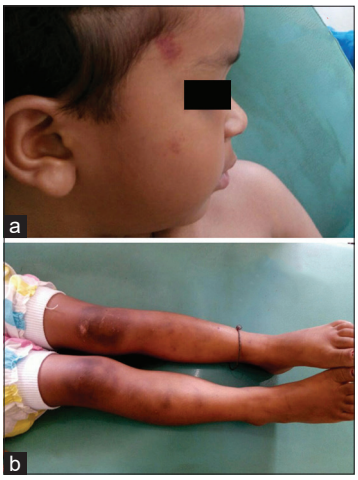

Figure 2: (a) Clinical photograph showing ecchymotic patches on the face and lower limb. (b) Clinical photograph showing ecchymotic patches on the lower limb

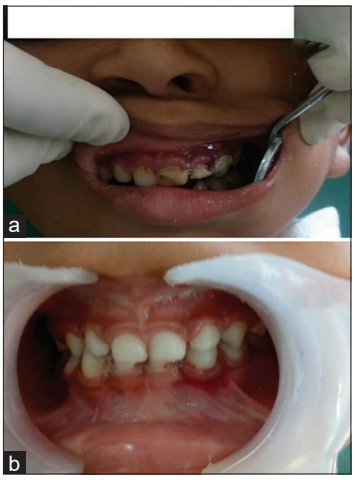

Figure 3: (a) Clinical posttreatment photograph showing splinting and topical botroclot application done with 62. (b) Clinical posttreatment photograph removal of splint with 62

Discussion

GT is a rare hereditary disease of blood clotting that affects clotting of blood after injury. This often results in moderate-to-severe bleeding on even a slightest injury to blood vessels as platelets fail to form a plug to halt

bleeding.[2] Depending on the severity, the disease is categorized as Type I – severe, Type II – less severe, and Type III – least severe.[2] Affected children show signs of bleeding during their first year of life, hence dentist can be the first to diagnose this disease. In the present case, the patient's diagnosis was done with platelet aggregation tests at the age of two. Although the disorder is related to consanguinity, the present case has no familial history of consanguineous marriages. The bleeding problem is caused by an abnormal gene which is not sex linked unlike hemophilia. Parents were well aware of the disorder and its consequences following even a slightest trauma to the patient. Hence, after dentoalveolar injury, topical application of hemocoagulant botroclot was done at home. Botroclot is a hemocoagulase isolated from venom of *Bothrops atrox* or *Bothrops jararaca*, a pit viper found in the tropical lowlands of South America. It consists of hemotoxins. The enzyme reptilase (botroxobin), derived from this snake's venom, is used to decrease or stop bleeding in patients with bleeding disorder in clinical treatments such as surgery, internal medicine, obstetrics and gynecology, ophthalmology, ear-throat-nose, and stomatology.[8] Besides botroxobin, other topical hemostatic agents used are absorbable hemostatic agents such as Surgicel (Johnson and Johnson, United Kingdom), Gelfoam (Pharmacia and Upjohn, Michigan, USA), fibrin glue, cyanoacrylate tissue adhesives, and splints. Perioperative antifibrinolytic agent like 10% solution of tranexamic acid used as mouthwash or for topical application to wound is the most preferred hemostatic agent.[9] However, in the present case, the oozing of blood continued for >10 h in spite of the application of topical hemostatic agent. Continuous oozing of blood for >12 h indicates clinically significant bleeding episode that may result in the development of hematoma or ecchymosis within the soft tissues and may also require blood product support.[10] The present case had subluxation of maxillary primary incisor tooth which is the most common dentoalveolar injury in children with primary teeth. It occurs due to high resiliency and less anchorage of alveolar bone, heal normally with or without any treatment in otherwise healthy patients. However, in view of the bleeding disorder in the present case, treatment plan was modified. Apart from simply keeping the patient under observation, a rigid splint was given using 21-gauge stainless steel wire and a high strength glass ionomer cement. Commonly used light cure composite material could not be used for fixation due to patients stressed, noncooperative status and continuous oozing of blood which would compromised required isolation. Splinting was done to prevent further oozing by movement of luxated tooth in socket. Such vertical or horizontal movements of tooth in its socket even in subluxation condition cause injury to microvasculature of periodontal ligament that precipitates rapid formation of platelet plug to form effective seals in the vessels within minutes. Splints enhance formation of firm well-organized clot and prevent them from being dislodged. Medications enhancing bleeding like nonsteroidal anti-inflammatory drug are to be avoided in such patients. However, due to patient's compromised general health with associated sickle cell disease, prophylactic antibiotics were prescribed to prevent infection.

Clinical implications

- Patients with GT must receive immunization against hepatitis B as they commonly receive blood transfusions
- They should also be advised to avoid contact sports
- Use of aspirin and nonsteroidal anti-inflammatory drugs must be restricted in such patients[10]
- Patients must be motivated to maintain excellent oral hygiene and participate in regular dental visits to mitigate the risk of gingival bleeding
- Women, on account of menorrhagic consequences, must be monitored for iron deficiency and should be placed on iron supplementation if necessary.

Conclusion

Pediatric dentist must be aware of bleeding disorders while evaluating continuous and prolong bleedings from minor injuries in playful preschool children. Role of pedodontist lies in giving proper anticipatory guidance to parents of children with GT with regard to avoid traumatic injuries, proper periodontal care, and encourage optimal oral hygiene maintenance pertaining to both local and systemic health in such children.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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