International Journal of Medical and Health Research
ISSN: 2454-9142
www.medicalsciencejournal.com
Volume 4; Issue 9; September 2018; Page No. 131-133

Dental management of Glanzmann's thrombasthenia patient: A case report

Krishnapriya Devan[1], Faizal C Peedikayil[2], Dhanesh N[3*]

[1] Post Graduate Student, Department of Pedodontics and Preventive Dentistry, Kannur Dental College, Anjarakandy, Kerala, India
[2] Professor and HOD, Department of Pedodontics and Preventive Dentistry, Kannur Dental College, Anjarakandy, Kerala, India
[3] Senior Lecturer, Department of Pedodontics and Preventive Dentistry, Kannur Dental College, Anjarakandy, Kerala, India

Abstract
Glanzmann's thrombasthenia is a rare inherited platelet disorder which can result in prolonged bleeding even from minor cuts or trauma. This case report presents the successful management of a 10 year old, known Glazmann's thrombasthenia patient who required extraction of a broken deciduous first molar with the topical application of crushed tranexamic acid tablet.

Keywords: glanzmann's thrombasthenia, rare bleeding disorder, tranexamic acid, dental management

Introduction

Hemostasis after an injury to the blood vessels is a complex process resulting from interaction between endothelium of blood vessel, coagulation factors and platelets. Inherited or acquired bleeding disorder can occur if there is a glitch in any of these steps [1-4]. Bleeding disorders that have a low prevalence in general population and constituting approximately 3-5% of all coagulation disorders are referred to as Rare Bleeding Disorders (RBD). RBDs include inherited deficiencies of coagulation factors such as fibrinogen, factor (F) II, FV, FV+FVIII, FVII, FX, FXI, FXIII, and multiple deficiency of vitamin K–dependent factors [5].

Glanzmann's Thrombasthenia (GT) is one such rare bleeding diathesis due to lack of platelet aggregation as the platelet glycoprotein IIb/IIIa (GP IIb/IIIa) complex required for platelet aggregation is either deficient of dysfunctional in these patients. GT was first described by Dr Eduard Glanzmann in 1918. It has an incidence of about 1:1000000 people, without any gender predilection and commonly occurs in offsprings of consanguineous parents [6].

Characteristic laboratory findings include normal platelet count, platelet morphology, Prothrombin time (PT) and Thromboplastin time (TT), prolonged activated Partial Thromboplastin time (aPTT) and bleeding time, deficient clot retraction and defective platelet aggregation. The common clinical findings in GT include purpura, epistaxis, easy bruising, gingival hemorrhage, and menorrhagia. Conditions that show clinical and laboratory findings similar to that of GT include von Willebrand's disease, Bernard–Soulier syndrome, and other platelet secretory defects [7, 8].

The dental treatment for patients with GT is a challenge because of the chances for prolonged bleeding after dental procedures like extraction. Therefore patients with GT must be treated with all the necessary precautions to control bleeding in case an episode of bleeding occurs.

Case History

A 10 year old girl reported to the Department of Pedodontics with the chief complaint of bleeding from gums in the lower left back teeth region since 2 months. Bleeding was reported when the patient chews food from the left side of the mouth and on brushing that area.

Patient is a known case of Glanzmann thrombasthenia. Patient's medical history revealed that at 4 years of age she was hospitalized and had history of multiple bleeding manifestations which included easy bruisability, prolonged bleeding from injury sites and ecchymotic patches on the body following trivial trauma. There was one history of bleeding from vaccination site and one episode of melena. During the episodes of prolonged bleeding, the child was transfused with packed cells and fresh frozen plasma. Family history revealed that the child was born of a non consanguinous marriage and there was no significant history of bleeding disorders in the family.

Since the bleeding from gums increased in frequency in the past few days and as there could be bleeding from gums during night, they came to Department of Pedodontics seeking treatment.

On examination, patient was conscious, cooperative and well oriented. Intraorally there was mild marginal gingival inflammation and a broken primary first molar tooth on lower left back teeth region (Figure 1).

The cause of bleeding was diagnosed to be due to gingival inflammation from the food lodgement and irritation from the broken piece of deciduous first molar. A treatment plan of extraction of the broken piece was made and as the patient belonged to the high risk category, they were advised to get physician's consent prior to procedure. Patient was advised hospital admission, if necessary.

After getting the signed consent from the parent, the paste of tranexamic acid (a 500mg tablet of Tranexamic acid [Pause 500,Emcure Pharmaceuticals] was crushed to fine powder with a mortar and pestle and mixed in one teaspoon of water to form a fine paste) was prepared to be applied locally on the extraction area immediately after extraction (Figure 2).

Extraction was done under local anesthesia. The prepared paste of tranexamic acid was applied and held in place with a wet gauze piece and mild biting pressure for 30 mins (Figure

3). After 30 mins there was considerable decrease in bleeding and the gauze piece was removed with the tranexamic acid left at the site to be swallowed slowly. After ensuring that the bleeding has stopped completely, the patient was send home after observation for 2 hours with strict instructions to not eat anything hot, not to touch area with tongue, not to spit forcefully and not to brush that area forcefully.

A review was done on the immediate day after extraction and after 7 days. The review after seven days (Figure 4) showed satisfactory healing of the extraction area with no new episodes of bleeding from the site.

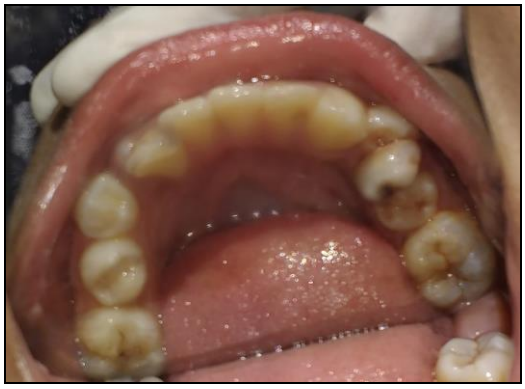

Fig 1: Intraoral photograph of lower arch showing the broken primary first molar on the left side

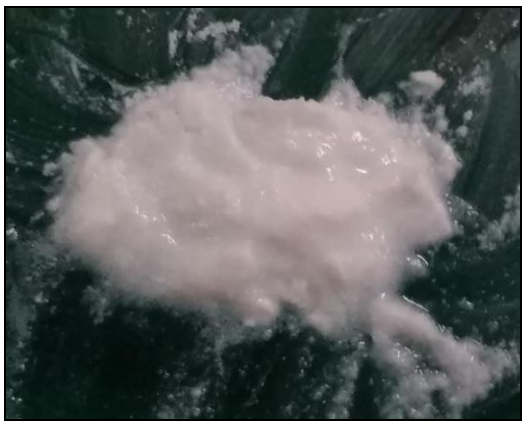

Fig 2: Tranexamic acid paste

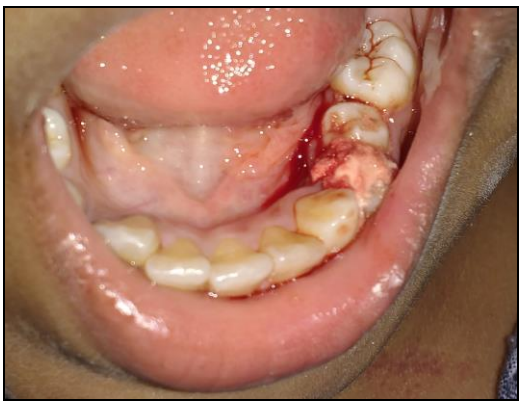

Fig 3: Tranexamic acid paste applied over the extraction site

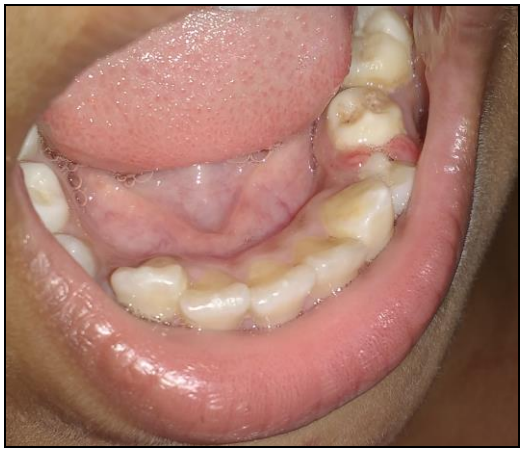

Fig 4: Review after 7 days

Discussion

Prolonged bleeding due to bleeding disorders can be a complication during surgical procedures like extractions in dentistry. A complete medical history and family history as well as dental history play an important role in identifying such patients.

The treatment for GT, is hematopoietic stem cell transplantation (HSCT). But due to risks associated with the procedure it is only recommended as a last resort when there is severe hemorrhage or when the patient develops antibodies leading to non-responsiveness to platelet transfusion [9].

Patients with GT usually seek dental treatment for bleeding from gingival [10] or bleeding from extraction socket. Gingival bleeding and bleeding from minor trauma to mucosa are common in patients with GT and to an extent can be prevented by taking special care in maintaining good oral hygiene.

Bleeding from surgical site or extraction site can be managed by local measures and severe or persistent bleeding requires systemic measures. The systemic hemostatic measures like platelet transfusion should be used judiciously and avoided if possible because of the risk of infection associated with repeated transfusions. The local measures to control minor bleeding includes application of cold compression, tranexamic acid (TXA), epsilon aminocaproic acid (EACA) or topical thrombin, gauze or gelatin sponge and laser coagulation.

Tranexamic acid is an antifibrinolytic agent derived synthetically from the amino acid lysine. The mechanism of action of TXA is prevention of blot clot breakdown. There is inhibition of fibrinolysis by blocking the lysine binding sites on plasminogen molecules and inhibition of the formation of plasmin [11]. A study by Mannucci [12] showed that TXA is ten times more potent than EACA.

In the literature there are several reports of antifibrinolytic agents that have been used successfully as an adjunct to platelet transfusion in inherited bleeding disorders [13, 14]. In the dental scenario, many studies report the efficacy of tranexamic acid as an oral rinse for prevention of post extraction/surgical bleeding in patients treated with anticoagulants [15, 16].

In a randomized control trial Nuvvula, Gaddam and Kamatham [17] concluded that freshly prepared tranexamic acid mouth wash (dissolving one packet of powder (500mg) in 10 ml of distilled water i.e. 5% TXA mouth wash) can be an

effective alternative to factor replacement therapy during dental scaling for controlling gingival hemorrhage in hemophiliacs.

Coetzee [18] stated that, most of the cases showed success when crushed TXA tablet of 500 mg suspended in water was placed on wet cotton piece that is put over the cavity/surgical site with mild biting pressure for about half an hour.

Children with GT should maintain their oral health meticulously. Adequate dental care should be provided to make sure that there are no untreated lesions as these can aggravate and lead to complex conditions that can result in bleeding. Utmost care should be taken while delivering dental treatment ensuring minimal trauma to the tissues and as little bleeding or no bleeding if possible.

Conclusion

Taking a detailed case history which include medical, dental and family history plays an important role in identifying patients with bleeding disorders. If proper identification is not done in such cases it can lead to serious or even life threatening conditions especially in rare bleeding disorders like GT where bleeding can be unpredictable. Treatment must be done taking all the necessary precautions and after educating the patients and parents about the risks associated and after getting their consent. As Pediatric dentists, in addition to rendering treatment, we can also educate the patient about the importance of maintaining good oral hygiene at an early age so that future bleeding complications from untreated lesions can be avoided.

References

1. Handin RI, Lux SE, Stossel TP. Blood Principles and Practice of Hematology. Philadelphia: JB. Lippincott Company, 1995, pp9-72.
2. Ogedegbe HO. An Overview of Hemostasis. Lab Med. 2002; 12(33):948-53.
3. Bick RL. Disorders of Thrombosis and Hemostasis Clinical and Laboratory Practice. 3rd edition. USA: Lippincott William's and Wilkins. Clinical assessment of patients with Hemorrhage, 2002, pp1-37.
4. Schiffman FJ. Hematologic Pathophysiology. Lippincottts Pathophysiology series. Philadelphia and New York: Lippincott-Raven Publishers Hemostasis and Thrombosis, 1998, pp161-223.
5. Peyvandi F, Palla R, Menegatti M, Mannucci PM. Rare Bleeding Disorders: General Aspects of Clinical Features, Diagnosis, and Management. Semin Thromb Hemost. 2009; 35:349-355.
6. Mehta BC, Agarwal MB, Bhanotra PC. Glanzmann's Thrombasthenia. J Postgrad Med. 1980; 26(3):210-212.
7. Levine PS. Qualitative disorders of platelet function. In: Pine WJ Jr, editor. Wintrobe's Clinical Heamatology. 10th ed. Maryland: Lippincott Williams & Wilkins. 1998; 2:1663-6.
8. Poornima G, Shivprasad S, Ashok L. Glanzmann's Thrombasthenia associated with a HBs Ag positive child: A case report. J Indian Soc Pedod Prev Dent. 2007; 25:S37-40.
9. Ishaqi MK, El-Hayek M, Gassas A, Khanani M, Trad O, Baroudi M, *et al*. Allogenic stem cell transplantation for Glanzmann's Thrombasthenia. Pediatr Blood Cancer. 2009; 52:682-3.
10. Kamatham R, Gaddam KR, Nuvvula S. Oral bleedings in hereditary blood coagulation disorders: A retrospective study. J NTR Univ Health Sci. 2016; 5:271-4.
11. Okamoto S, Hijikata-Okunomiya A, Wanaka K, Okada Y, Okamoto U. Enzyme controlling medicines: introduction. Seminars in thrombosis and hemostasis 1997; 23:493-501.
12. Mannucci PM. Prevention and treatment of major blood loss. New England Journal of Medicine 2007; 356:2301-11.
13. Bolton-Maggs PH, Chalmers EA, Collins PW, *et al*. A review of inherited platelet disorders with guidelines for their management on behalf of the UKHCDO. Br J Haematol. 2006; 135:603-33.
14. Alamelu J, Liesner R. Modern management of severe platelet function disorders. Br J Haematol. 2010; 149:813-23.
15. Sindet-Pedersen S, Ramstrom G, Bernvil S, Blomback M. Hemostatic effect of tranexamic acid mouthwash in anticoagulant-t patients undergoing oral surgery. N Engl J Med. 1989; 320(13):840-843.
16. Borea G, montebugnoli L, capuzzi P, magelli C. Tranexamic acid as a mouthwash in anticoagulant-treated patients undergoing oral surgery. An alternative method to discontinuing anticoagulant therapy. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1993: 75:29-31.
17. Nuvvula S, Gaddam KR, Kamatham R. Efficacy of tranexamic acid mouthwash as an alternative for factor replacement in gingival bleeding during dental scaling in cases of hemophilia: A randomized clinical trial. Contemp Clin Dent. 2014; 5:49-53.
18. Coetzee MJ. The use of topical crushed tranexamic acid tablets to control bleeding after dental surgery and from skin ulcers in haemophilia. Haemophilia. 2007; 13:443-4.