Case Report

Management of siblings with Glanzmann's thrombasthenia: A case report

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Abstract

Glanzmann's thrombasthenia is a rare, genetically inherited platelet disorder characterized by a lack of platelet aggregation. Until date, only close to 500 cases have been reported. GT is associated with clinical variability: some patients have only minimal bruising while others have frequent, severe and potentially fatal hemorrhages often making diagnosis difficult. Children are mostly diagnosed very early in life due to the spontaneous and unexplained mucocutaneous bleeding. The present case report deals with two siblings who reported with spontaneous gingival bleeding who were successfully managed by removal of local irritant factors and proper supportive care.

Keywords: Gingival bleeding, Glanzmann's thrombasthenia, hemorrhage, platelet disorder

Introduction

Platelets play a pivotal role in hemostasis. During hemostasis, damaged subendothelium releases sticky proteins and fibrinogen, which bind with aggregated platelets at the site of injury, to form a platelet plug. Platelets then provide a surface and phospholipid source for attachment of coagulation cofactors. Subsequent activation of the coagulation pathways prompts fibrin attachment to activated platelets, creating a thrombus. Any disruption in platelet function, whether acquired or inherited, will generate bleeding.[1,2]

Glanzmann thrombasthenia(GT) is a rare autosomal recessive platelet surface receptor disorder of GPIIb/IIIa (ITG αIIbβ3), either qualitative or quantitative, which results in faulty platelet aggregation and diminished clot retraction.[1]

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Children with GT are often diagnosed early in life, and often before the age of five, usually by unexplained spontaneous mucocutaneous bleeding. Life expectancy is normal.[1,2] Dental management of these patients is unpredictable as the severity of bleeding is unpredictable.[3] In the present paper, we report the rare occurrence of dental management of GT in two siblings.

Case Report

Two siblings of 14 years (female) and 8 years (male) reported to the dental clinic with the chief complaint of bleeding gums. The elder sibling had a history of bleeding for the past 5 days while the younger sibling had bleeding for 4 days. The children were apparently well until bleeding spontaneously started while brushing their teeth. The children reported late to the dental office, as they lived in a remote village 80 km away from where there was no access to health care.

On examination, a blood clot was seen extending from 12 to 16 for the elder sibling, whereas the younger sibling had a blood clot from 62 to 26 [Figure 1a]. The removal of the clot initiated fresh bleed from the gingiva [Figure 1b].

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The elder sibling was the first of four children of a consanguineous marriage [Figure 2] while the younger sibling was the youngest child. They gave no family history of any bleeding disorder. The girl had a history of purpuric patches since birth and prolonged bleeding after vaccination with recurrent episodes of mucocutaneous bleeding. The parents did not take the situation seriously. Blood investigation revealed the elder sibling had anemia. Both siblings had normal platelet counts, prolonged bleeding times, and abnormal platelet aggregation.

Past hematological evaluation reports of both siblings showed prolonged bleeding time, decreased clot retraction, and abnormal platelet aggregation response to physiologic stimuli. Platelet counts and morphology were normal. Their immunization schedule was complete and she had achieved all the milestones of growth at the correct time. The siblings were referred to a higher center for diagnosis where they were diagnosed to have Glanzmann's thrombasthenia. A pedigree chart was made which showed three other relatives were suffering from the same disorder.

The patients returned to the dental clinic for treatment. OPG for the elder sibling revealed bone loss interdentally with 12 and 13 [Figure 3a] whereas IOPA for the younger sibling revealed 62 [Figure 3b] had resorbed roots.

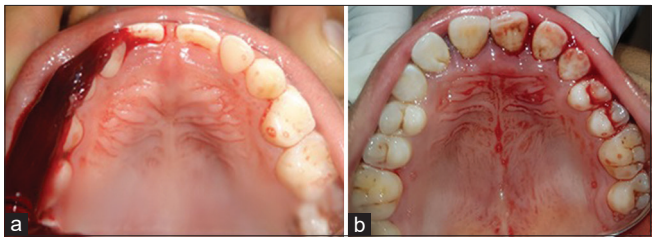

Figure 1: (a) Blood clot extending from primary lateral incisor to permanent first molar in younger sibling. (b) Fresh bleeding after removal of clot in older sibling

After consultation with a pediatrician, treatment was planned for both siblings. Both patients were hospitalized in case any future bleeding episode would occur. The elder sibling received a transfusion of 20 platelet concentrates and 10 packed RBC for anemia. The Hb level rose from 4 gm/dl to 9 gm/dl.

Considering the patient's past medical history, the pediatricians recommended one unit of platelet transfusion 2 h preoperatively and one unit postoperatively as blood loss was anticipated.

Oral prophylaxis was done for the elder sibling. Gauze dipped in tranexamic acid solution was placed from 12 to 14. The bleeding stopped after 1 h. Extraction of 62 was done for the younger sibling under local anesthesia infiltration (2% lignocaine in 1:200,000 dilution adrenaline, Neon Laboratories Ltd.). Hemostasis was achieved by compression of the socket and ice pack application. Both patients were admitted in the hospital for 5 days more for observation and then discharged.

Discussion

Pediatric dentists often come across different medical conditions when children are brought for dental management. Any episode of excessive bleeding after extraction or exfoliation of primary teeth, unexplained and spontaneous bleeding from the gingiva, and mucous membranes should alert the dentist about the possibility of bleeding disorders.[3]

GT is a rare autosomal recessive blood clotting disorder that can be present from birth. Signs and symptoms vary greatly from person

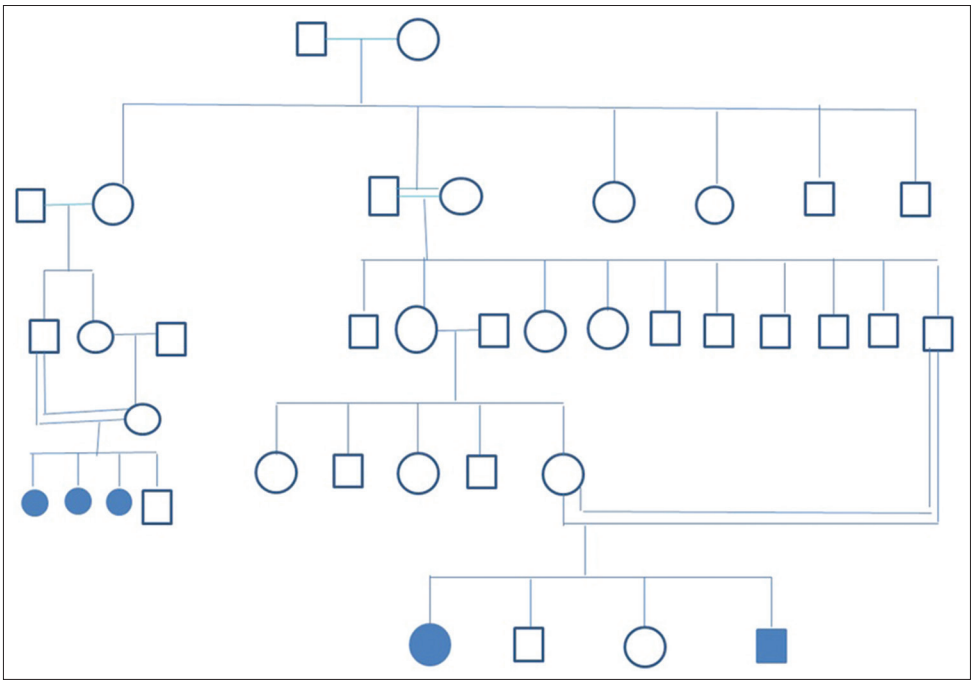

Figure 2: Pedigree chart

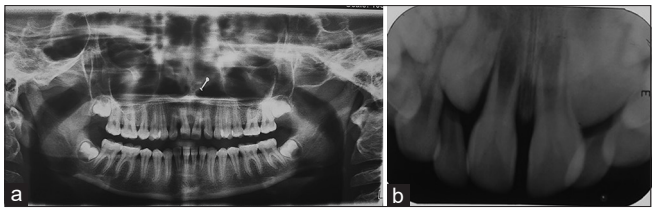

Figure 3: (a) OPG of elder sibling. (b) IOPA of younger sibling

to person. Until date, only 500 cases have been reported and the pathogenesis is still under debate. It is now believed that a defect in the platelet surface receptor of GPIIb/IIIa (ITG αIIbβ3) causes GT by the prevention of platelet plug formation during bleeding.[4] Hematological investigations in GT show prolonged bleeding time, defective or absent clot retraction, and failure of platelets to aggregate in response to adenosine diphosphate, epinephrine and collagen while coagulation tests are normal.[5]

There is always an underlying risk of postsurgical hemorrhage as bleeding in GT is unpredictable. Compression and gelatin sponge have been used as adjuncts to control bleeding.[6]

Therapy in GT is based on both the prevention and management of specific bleeding episodes. Since these patients require multiple transfusions in a lifetime, measures to avoid platelet alloimmunization should be taken, which is best accomplished by leukocyte-depleted blood products and the use of human leukocyte antigen (HLA)-matched platelets.[7]

In the present case, the removal of local irritant resulted in the stoppage of bleeding. Hence, the maintenance of good oral hygiene should be reinforced for such patients. Another unique feature was that both siblings had bleeding episodes around the same time and reported to the dental clinic together.

Conclusion

GT is one of the rarest bleeding disorders. The chances of patient and practitioner to be unaware of the condition are high. Any episode of uncontrolled bleeding should be investigated and proper medical history should be taken to prevent any post-operative complications.

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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Conflicts of interest

There are no conflicts of interest.

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