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Abstract

Aim: The aim of this report is to present a case of **Glanzmann thrombasthenia (GT) with oral manifestations** requiring periodontal management along with a discussion of the clinical, hematologic, and molecular level features of the disease.

Background: GT is a rare hematological disorder with oral manifestations affecting platelets and clotting. It is characterized by **spontaneous bleeding from mucosal tissues** and **excessive bleeding following skin damage**. It belongs to the group of hereditary platelet disorders and is due to a defect in one of two genes, **platelet membrane glycoprotein (GP) IIb/IIIa.**

Report: A 22-year-old female patient with **a history of Type I GT** and long-term care using **platelet transfusion** was referred for management of her gingival bleeding. Dental treatment included **scaling, polishing, oral hygiene instructions**, along with a prescription for anti-plaque agents. There was pronounced **improvement in her periodontal condition** after treatment. Reduced gingival bleeding resulted in an **increase in her hemoglobin level** from a pre-treatment level of 2 gm% to 8 gm% during her last visit. The patient was followed for eight months with no further periodontal complications.

Summary: A pronounced improvement in the periodontal condition of a GT patient occurred following routine scaling and polishing procedures along with proper oral hygiene maintenance instructions. The result was a reduction of the patient's gingivitis and associated spontaneous bleeding and an improvement in her hemoglobin status. GT patients should be managed for their periodontal problems following a hematological consultation. The importance of **proper maintenance of oral hygiene** as well as **regular recall visits** should be emphasized.

Introduction

- Glanzmann thrombasthenia (GT) is a rare autosomal recessive bleeding disorder characterized by a **quantitative or qualitative abnormality** of **platelet membrane glycoprotein (GP) IIb/IIIa receptors**.
- The features of this disorder include a **normal platelet count**, **normal prothrombin time**, **normal partial prothrombin time**, a **prolonged bleeding time**, and an **absence of platelet aggregation**.
- Clinical signs:

1. mucocutaneous hemorrhage with vulnerability to bruising,
2. menorrhagia,
3. epistaxis,
4. gingival hemorrhage,
5. intermittent episodes of gastrointestinal bleeding

- GT is important from a periodontal point of view because of the **frequent gingival hemorrhage** observed in these patients.
- The most frequent causes of bleeding: **Trauma** and **pressure.**

- Dental management of these patients is considered to be complicated because **the severity of bleeding** is unpredictable and **special precautions** are warranted before any surgical procedure is initiated.
- very few reports have appeared regarding the gingival and periodontal manifestations of GT and their management.

Case Report

- A 22-year-old female patient was referred for periodontal therapy from the Hematology department of a medical care facility for the management of severe spontaneous gingival bleeding.
- History:

1. 2 y/o the first episode spontaneous bleeding from her nasal cavity
2. 5 y/o severe gingival bleeding→she could only use a powdered dentifrice and her fingers to clean her teeth
3. 10 y/o increased bleeding was noted during menarche→blood transfusion
4. Initially a transfusion was required only yearly. The frequency of transfusion has been increased to twice a week for the past year.

- A general examination revealed her to be moderately built for her age with severe pallor; vital statistics appeared to be within normal range.

	Values from Patient Samples	Normal Values
Hemoglobin	2 gm/dL	12-16 gm/dL
Total white blood cell count	6200 cells/µL/cu mm	4300-10800 cells/µL/cu mm
Differential Count Neutrophils Lymphocytes Eosinophils	 67% 29% 4%	 50-70% 25-35% 1-3%
Platelet Count	260000/mL	150000-350000/mL
Packed Cell Volume	30.4%	36-48%
Bleeding Time (Ivy Method: Normal 2-6 min)	More than 15 min.	Up to 10 min.
Clotting Time (Lee and White: Normal 10-20 min)	13 min.	5-10 min.
Prothrombin Time	15.9 min.	Up to ±3 seconds
Activated Partial Thromboplastin Time	29.9 min.	45-60 sec.
Clot Retraction	Nil	
Platelet Aggregation with Adrenalin Collagen ADP Ricocetin	 Absent Absent Absent Normal	
Blood Picture	Mildly microcytic hypochromic with normal platelet morphology	

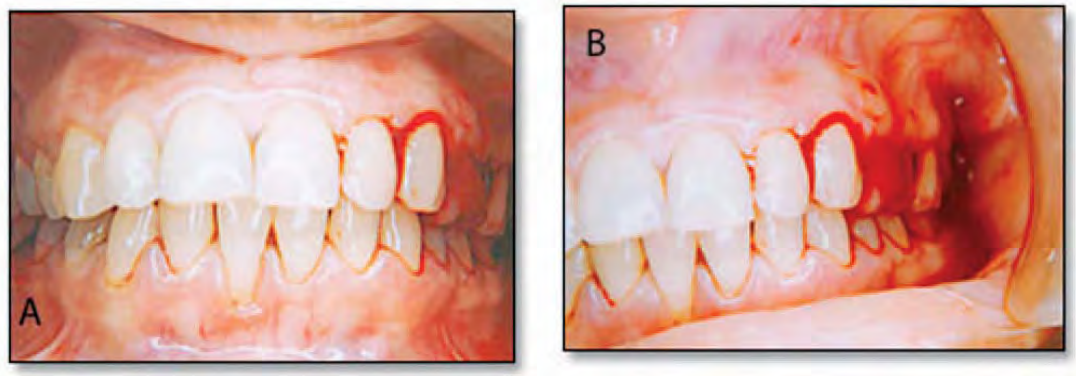
- **Intraoral examination:**

1. a moderate amount of dental calculus on the buccal aspect of maxillary first

molars and the lingual aspect of mandibular posterior teeth.

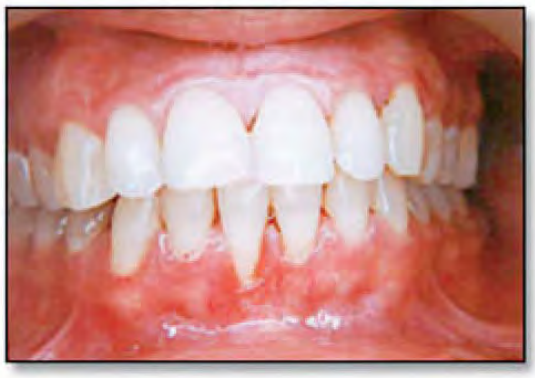
2. gingiva appeared to be very pale with spontaneous bleeding from the maxillary left and the mandibular right posterior regions, even with slight provocation of the tissue.
3. The consistency, contour, and position of gingiva were satisfactory except for slight gingival recession on labial aspect of tooth #41.

Data Element	Value
Plaque Index	1.3±0.25
Gingival Index	1.9±0.12
Calculus Index	2.2±0.38
Probing Depths	1.46±0.08
Clinical Attachment Level	0.98±0.24



Treatment

- Supragingival and subgingival scaling along with platelet transfusion
- Oral hygiene instructions were also given.
- Antiplaque agents like toothpaste and 0.12% chlorhexidine mouth rinse were prescribed.
- Weekly follow-up recall visits for the first month
- Monthly visits for the next eight months



The gingival condition improved in terms of color, consistency, and a reduction in bleeding tendency.

Discussion

- Glanzmann thrombasthenia (GT) is an autosomal recessive disorder affecting one of two genes, either platelet membrane glycoprotein IIb or IIIa.
- In GT, **one or both of these proteins are mutated** resulting in failure of platelet aggregation following stimulation and blood clot formation.
- GP IIb/IIIa as a dimer , once activated, will **bind to one end of fibrinogen**. **Another platelet** with its own GP IIb/IIIa can then **bind to the other end of fibrinogen** allowing formation of a large collection or aggregation of blood platelets commonly called a blood clot.

- The platelets of GT patients are functionally indistinguishable in that they **do not aggregate** in response to physiologic agents promoting clot formation like adenosine diphosphate (ADP), thrombin, or epinephrine
- The diagnosis: laboratory tests or cDNA probe

Test	In cases with Glanzmann Thrombasthenia
Bleeding Time	Prolonged
Aggregation of platelets in response to physiologic agents (Adrenalin, Thrombin)	Failure
Platelet Count	Within reference range
Platelet Morphology	Normal
Prothrombin Time and Activated Partial Thromboplastin Time	Within reference range
Urinanalysis	Proteineuria and microscopic hematuria
Sodium Dodecyl Sulfate – polyacrylamide gel electrophoresis of radiolabeled platelet proteins	Absence of GP IIb/IIIa

- GT is very rare, only 300 cases reported worldwide in the literature.
- Classified as Type 1, Type 2, or a variant type
  1. Patients with Type 1, the most severe form of disease, have less than **5%** of the normal amount of GP IIb/IIIa present in their platelets and an **absence of fibrinogen binding and clot retraction**
  2. Individuals with Type 2 GT have only **10 to 20%** of the normal amount of GP IIb/IIIa that can bind to fibrinogen resulting in a **moderately deficient clot retraction capability.**
  3. One other form of GT is a variant type having **greater than 50%** of the normal amount of GP IIb/IIIa resulting in extremely **variable fibrinogen binding and clot retraction.**
- The **probability of death** among GT patients of all types is estimated at the range of **5 to 12.8%**.
- The patient in this report had parents with second-degree relatives (cousins) who also suffered from Type 1 GT
- Previous reports and clinical management:
  - ✓ Palliative measures without using hemostatic agents
    →not considered because of the complex nature of the disease
  - ✓ Desmopressin (DDAVP) was successfully used following a dental procedure in one person→but its use is not routinely recommended
  - ✓ Transfusion of normal platelets, and the use of human leucocyte antigen-matched platelets to prevent alloimmunization complications is recommended
  - ✓ The last resort were bone marrow transplants and administration of the recombinant factor VIIa
- Pretreatment **consultation with a hematologist** and **platelet transfusion** is often

necessary in GT patients **prior to surgical or dental procedures and childbirth**

- The patient was able to **use an atraumatic soft bristle brush** as an adjunct to the treatment performed. Anti-plaque agents like **0.12% chlorhexidine and toothpaste** were prescribed and were found to be helpful in controlling plaque accumulation
- It is recommended all medications that might increase the risk of bleeding should be avoided.

Clinical Significance

- GT should be considered in the differential diagnosis of conditions with **severe bleeding from mucous membranes**. Studies report **the most difficult bleeds to correct** are those **from mucosa of the oral cavity and nose**. Rarely are deeper gastrointestinal hemorrhages reported.
- Any dental procedure performed in these patients should be preceded by **hematological consultation**. **Proper follow up** is considered to be an essential part in these cases.
- Patients as well as relatives should be made aware of the importance of oral hygiene measures as well as simple tips in preventing the occurrence of bleeding episodes.

題號	題目
1	下列何者非Glanzmann Thrombasthenia凝血功能疾病病人在牙科治療時應採取的措施? (A) 照會血液科醫師 (B) 檢查病人出血時間(bleeding time) (C) 治療時不需血小板輸血 (D) 治療後須定期回診
答案(C)	出處：
題號	題目
2	Glanzmann Thrombasthenia凝血功能疾病病人的日常口腔清潔最好為下列何種方法
	(A) 僅需使用漱口水 **(B)** 使用手指沾牙膏清潔 **(C)** 使用不會造成傷害的軟毛牙刷配合牙膏與漱口水清潔 **(D)** 不需清潔，只需定期回診檢查
答案(C)	出處：