Managing Acute Hemorrhage in Glanzmann Thrombasthenia in the Emergency Department : A Two-Case Report

Rim Karray[1], Fedi Ben Ali[1], Hela Ben Jmeaa[2], Racem Mnif1, Hana Kessentini[1], Olfa Chakroun[1], Imed Frikha[2], Noureddine Rekik[1].

1. ***Emergency Department, Habib Bourguiba University Hospital, Sfax, Tunisia.***
2. ***Cardiovascular and Thoracic Surgery Department, Habib Bourguiba University Hospital, Sfax, Tunisia.***

Abstract

Background: Glanzmann Thrombasthenia (GT) is a rare inherited platelet function disorder caused by a quantitative or qualitative deficiency of the glycoprotein IIb/IIIa complex, leading to defective platelet aggregation. Although typically associated with mucocutaneous bleeding, life-threatening hemorrhagic complications may occur and pose significant challenges in emergency settings.

Cases presentation: We report two cases of acute hemorrhage in patients with GT managed in the emergency department. The first case involved a 32-year-old man presenting with acute dyspnea due to a massive spontaneous left hemothorax. Despite prompt resuscitation, blood and platelet transfusions, antifibrinolytic therapy, and emergency chest drainage, the patient developed respiratory failure and subsequently died from septic shock. The second case concerned a 19-year-old man admitted after facial trauma complicated by persistent epistaxis refractory to local measures and tranexamic acid. Hemostasis was successfully achieved after platelet transfusion.

Conclusion: These cases highlight the wide clinical spectrum of hemorrhagic emergencies in GT and the limitations of standard coagulation tests, which are often normal. Management relies on rapid assessment, early antifibrinolytic therapy, and platelet transfusion, with recombinant activated factor VII as a valuable option in refractory or allo-immunized patients. Emergency physicians should be aware of this rare condition, as timely multidisciplinary management is crucial to reduce morbidity and mortality.

Keywords: Glanzmann Thrombasthenia, emergency, acute hemorrhage, management.

INTRODUCTION

The management of rare or complex hemorrhagic emergencies represents a real challenge for emergency department teams. Glanzmann's thrombopathy (GT), a rare congenital platelet disorder caused by a qualitative or quantitative deficiency of the glycoprotein IIb/IIIa complex, is one such condition. Although it is classically

associated with mucosal bleeding (epistaxis, gingival bleeding, menorrhagia, etc.), serious hemorrhagic complications such as spontaneous hemothorax have been reported, even in the absence of trauma. The risk is potentially higher in the presence of stress factors.

We report the cases of two patients who were admitted to the emergency department for bleeding associated with Glanzmann's disease, massive hemothorax, and epistaxis.

Case 1

A 32-year-old man was admitted to the emergency department with dyspnea. Initial examination using the ABCDE approach showed a clear upper airway, a respiratory rate of 22 breaths per minute, and a pulse oxygen saturation of 94% on room air. Pulmonary auscultation revealed decreased vesicular breath sounds over the left hemithorax, with no rales or pleural friction rub. The patient's blood pressure was 120/70 mmHg, and heart rate was 97 beats per minute. He was conscious, cooperative, and apyretic. The medical history revealed that the patient had been treated since childhood for GT and had stopped treatment several years ago. There was no cough or expectoration, nor any recent falls or trauma.

A point-of-care ultrasound showed a large left pleural effusion. An urgent chest X-ray revealed homogeneous opacity of the left hemithorax, associated with contralateral tracheal deviation, suggestive of a large pleural effusion. **(Figure 1)**

An urgent chest scan confirmed the presence of a large left pleural effusion, with blood density, causing total lung collapse and mediastinal deviation compressing the right heart chambers.

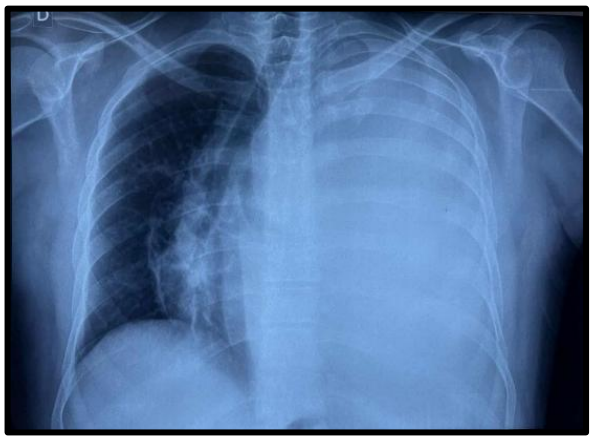

Figure 1: A chest X-ray revealing homogeneous opacity of the left hemithorax, associated with contralateral tracheal deviation, suggestive of a large pleural effusion.

Biological tests, including hemostasis, showed anemia, with hemoglobin at 8.2 g/dl, platelets at 229,000/mm3, PT 67%, APTT 28/30, and fibrinogen 2.7g/L.

The collegial decision with the cardiovascular and thoracic surgery team was to insert a chest drain on the left side. The patient was prepared before the interventional procedure. A transfusion of red blood cell concentrates was administered, supplemented by apheresis platelet concentrate to rapidly correct the hemostasis disorder. Treatment with tranexamic acid was also started to limit the extent of blood loss. After this preparatory phase, emergency chest drainage was performed laterally at the fourth intercostal space. It allowed for the immediate evacuation of 400 ml of old-looking blood, after which the drain was clamped and then unclamped in order to limit further blood loss while allowing for careful monitoring of subsequent flow. It brought back a total of 1600 cc/24 hours. The patient's respiratory condition progressively worsened the following day, requiring emergency orotracheal intubation and mechanical ventilation.

He was then transferred to intensive care. The prognosis was unfavorable, and the patient died after one month in septic shock following pneumonia acquired while on mechanical ventilation.

Case 2

A 19-year-old man, known to have GT since childhood, was admitted to the emergency room for craniofacial trauma following an assault. The initial examination following the ABCDE approach was normal. There was epistaxis, blood pressure of 110/70 mmHg, heart rate of 82 bpm, and oxygen saturation of 96% in ambient air. The patient had a periorbital ecchymosis and a bleeding wound on his upper lip, which was sutured. No other signs of physical assault were noted. The patient presented with moderate epistaxis that did not resolve after digital compression, anterior packing, and administration of tranexamic acid.

A brain scan was performed and revealed no abnormalities. The biological assessment, in this case hemostasis, showed no abnormalities (hemoglobin at 12.8 g/dl, platelets at 258,000/mm3, PT 68%, APTT 29/30, and fibrinogen 3 g/L).

The joint decision with the otolaryngology and hematology teams was to administer platelet concentrates to optimize the patient's hemostasis.

The outcome was favorable. The epistaxis subsided after the platelet concentrate transfusion.

He was discharged after 24 hours of monitoring.

DISCUSSION

Glanzmann's thrombasthenia is a rare disease characterized by a quantitative or qualitative deficiency of the GPIIb/IIIa complex (platelet integrin αIIbβ3), which is essential for platelet aggregation and clot formation. The clinical presentation of Glanzmann's thrombasthenia is dominated by severe mucocutaneous bleeding and the risk of major hemorrhagic complications (1,2). The diagnosis of this disease may be delayed because platelet counts and standard hemostasis tests (PT, APTT, fibrinogen) are generally normal: only aggregometry and immunophenotyping can confirm the diagnosis. It is crucial not to eliminate Glanzmann's thrombasthenia in cases of severe bleeding despite normal standard test results (1).

In an emergency, it is imperative to rapidly evaluate the severity of bleeding and administer appropriate treatment to prevent life-threatening risks (3).

Globally, the management of hemorrhage relies on rapid recognition of its severity, immediate control of the source of bleeding, and hemodynamic stabilization of the patient according to the ABCDE approach. It includes early vascular access, judicious fluid resuscitation with transfusion if necessary, and early administration of tranexamic acid. The correction of coagulation disorders and the rapid referral to specialized care are essential to improve prognosis (4).

In Glanzmann's thrombasthenia, initial control of bleeding relies on local measures such as compression and cryotherapy, combined with systemic administration of antifibrinolytics, particularly tranexamic acid, which is well documented in the literature as helping to control menometrorrhagia and other superficial blood loss. However, these measures are rarely

sufficient in cases of major hemorrhage, where transfusion of HLA-compatible platelet concentrates remains the gold standard (5-8).

Platelet concentrate transfusion is the cornerstone of treatment for moderate to severe bleeding. However, alloimmunization against platelet glycoproteins can lead to transfusion resistance, making management more complex (1,6).

For refractory patients, the use of recombinant factor VIIa (rFVIIa, Novo Seven) often achieves effective hemostasis, with a precise dosing regimen and strict monitoring to prevent thromboembolic complications (8, 9). This factor has proven to be a promising therapeutic option for the treatment of bleeding episodes in patients with Glanzmann's thrombasthenia, particularly in cases of alloimmunization or refractoriness to traditional platelet transfusions (10).

Optimal management requires multidisciplinary coordination, integrating emergency services, hematology, anesthesia, and blood banks, ensuring rapid access to appropriate treatments and personalized follow-up. As part of a comprehensive approach, it is essential to plan for long-term coordination with reference centers to anticipate bleeding episodes, particularly during trauma, surgery, or obstetric procedures (9,11).

These recommendations are supported by international consensus and data from prospective registries, which emphasize the importance of individualized protocols and rigorous follow-up to improve patient prognosis (1,9,11).

CONCLUSION

The management of acute bleeding in a patient with Glanzmann's thrombasthenia must be rapid and coordinated to avoid severe blood loss or hemorrhagic shock. Immediate measures include local treatment, antifibrinolytics, and appropriate platelet transfusions. Alloimmunization can cause these measures to be ineffective; in this situation, recombinant factor VIIa proves to be a crucial treatment. Multidisciplinary collaboration between emergency services, hematology, blood banks, surgeons, and the implementation of personalized protocols are essential to optimize hemorrhage control and limit mortality.

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