

CASE REPORT

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HEMODYNAMIC INSTABILITY IN A HEMOPHILIC PATIENT: CLINICAL CASE REPORT

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ABSTRACT

Hemophilic patients may present emergency situations related to large volume bleeding and secondary hemodynamic instability, such as hypovolemia. A 33-year-old female patient with a history of type 1 Diabetes Mellitus since childhood, myelodysplasia diagnosed 4 years ago with a history of monthly blood transfusions, was referred to the emergency room of a public hospital due to a persistent hemorrhage in right upper limb raffia. The patient was slightly overweight (BMI = 25.8kg / m²). Regarding the nursing process, after multiple semiological evaluations, the priority diagnosis for the patient was "Ineffective protection related to changes in coagulation, characterized by abnormal blood profile". Prescriptions made by the nurse conducting of the case assisted the patient in resolving the above diagnosis. Nursing and physician procedures for hemorrhage control are essential, especially when some signs of hemodynamic instability are found in the semiological evaluation, as well as personal care in preventing future trauma becomes important.

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INTRODUCTION

Platelets, along with proteins, are related to a series of biochemical reactions that lead to blood clotting. Dysfunctions in this cascade of reactions may lead to hemophilia, a disease characterized by absence of coagulant activity of factor VIII (Hemophilia A) or factor IX (Hemophilia B) due to changes in the genes that encode them (CAMERINI *et al.*, 2018). Coagulation factors VIII or IX and von Willebrand's disease, related to hemophilia, are the most prevalent hereditary coagulopathies in the world population. The most severe cases are identified by the occurrence of hemorrhages in the form of hematuria, epistaxis, hematemesis and hematoma formation (DE ARAÚJO *et al.*, 2019). The diagnostic is based in many factors but mainly presence of hemorrhagic cases in the family history and laboratory analysis through a blood analysis, which identifies changes in the blood clotting time. Gene therapy is an alternative treatment, as it provides the production of deficient factors by the body itself (RODRIGUES, 2018).

Hemophilic patients may present emergency situations related to large volume bleeding and secondary hemodynamic instability, such as hypovolemia.

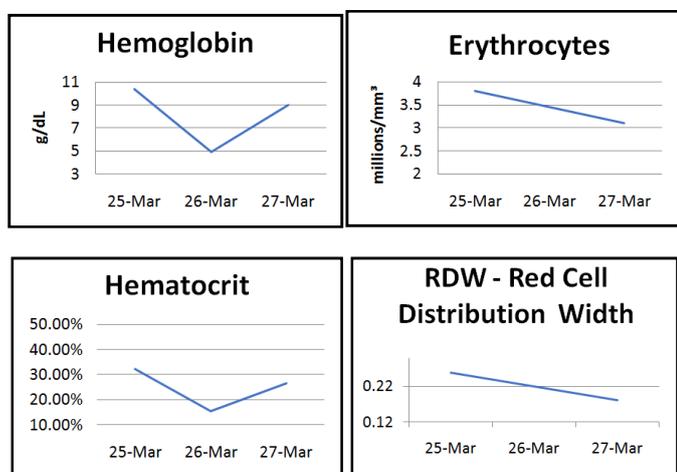
Clinical Case Report

A 33-year-old female patient with a history of type 1 Diabetes Mellitus since childhood, myelodysplasia diagnosed 4 years ago with a history of monthly blood transfusions, was referred to the emergency room of a public hospital in the state of Minas Gerais, due to a persistent hemorrhage in right upper limb raffia (RUL) complaint due to injury at the antecubital fossa after lipothymia. There is no history of coagulopathies in the family. At ectoscopy, the patient had a pale conjunctiva. Regarding the integumentary system, there was a significant amount of bruising on the upper and lower limbs due to pressure sensitivity, presence of a compressive dressing on the right upper limb, peripheral venous access (PVA) on the left upper limb, central venous access (CVA) in the right external

jugular and vesical delay probe (VDP). At cardiovascular clinical examination, she was tachycardic (135 bpm), normotensive (122/70 mmHg), with hyperphonic rhythmic sounds with systolic ejection murmur. The patient reported in-hospital constipation, although denied pain on abdominal palpation. In addition, she was slightly overweight (BMI = 25.8 kg / m²). At the time of hemodynamic emergence, patient received three blood transfusions and remained under observation for hematological and biochemical analyzes, as shown in the tables and graphs below.

Table 1. Sequential analysis of Prothrombin Activity Time (PAT) of the patient with hemophilia and RUL hemorrhage in raffia, 2019

Prothrombin Time (PAT)	Activity	Result	Reference values
Analysis 1		Confirmed incoagulable	70-100%
Analysis 2		Incoagulable	70-100%
Analysis 3		60%	70-100%



Graph 1. Hematological analysis of Hemoglobin, Erythrocytes, Hematocrit and RDW of a hemophilic patient with RUL hemorrhage in raffia condition, 2019

Blood smear analysis revealed marked anisocytosis (discrepancy in the size of some cells, especially red blood cells) (on March 25), progressing to mild (on March 27). In addition, there was mild poikilocytosis, which is the change in the shape of red blood cells, which characterizes an anemic profile. On March 27, it was possible to identify the presence of dacryocytes, which are droplet-shaped erythrocytes. Regarding the nursing process, after multiple semiological evaluations, the priority diagnosis for the patient was "Ineffective protection related to changes in coagulation, characterized by abnormal blood profile". Prescriptions made by the nurse conducting the case assisted the patient in resolving the above diagnosis. Guidance on the importance of protecting against trauma that could cause bleeding was prescribed; indication of the use of soft bristled toothbrush for oral hygiene; advice to avoid lifting heavy objects. Besides, the following were prescribed: increased intake of vitamin K-rich foods and general skin care such as therapeutic mattress use, given its sensitivity to pressure.

DISCUSSION

Hemophilia is a bleeding disorder caused by a coagulation factor VIII or IX deficiency, a sex-linked inheritance with a recessive pattern (VANDERHAVE *et al.*, 2012). As both are

X-linked disorders, the most affected individuals are men. The incidence of hemophilia A is 1 in 5,000 men while that of hemophilia B is 1 in 30,000 men. (CARCAO, 2012). Individuals with mild or moderate hemophilia have factor VIII C or factor IX C levels from 1 to 5% and 6 to 40%, respectively. These individuals tend not to develop spontaneous bleeding, but there are bleeding due to trauma or surgical or dental procedures (CARCAO, 2012). In the emergence of hemophilic hemorrhage of any kind, the UK Hemophilia Center Physician's Organization guidelines suggest that the maximum time between hospital arrival and clinical evaluation should not exceed 15 minutes and if bleeding treatment is required, the maximum transfusion time should not exceed 30 minutes (FOWLER *et al.*, 2011). In cases of emergence of trauma involving hemophiliacs, laceration or abrasion, injuries may occur. Therefore, it is recommended to treat superficial lacerations by cleaning the wound, applying pressure and using *steri-strips* adhesive sutures. For deep lacerations, it is necessary to increase the level of the factor corresponding to the disease and then suture the wound. Sutures can be removed under concentrated factor (SRIVASTAVA *et al.*, 2012).

After emergency care and the appropriate multiprofessional intervention, clinical follow-up turns to self-care, especially related to skin lesions. At this moment, the conduct taken by the nurse and his team through the nursing process, multiple semiological evaluations, identification of the nursing diagnosis (ND) and also the nurse's prescriptions are indispensable. (DE OLIVEIRA SANTOS *et al.*, 2019; NANDA INTERNATIONAL, 2018; HORTA, 1979). There are few articles found in the literature that specifically address the nursing process for patients with hemophilia. The articles found approach general issues related to the care process and guidance to patients and families, but do not mention clear diagnoses and nursing prescriptions (THOMAZELLI; DE LEMOS; MARQUES, 2019; VILLELA, 2019; NASCIMENTO SOUZA *et al.*, 2016). However, the need for skin self-care and other lesions is evident in order to reduce bleeding cases with hemodynamic complications in the hemophiliac patient (VILLELA, 2019). A recent study has shown several cautions and implications for daily life activities of hemophiliac patients, as well as warning signs for bleeding (DE ARAÚJO *et al.*, 2019). Such implications in the activities of daily living occur mainly in children, a phase that self-care is still under development and the clinical engagement with the condition is not understood, presupposing special care in the school phase (DOS SANTOS; LOPES, 2017). The management of oral health, besides being a nursing prescription, is addressed in several multidisciplinary studies, specifically dental surgeons, denoting the major importance of using soft bristle brushes, adequacy of other forms of cleaning and operative management of possible oral procedures to be performed (SREENIVAS NAGARAKANTI *et al.*, 2019; ANSARI *et al.*, 2019; REY, 2019; LIRAS; ROMEU, 2019). Similarly, vitamin K-rich foods, as well as the use of non-vitamin K antagonist medications, have been discussed in the literature as protective factors for bleeding in hemophiliac patients (GREMMEL *et al.*, 2018).

Conclusion

The clinical case reported showed a rare situation, in which from a raffia in RUL, a hemophiliac patient underwent with secondary hemodynamic instability a bleeding in surgical

wound. Considering the lack of familiarity with this disease and case specificity, extreme emergency treatment measures in cases of hemophiliacs with hemorrhage become important, as appropriate measures increase the chances of good prognosis and absence of more severe sequelae such as anemia and hypovolemic shock. Nursing and physicians procedures for hemorrhage control are essential, especially when some signs of hemodynamic instability are found in the semiological evaluation, as well as personal care in preventing future trauma becomes important.

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