Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Medicine

Perioperative Management of Hemophilia A: An Uncommon Inherited Blood Disorder

Safae Dehbi[1*], Taha Janfi[1], Larbi ED-Dafali[1], Aziza Bentalha[1], Salma ECH Cherif El kettani[1], Alae El Koraichi[2]

[1]Pediatric Intensive Care Unit, Children's Hospital, IBN SINA University Hospital, RABAT, MOROCCO
[2]Faculty of Medicine and Pharmacy of Rabat, MOHAMMED V University, RABAT, MOROCCO

DOI: https://doi.org/10.36347/sjmcr.2024.v12i12.004 | **Received:** 07.11.2024 | **Accepted:** 04.12.2024 | **Published:** 06.12.2024

***Corresponding author:** Safae Dehbi
Pediatric Intensive Care Unit, Children's Hospital, IBN SINA University Hospital, RABAT, MOROCCO

Abstract **Case Report**

Hemophilia A is a rare hematological disorder caused by an inherited X-linked factor VIII deficiency. It can be responsible for significant perioperative bleeding with fatal consequences if not diagnosed and managed correctly in the preoperative phase. We report the case of a 6-month-old infant scheduled for a cure for bilateral inguinal hernia. During the pre-anesthetic consultation, given that the child was uncircumcised and had not yet reached walking age, a blood workout was made, which revealed a prolonged aPTT with a patient/control aPTT ratio of 3.27, without hemorrhagic signs. An activity work-up of clotting factors showed less than 1% of normal levels of active clotting factor VIII. Thus, the diagnosis of a severe form of hemophilia A was made. Recombinant factor VIII was administrated in the preoperative period, with an activity factor VIII control over 60%. The surgical procedure was uneventful, with minimal intraoperative bleeding. Our case emphasizes the importance of prophylactic treatment and a proper pre-anesthetic assessment, which are essential to establish an appropriate management strategy and avoid complications, most of which are fatal.

Keywords: Hemophilia A, Anesthesia Management, Pediatrics.

INTRODUCTION

Hemophilia A is a rare hematological disorder caused by an inherited X-linked factor VIII deficiency. It can be responsible for significant perioperative bleeding with fatal consequences if not diagnosed and managed correctly in the preoperative phase. We report the case of a 6-month-old infant diagnosed with hemophilia A following a pre-anesthetic consultation.

CASE REPORT

A 6-month-old male infant, weighing 7 kg, uncircumcised, his parents are non-consanguineous, with a family history of hemophilia A of the maternal grandfather. He had been diagnosed since birth with left hemicorporeal hypertrophy. The malformative evaluation revealed right testicular ectopia, a bilateral inguinal hernia, and a suspicious left scapular mass. After reviewing his case, it was decided to operate him and have a biopsy of the scapular mass.

During the pre-anesthetic consultation, given that the child was uncircumcised and had not yet reached walking age, a blood count, an activated partial thromboplastin time (aPTT), and a prothrombin time (PT) were carried out. The results showed a prolonged aPTT with a patient/control aPTT ratio of 3.27, without hemorrhagic manifestations. Given the family history of hemophilia A, an activity work-up of clotting factors (V, VIII, Von Willebrand, IX, XI, XII) was done and showed less than 1% of the normal level of active clotting factor VIII. Thus, the diagnosis of a severe form of hemophilia A was made.

For the preoperative management, the patient received recombinant factor VIII concentrate at a dose of 1000 IU / 12 hours, with a target clotting factor VIII activity of over 50% according to our local protocol. The treatment was initiated on day-1, with a preoperative control at 60%. The patient was subsequently admitted to the operating theatre with a 24-gauge venous line and was monitored by electrocardiogram, pulse oximeter, and non-invasive blood pressure. The surgical procedure was uneventful, with minimal intraoperative bleeding estimated at 30cc. During the postoperative period, factor VIII concentrate was infused with the same dosage (1000 IU/12h) for 4 days. On postoperative day 5, clotting Factor VIII activity was 55%, with hemoglobin

levels remaining approximately 11g/dL, with no hemorrhagic manifestations. He was then discharged on day 6.

DISCUSSION

Hemophilia's history dates back to the 2nd century B.C. with Hebrew writings recounting that male babies did not need to undergo circumcision if two of their brothers had already succumbed to the procedure before them [1]. It was not until 1828, thanks to HOPFF's work at the University of Zurich, that the term hemophilia appeared [2]. It is an X-linked recessive hereditary disease, which explains its predominance in males with an incidence of 1/5000 [3]. Still, it can also be seen in girls carrying the mutation with a generally minor form.

The diagnosis is usually made in the presence of severe spontaneous or post-traumatic bleeding, following a screening of children with a family history of hemophilia A using molecular biology techniques to search for the gene mutation coding for factor VIII [4], located on the long arm of the X chromosome (Xq28), or an incidental finding in which the anesthesiologist plays a pivotal role. Clinical examination, searching for personal and family history of any bleeding complications, and asking if the child has reached walking age are fundamental in the pre-anesthetic consultation. A standardized questionnaire use is recommended to facilitate the process. However, none has yet been validated in French for children. The bleeding assessment tool of the International Society of Thrombosis and Hemostasis (ISTH BAT), the only one validated in pediatrics, includes 12 items graded from -1 to +4. A score greater than 2-3 in children indicates a significant hemorrhagic syndrome [5], with a negative predictive value of 99%. However, it includes so many items making its application systematically in the pre-anesthetic consultation complex, with an average completion time of 20 minutes. After completing the interrogatory step, the anesthesiologist must prescribe blood tests to look for clotting disorder in case of a positive bleeding diathesis or if walking age is not reached yet. However, a normal aPTT value does not rule out congenital coagulopathy. The minor the deficit is, the lower the test sensitivity becomes. So, a biological work-up must be interpreted according to the clinical context.

Three degrees of severity can be distinguished, the minor form with factor VIII activity varying between 5% and 40%, in which case patients tend to bleed in major trauma. In the moderate form, factor VIII activity varies between 1% and 5%, and bleeding can be caused by mild to minor trauma. Finally, the severe form with a factor VIII activity less than 1%, which was the case of our patient, with a high risk of spontaneous bleeding that could be life-threatening [6].

Except for situations with major bleeding, perioperative management of patients with hemophilia A is based on prevention by restoring an adequate factor VIII activity to ensure effective hemostasis. The classification of surgery into minor, moderate, and major hemorrhagic risk, recommended for antiplatelet agent management [7], is not adapted to risk assessment in the case of congenital coagulopathy [8]. The risk is generally considered minor for the following procedures: insertion and removal of central venous lines, synoviorthesis, dental extraction of less than three teeth (excluding wisdom teeth), treatment of congenital cataracts, and removal of skin lesions. On the other hand, adenoidectomy, tonsillectomy, osteotomies and arthrodesis, reduction of fractures, arthroscopy, extraction of three or more teeth or wisdom teeth, and digestive endoscopy biopsies involve a major risk. However, it is up to the anesthesiologist and surgical team to provide the most relevant information on the invasiveness of the interventional procedure, so that the hemostasis specialist can recommend the most appropriate preventive treatment according to local protocols. The latest recommendations from the World Federation of Hemophilia [6], preconize Factor VIII activity between 80% and 100% for major surgery (e.g. cardiovascular surgery and neurosurgery) and above 50% for the other procedures. The dose to be administered is calculated according to the following formula: Dose (IU) = Weight (kg) x Desired rate of increase in FVIII activity x 0.5 [9].

Some precautions must be taken in the operating theatre [10]. We tend to avoid nasal intubation to prevent submucosal hemorrhages for airway management unless there is a formal indication. The pressure points must be protected when positioning the patient to prevent intramuscular hematomas or hemarthrosis. Hemodynamic stability is a major requirement since tachycardia and hypertension can worsen bleeding and make surgical hemostasis more delicate. And let's not forget the surgeon's role in ensuring meticulous hemostasis, particularly of small vessels.

Post-operatively, supplementation may be necessary for 5 to 7 days, depending on the nature of the procedure, to reinforce hemostasis, with close clinical monitoring.

CONCLUSION

Thanks to advances in available therapies and the use of long half-life drugs with the concept of prophylactic treatment, it has been possible to improve the mortality rate and quality of life of hemophilic A children. However, pre-operative diagnosis and a proper pre-anesthetic assessment are essential to establish an appropriate management strategy and avoid complications, most of which are fatal.

Authors' Contributions

- Safae Dehbi, Taha Janfi: patient management

- Safae Dehbi, Larbi Dafali, Alae El koraichi : data collection
- Safae Dehbi: manuscript drafting
- Alae El Koraichi, Aziza Bentalha and Salma ECH Cherif El Kettani: manuscript revision.

All the authors have read and approved the final version of the manuscript.

Consent: The author obtained written informed consent from the patient's family for submission of this manuscript for publication.

REFERENCES

1. Rosner, F. (1995). *Medicine in the Bible and the Talmud: Selections from Classical Jewish sources* (Vol. 5). KTAV Publishing House, Inc..
2. Nilsson, I. M. (1994). Blödarsjuka--förr och nu [Haemophilia--then and now]. *Sydsven Medicinhist Sallsk Arsskr*, *31*, 33-52.
3. Kar, A., Phadnis, S., Dharmarajan, S., & Nakade, J. (2014). Epidemiology & social costs of haemophilia in India. *Indian Journal of Medical Research*, *140*(1), 19-31.
4. Craig, D., Seaman, F., Xavier, F., Ragni, M. V., & Hemophilia. A. (2021). (Factor VIII Deficiency). *Hematology/Oncology Clinics of North America*, *35*(6), xiii–xiv.
5. Rodeghiero, F., Tosetto, A., Abshire, T., Arnold, D. M., Coller, B., James, P., ... & Lillicrap, D. (2010). ISTH/SSC bleeding assessment tool: a standardized questionnaire and a proposal for a new bleeding score for inherited bleeding disorders. *Journal of Thrombosis and Haemostasis*, *8*(9), 2063-2065.
6. Srivastava, A., Santagostino, E., Dougall, A., Kitchen, S., Sutherland, M., Pipe, S. W., ... & WFH Guidelines for the Management of Hemophilia panelists and co-authors. (2020). WFH guidelines for the management of hemophilia. *Haemophilia*, *26*, 1-158.
7. National Guideline Centre (UK). (2016). *Preoperative Tests (Update): Routine Preoperative Tests for Elective Surgery*. NICE Guideline, No. 45. London: National Institute for Health and Care Excellence (NICE).
8. Solimeno, L. P., Escobar, M. A., Krassova, S., & Seremetis, S. (2018). Major and minor classifications for surgery in people with hemophilia: a literature review. *Clinical and Applied Thrombosis/Hemostasis*, *24*(4), 549-559.
9. Kwak, J., Mazzeffi, M., Boggio, L. N., Simpson, M. L., & Tanaka, K. A. (2020). Hemophilia: A review of perioperative management for cardiac surgery. *Journal of Cardiothoracic and Vascular Anesthesia*, *34*(9), 2455–64.
10. Flores, R. P. G., Bagatini, A., Santos, A. T. L. D., Gomes, C. R., Fernandes, M. S., & Molon, R. P. (2004). Hemofilia e anestesia. *Revista brasileira de anestesiologia*, *54*, 865-871.