Open Access Library Journal
2023, Volume 10, e9826
ISSN Online: 2333-9721
ISSN Print: 2333-9705

# From Rheumatic Fever to Hemophilia a Major: A Case Report

**Najlae Nassiri, Hind Zahiri, Ayyad Ghanam, Aziza El Ouali, Abdeladim Babakhouya, Maria Rkain, Noufissa Benajiba**

Department of Pediatrics, Mohamed VI University Hospital, Oujda, Morocco
Email: najlae.nassiri93@gmail.com

**How to cite this paper:** Nassiri, N., Zahiri, H., Ghanam, A., El Ouali, A., Babakhouya, A., Rkain, M. and Benajiba, N. (2023) From Rheumatic Fever to Hemophilia a Major: A Case Report. *Open Access Library Journal*, **10**: e9826.
https://doi.org/10.4236/oalib.1109826

**Received:** January 31, 2023
**Accepted:** February 25, 2023
**Published:** February 28, 2023



## Abstract

Severe bleeding after tonsillectomy can lead to airway obstruction and be life-threatening. We report a case of post-tonsillectomy bleeding in a ten-year-old patient with previously undiagnosed hemophilia A, with a history of bleeding after circumcision and hematomas following minimal trauma. He was treated for rheumatic fever because of arthritis and repeated sore throats, for which reason he underwent tonsillectomy. In the postoperative period, the patient presented active bleeding even after ligation of the left pillars, with a prolonged APTT in the biological work-up and a factor VIII dosage that collapsed to 0. The patient was diagnosed with hemophilia A major at 7% and was treated with an anti-hemophilic factor with good improvement.

## Subject Areas

Hematology

## Keywords

Tonsillectomy, Hemophilia A, Rheumatic Fever, Postoperative Bleeding

## 1. Introduction

Hemophilia is a ubiquitous condition with an incidence of one case per 10,000 births [1]. Great progress has been made in recent years in all aspects of the management of patients with hemophilia [2]. Hemophilia is a genetic disease of recessive transmission linked to the X chromosome. It affects males and is transmitted by females, therefore known as carriers [3]. It is due to a decrease in the level of the coagulation factor, factor VIII (hemophilia A) and/or factor IX (hemophilia B), and is particularly associated with a risk of bleeding at the time of surgery and during the perioperative period.

We report a case of post-tonsillectomy bleeding in a ten-year-old patient with previously undiagnosed hemophilia A.

## 2. Observation

This is a 10-year-old male child from a 1st degree consanguineous marriage, with a history of recurrent angina since the age of 3 years at a rate of 7 episodes per year, tonsillar hypertrophy, and fleeting arthritis affecting the large joints, The diagnosis of rheumatic fever was evoked as we are in an endemic country, in this sense the patient was put on benzathine benzylpenicillin (extencillin) and was scheduled for a tonsillectomy.

After the tonsillectomy procedure, the patient presented a hemorrhagic syndrome with persistent bleeding for three days, requiring a repeat operation for ligation of the left pillars, and then referred to our training center for further management, Clinical examination on admission to the pediatric emergency room showed a conscious child with a Glasgow score of 15/15, conjunctiva slightly discolored, normotensive at 100/65mmHg, tachycardia at 130 beats/minute, eupneic at 18 cycles/minute, with saturation in ambient air a 98% and apyretic at 36.9˚C, the endo buccal examination showed the presence of a blood clot in front of the surgical site with no active bleeding, the rest of the clinical examination was without anomaly. On further questioning, we noted in the history the notion of post-circumcision bleeding and bruises from minimal trauma with no similar cases in the family.

The blood count showed anemia at 8.7 g/dl normochromic normocytic (mean corpuscular volume 79 and mean corpuscular hemoglobin 27), in front of the two episodes of postoperative bleeding in the history of the patient: after the circumcision and after the tonsillectomy procedure, a hemostasis check-up was requested and showed a prolongation of the activated partial thromboplastin time to 3.13, a normal prothrombin level, and a factor VIII level that had collapsed to 0.7%, with factor IX and Willbrand factor levels returning to normal. Thus, the diagnosis of major hemophilia A was made in view of the postoperative bleeding in a male patient with biological confirmation. The patient benefited from the administration of an anti-hemophilic factor and is currently being followed in our training center with good improvement.

## 3. Discussion

Hemophilia A is an X-linked bleeding disorder caused by mutations in the genes for coagulation factor VIII. This factor is involved in the intrinsic pathway of blood clotting. The prevalence of hemophilia A is one per 5000 live male births [4] [5].

The transmission of hemophilia follows the laws of X-linked recessive inheritance. However, the absence of a known family history is a frequent situation when a new case of hemophilia is diagnosed.

Hemorrhagic manifestations in hemophilia A major occur most often follow-

ing minimal trauma, with the characteristic hemorrhages: hemarthroses (70% - 80%), hematomas (ten-20%), hematuria, and visceral hemorrhages being less frequent.

The severity depends on the severity of the deficiency:

Severe hemophilia: characterized by a profound deficiency in antihemophilic factor, the level of FVIII or FIX is less than 1% (compared to the normal level which is 0.1 mg/L of plasma for FVIII and 5 mg/L of plasma for FIX). These patients bleed spontaneously and are particularly prone to intramuscular and joint bleeding;

Moderate hemophilia: the level of factor VIII or IX is between 1% and 5%. Hemorrhagic bleeding is more moderate and usually secondary to trauma;

Minor hemophilia: the level of FVIII or FIX is between six and 30%. Accidents are rare and only provoked. It is rarely detected at birth.

Randhawa A [6] reported the case of a 5-year-old boy with no history of bleeding disorders, who presented with recurrent bleeding after a tonsillectomy. Initial coagulation tests showed a slight prolongation of the activated partial thromboplastin time (APTT) and further biological examinations showed a collapsed factor VIII level, which confirmed the diagnosis of previously undiagnosed mild hemophilia.

This can lead to serious complications. For this reason, FVIII testing should be an integral part of the preoperative workup for surgical procedures in male children.

## 4. Conclusion

As in our case, hemophilia may be discovered incidentally after tonsillectomy, even if the patient did not report a bleeding tendency and the preoperative workup was normal. A careful history of bleeding history is important for uncomplicated surgery.

## Conflicts of Interest

The authors declare no conflicts of interest.

## References

[1] Nathwani, A.C. and Tuddenham, E.G.D. (1992) Epidemiology of Coagulation Disorders, in Bailliere's Clinical. *Haematology*, **5**, 383-439. https://doi.org/10.1016/S0950-3536(11)80025-9

[2] Kasper, C.K., Mannucci, P.M., Boulyjenkov, V., Brettler, D., Chuansumrit, A., Heijnen, L., *et al.* (1992) Hemophilia in the 1990s: Principles of Treatment and Improved Access to Care. *Seminars in Thrombosis and Haemostasis*, **1**, 1-10. https://doi.org/10.1055/s-2007-1002404

[3] Legat-Fagnoni, C., Bertrand, M.A., Woronoff-Lamsi, M.C., Clerc, B., De'moly, P., *et al.* (2008) Traitement des anomalies de la coagulation pharmacie clinique et the'rapeutique. 3e Ed., Elsevier, France, 509-527. https://doi.org/10.1016/B978-2-294-06234-6.50027-2

[4] Srivastava, A., Brewer, A.K., Mauser-Bunschoten, E.P., Key, N.S., Kitchen, S., Llinas, A., Ludlam, C.A., Mahlangu, J.N., Mulder, K., Poon, M.C. and Street, A. (2013) Treatment Guidelines Working Group on Behalf of The World Federation of Hemophilia. Guidelines for the Management of Hemophilia. *Haemophilia*, **19**, e1-e47. https://doi.org/10.1111/j.1365-2516.2012.02909.x

[5] Francùhini, M. and Mannucci, P.M. (2012) Past, Present and Future of Hemophilia: A Narrative Review. *Orphanet Journal of Rare Diseases*, **7**, Article No. 24. https://doi.org/10.1186/1750-1172-7-24

[6] Randhawa, A., Bondin, D. and Kumar, B.N. (2014) A Surgical Presentation for Hemophilia A. *BMJ Case Reports*, bcr2014203687. https://doi.org/10.1136/bcr-2014-203687