Research Article

Acute Subdural Hematoma In A Patient With Hemophilia A: Case Report

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Abstract

Background: Intracranial hemorrhage (ICH) is a rare but life-threatening complication in patients with hemophilia, with a markedly higher incidence than in the general population. Even minor head trauma can precipitate severe bleeding, particularly in the absence of prophylactic therapy.

Case: A 6-year-old boy with moderate hemophilia A (factor VIII ~1.2%) developed an acute subdural hematoma (SDH) after minor head injury. He presented with progressive headache, vomiting, and decreased consciousness. Laboratory evaluation showed prolonged activated partial thromboplastin time consistent with factor VIII deficiency. Immediate factor VIII replacement was administered according to World Federation of Hemophilia (WFH) guidelines, along with adjunctive tranexamic acid. Cranial computed tomography revealed a large SDH with significant midline shift. The patient underwent emergent decompressive hemicraniectomy and hematoma evacuation with meticulous perioperative hemostatic management. Postoperatively, intensive care monitoring and sustained factor replacement maintained adequate hemostasis, resulting in stabilization and full neurological recovery without deficits.

Conclusion: This case demonstrates that life-threatening ICH can occur even in moderate hemophilia following trivial trauma. Early recognition, prompt factor replacement, and coordinated multidisciplinary management are critical to improving survival and neurological outcomes.

Keywords: Hemophilia A; Subdural hematoma; Paediatric neurosurgery; factor VIII; case report.

Introduction

Hemophilia A and B are rare X-linked recessive bleeding disorders caused by deficiencies of coagulation factor VIII and factor IX, respectively.[1] Although recurrent musculoskeletal bleeding is characteristic, intracranial hemorrhage (ICH) remains the most devastating complication, contributing significantly to mortality and long-term neurological disability.[2] Among ICH subtypes, subdural hematoma (SDH) is commonly reported and may occur after minor trauma or even spontaneously due to impaired hemostasis.[3]

The risk of ICH in hemophilia is markedly elevated compared to the general population, with incidence rates of 2.3–7.4 per 1,000 person-years, approximately 20–50 times higher than in unaffected individuals and a pooled neonatal incidence of 2.1%.[4,5] Lifetime prevalence ranges from 1.1% to 10.8%, influenced by disease severity and access to care.[4,6,7] Although severe hemophilia A accounts for a substantial proportion of cases, ICH also occurs in moderate disease, particularly in patients without prophylaxis. Additional risk factors include inhibitor development and delayed or inadequate treatment.[6,8] Despite therapeutic advances, ICH remains a leading cause of mortality, with pediatric case fatality rates of 20%–25%, especially in resource-limited settings.[4,8,9]

This case is notable for the occurrence of life-threatening SDH following trivial trauma in a child with moderate hemophilia A, underscoring that severe ICH is not limited to patients with severe disease.

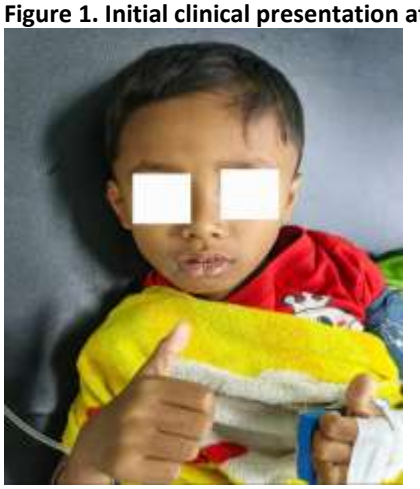
Case Report

A 6-year-old boy (weight 18 kg) was brought to the emergency department of a local hospital with severe headache and recurrent vomiting requiring urgent evaluation. His symptoms developed several hours after an accidental low-height fall at home one day prior. There was no reported loss of consciousness, seizure activity, or external signs of significant head trauma at the time of injury.

The patient had been diagnosed with moderate hemophilia A six months earlier, following episodes of easy bruising and gingival bleeding. Initial laboratory evaluation at diagnosis revealed a markedly prolonged activated partial thromboplastin time (aPTT) of 83.6 seconds, a normal prothrombin time of 10.4 seconds, and low factor VIII activity of approximately 1.2%, confirming moderate factor VIII deficiency. He was not on regular prophylactic factor VIII replacement therapy and had only received on-demand treatment for prior minor bleeding episodes, with good clinical response. There was no known family history of bleeding disorders or hemophilia. Psychosocial history was unremarkable, and there were no prior hospitalizations for major bleeding events.

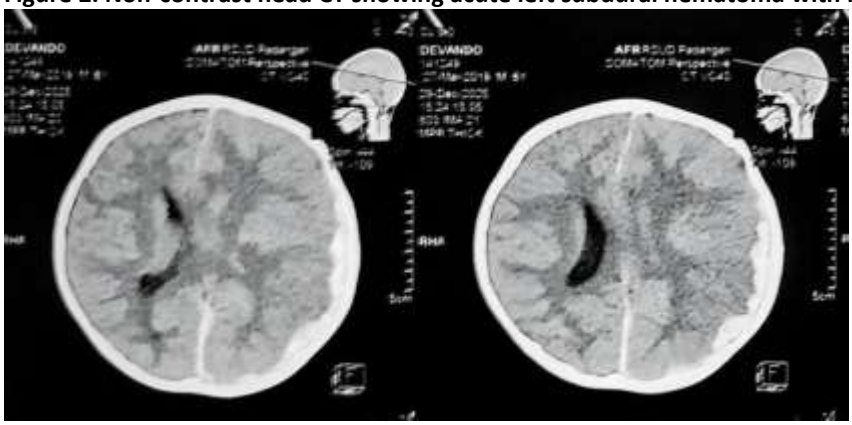
On initial examination, the patient appeared well-nourished, alert, and normocephalic, with no external signs of head trauma such as scalp hematoma or bleeding. Cardiopulmonary and abdominal examinations were unremarkable. Neurological assessment at presentation showed intact orientation without focal deficits. However, over several hours of observation, his neurological status deteriorated, with progressive drowsiness and reduced responsiveness. Upon transfer to our tertiary care center, he was lethargic, breathing spontaneously, and responsive only to verbal stimuli. Pupils were equal and reactive to light. A new right-sided hemiparesis was noted, indicating focal neurological involvement. Vital signs revealed low-grade fever at 37.9 °C, heart rate 98 beats/min, blood pressure 96/68 mmHg, and oxygen saturation 99% on room air, consistent with overall hemodynamic stability (Figure 1).

Figure 1. Initial clinical presentation at admission



Initial laboratory evaluation at the referring hospital demonstrated mild anemia with hemoglobin 10.6 g/dL, elevated platelet count 499 × 10^3/µL, slightly prolonged prothrombin time of 13.7 s, and markedly prolonged activated partial thromboplastin time of 56.2 s, consistent with underlying hemophilia A. On arrival at our center, hemoglobin was 10.2 g/dL with persistently prolonged aPTT of 68.1 s and mild hyponatremia of 129.6 mmol/L; other biochemical parameters were unremarkable. Given the history of head trauma and known coagulopathy, ICH was strongly suspected. Urgent non-contrast head CT confirmed an acute left parietal SDH, demonstrating a crescent-shaped hyperdense collection measuring approximately 14 mm in thickness, with significant mass effect, 9 mm rightward midline shift, and associated cerebral edema. No skull fractures or additional ICHs were identified.

Figure 2. Non-contrast head CT showing acute left subdural hematoma with midline shift



The primary diagnosis was traumatic acute SDH in the setting of hemophilia A. Alternative causes of decreased consciousness, including central nervous system infection and metabolic encephalopathy, were considered less likely. A key diagnostic and management challenge was limited access to factor VIII concentrate at the referring facility, necessitating transfer after initial supportive correction with fresh frozen plasma. Management prioritized rapid correction of coagulopathy and control of intracranial pressure. At the referring hospital, the patient received 1 unit of packed red blood cells and 2 units of fresh frozen plasma. Upon arrival at our center, 500 IU of factor VIII concentrate (~30 IU/kg) and intravenous tranexamic acid 250 mg were

administered immediately. Supportive therapy included intravenous fluids, acetaminophen, ondansetron, and mannitol 1 g/kg for intracranial pressure control.

Following initial stabilization, the patient underwent emergent left decompressive hemicraniectomy with evacuation of the SDH under general anesthesia (Figure 3). An additional 1000 IU of factor VIII (~60 IU/kg) was given preoperatively to achieve adequate hemostasis. Intraoperative blood loss was minimal (~50 mL) and controlled with standard measures; a small transfusion of packed red cells (~75 mL) was required. The procedure was completed without complications.

Figure 3. Intraoperative view showing evacuation of a large acute subdural clot (~10 mm)

Postoperatively, factor VIII replacement was continued with 500 IU every 12 hours for 3 days, tapered to 250 IU every 12 hours until day 7, and 250 IU daily until day 14 to maintain adequate hemostasis. Tranexamic acid was continued as adjunct therapy. The patient was managed in the ICU with close monitoring, head elevation, and supportive care, showing progressive neurological improvement without re-bleeding or thrombotic complications**.**

By postoperative day 14, the patient demonstrated complete clinical recovery and was discharged in stable condition. He was fully alert, interactive, and neurologically intact, with normal motor strength, independent ambulation, and no residual deficits. The surgical wound healed well without signs of infection. Follow-up laboratory evaluation showed improved hemoglobin of 11.1 g/dL and normalized aPTT of 32.4 s, indicating adequate correction of coagulopathy. Therapeutic adherence to factor VIII replacement and adjunctive treatment was maintained throughout hospitalization, with good tolerability and no adverse effects. No postoperative complications, including re-bleeding, thrombosis, or infection, were observed. The patient and family received education on hemophilia management, emphasizing early recognition of bleeding and injury prevention. Secondary prophylaxis with factor VIII was planned, and the patient was referred to a specialized hemophilia center for long-term care. At one-month follow-up, he remained asymptomatic with full return to normal activities and no neurological sequelae.

Discussion

This case illustrates a life-threatening ICH in a child with moderate hemophilia A following trivial head trauma, emphasizing that severe bleeding is not confined to patients with severe disease. Although moderate hemophilia is typically associated with bleeding after significant injury, even minor trauma can precipitate catastrophic hemorrhage.[10,11] SDH is a recognized complication in hemophilia and may occur after minimal trauma or even spontaneously due to impaired hemostasis.[3] Our patient's borderline factor VIII level (~1.2%) further highlights that the clinical phenotype may overlap with severe disease, necessitating similar vigilance and management.

A key strength of our approach was the immediate initiation of factor VIII replacement at the first suspicion of intracranial bleeding. Early correction of coagulopathy is critical to limit hematoma expansion and prevent irreversible brain injury, and delays or inadequate dosing have been associated with poorer outcomes.[2,8] The addition of tranexamic acid provided adjunctive stabilization of clot formation through antifibrinolytic effects.[12] However, a notable limitation was the lack of access to factor VIII concentrate at the referring facility, necessitating interim use of fresh frozen plasma. While this provides partial correction, it is suboptimal due to the large volumes required and difficulty achieving adequate factor levels.[2] This delay underscores the importance of timely access to appropriate hemostatic therapy, particularly in emergency settings.

Definitive management required urgent neurosurgical intervention following adequate hemostatic preparation. Decompressive hemicraniectomy with hematoma evacuation was performed after achieving near-normal factor levels using weight-based dosing, consistent with established replacement principles.[13] Surgical management in hemophilia patients presents unique challenges, as insufficient factor coverage can lead to uncontrolled bleeding, while excessive dosing may increase thrombotic risk. In this case, meticulous perioperative planning and close coordination between hematology, anesthesia, and neurosurgery teams enabled effective intraoperative hemostasis and minimal blood loss.[13,14] This multidisciplinary approach represents a major strength and was essential to the favorable outcome.

Postoperative management was equally critical. Sustained factor VIII replacement over two weeks ensured maintenance of adequate hemostatic levels during the period of highest re-bleeding risk and tissue repair.[2,13] The absence of re-bleeding or thrombotic complications in our patient reflects both appropriate dosing and close monitoring. Continued use of tranexamic acid further supported clot stability. These measures highlight the importance of structured postoperative protocols in preventing secondary complications and optimizing recovery.[2,15]

Comparison with previously reported cases reveals consistent themes regarding the importance of early intervention and coordinated care. Many reports describe ICH in hemophilia following minor trauma, often with initially subtle clinical findings but rapid neurological deterioration.[4,8] Our case aligns with these observations, as the patient initially appeared stable before developing progressive neurological decline. This reinforces the need for a high index of suspicion and early imaging in any hemophilia patient with head injury, regardless of initial presentation. Additionally, the absence of factor VIII inhibitors in this patient simplified management, as inhibitor development is associated with treatment resistance and worse outcomes.[11]

This case also highlights important considerations for long-term management. The absence of prior prophylaxis likely contributed to the severity of bleeding in this patient. Initiation of secondary prophylaxis following recovery is essential to reduce the risk of recurrence. Advances in hemophilia therapy, including extended half-life factor VIII products and non-replacement therapies such as emicizumab, provide additional options to improve adherence and long-term outcomes.[1] Furthermore, comprehensive follow-up in a specialized hemophilia center is critical to ensure optimal management, monitor for inhibitor development, and provide education on injury prevention.

Patient Perspective

As the patient is a young child, a first-person perspective is unavailable. His parents reported initial fear followed by relief after recovery, and emphasized increased awareness of hemophilia management, with commitment to prophylaxis, injury prevention, and prompt care after any head trauma.

Informed Consent

Written informed consent for anonymous publication of this case and accompanying images was obtained from the patient's parents.

Conclusion

This case demonstrates that severe ICH can occur in moderate hemophilia A following trivial trauma. Early recognition, immediate factor replacement, and rapid multidisciplinary intervention are essential to achieving favorable outcomes. Any head injury in a patient with hemophilia should be treated as a medical emergency, regardless of initial clinical appearance.

Disclosure

Conflict of Interest:

The authors declare that the study was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Author Contributions:

HH and PK contributed to the conception, data collection, analysis, and drafting of the manuscript. All authors contributed to and approved the final manuscript.

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References

1. Mancuso M, Santagostino E. Outcome of Clinical Trials with New Extended Half-Life FVIII/IX Concentrates. Journal of Clinical Medicine. 2017;6(4):39. doi:https://doi.org/10.3390/jcm6040039
2. Srivastava A, Santagostino E, Dougall A, et al. WFH Guidelines for the Management of Hemophilia, 3rd Edition. Haemophilia. 2020;26(s6). doi:https://doi.org/10.1111/hae.14046
3. Stieltjes N, Calvez T, Demiguel V, et al. Intracranial Haemorrhages in French Haemophilia Patients (1991-2001): Clinical presentation, Management and Prognosis Factors for Death. Haemophilia. 2005;11(5):452-458. doi:https://doi.org/10.1111/j.1365-2516.2005.01090.x
4. Zwagemaker AF, Gouw SC, Jansen JS, et al. Incidence and Mortality Rates of Intracranial Hemorrhage in Hemophilia: a Systematic Review and Meta-analysis. Blood. 2021;138(26):2853-2873. doi:https://doi.org/10.1182/blood.2021011849
5. Shapiro AD, Hardesty BM, Peyvandi F, Iorio A. Prevalence of Selected Bleeding and Thrombotic Events in Persons with Hemophilia versus the General population: a Scoping Review. Research and Practice in Thrombosis and Haemostasis. 2023;7(1):100007-100007. doi:https://doi.org/10.1016/j.rpth.2022.100007
6. Hu J, Chandler M, Manuel C, et al. Risk of Intracranial Hemorrhage in Persons with Hemophilia A in the United States: Real-World Retrospective Cohort Study Using the ATHNdataset. Journal of Blood Medicine. 2024;15:191-205. doi:https://doi.org/10.2147/jbm.s443380
7. Gringeri A, Mannucci PM, Gringeri M, Castaman G, Peyvandi F. Global Estimation of the Bleeding Episodes Treatable with Desmopressin in Von Willebrand Disease and Hemophilia A. Haematologica. 2025;110(8). doi:https://doi.org/10.3324/haematol.2025.287572
8. Zanon E, Pasca S. Intracranial Haemorrhage in Children and Adults with Haemophilia A and B: a Literature Review of the Last 20 Years. Blood Transfusion. 2019;17(5):378-384. doi:https://doi.org/10.2450/2019.0253-18
9. Chalmers EA, Alamelu J, Collins PW, et al. Intracranial Haemorrhage in Children with Inherited Bleeding Disorders in the UK 2003-2015: a National Cohort Study. Haemophilia. 2018;24(4):641-647. doi:https://doi.org/10.1111/hae.13461
10. Blanchette VS, Key NS, Ljung LR, Manco-Johnson MJ, van den Berg HM, Srivastava A. Definitions in Hemophilia: Communication from the SSC of the ISTH. Journal of Thrombosis and Haemostasis. 2014;12(11):1935-1939. doi:https://doi.org/10.1111/jth.12672
11. Doncel SS, Mosquera GAD, Cortes JM, Rico CA, Cadavid FJM, Peláez RG. Haemophilia A: a Review of Clinical Manifestations, Treatment, Mutations, and the Development of Inhibitors. Hematology Reports. 2023;15(1):130-150. doi:https://doi.org/10.3390/hematolrep15010014
12. Poston JN, Kruse-Jarres R. Perioperative Hemostasis for Patients with Hemophilia. Hematology / the Education Program of the American Society of Hematology. 2022;2022(1):586-593. doi:https://doi.org/10.1182/hematology.2022000387
13. Lowell AE, Calgi MP, Caruso JJ, Man LM, McNeil JS. Perioperative Management of Hemophilia Patients. Current Anesthesiology Reports. 2024;14(3):354-365. doi:https://doi.org/10.1007/s40140-024-00635-y
14. Kwak J, Mazzeffi M, Boggio LN, Simpson ML, Tanaka KA. Hemophilia: a Review of Perioperative Management for Cardiac Surgery. Journal of Cardiothoracic and Vascular Anesthesia. 2020;36(1):246-257. doi:https://doi.org/10.1053/j.jvca.2020.09.118
15. Esposto MP, Bonetti E, Balter R, et al. Changes over the Last Two Decades in Hemophilia A and B Management in a Tertiary Care Pediatric Centre. Clinical Epidemiology and Global Health. 2024;29:101762-101762. doi:https://doi.org/10.1016/j.cegh.2024.101762