A Rare Case of Acquired Hemophilia A in a Patient with Suspected Plasma Cell Dyscrasia

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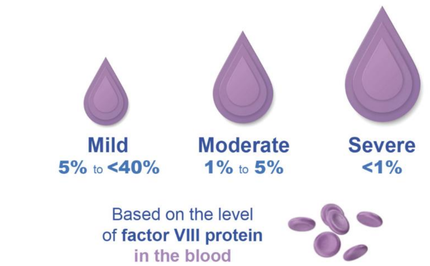
Introduction

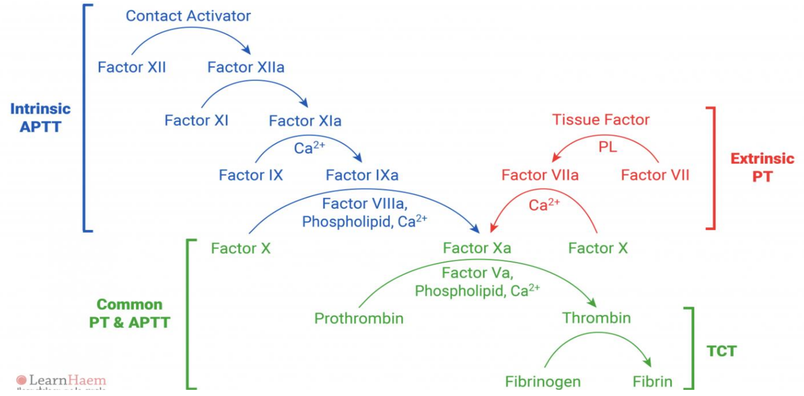
Acquired hemophilia A (AHA) is a rare but life-threatening autoimmune bleeding disorder caused by autoantibodies against factor VIII. With an annual incidence of ~1.5 cases per million, more than half of the cases are idiopathic, while the remainder are associated with malignancy, autoimmune disease, postpartum states, or medications.[2] Mortality is often related to delayed diagnosis, severe bleeding, or complications of immunosuppression. Hemostatic control typically requires bypassing agents such as recombinant activated factor VII (rFVIIa) such as NovoSeven or activated prothrombin complex concentrate (FEIBA). Additional inhibitor eradication is needed and relies on immunosuppressive therapy with corticosteroids alone or in combination with rituximab or cyclophosphamide.[5] Recent registry analyses (EACH2, GTH) confirm that early use of combination therapy significantly improves remission rates and survival.[4] This case highlights the diagnostic and therapeutic challenges of AHA in an elderly patient with suspected plasma cell dyscrasia.



https://www.alturviio.com/hemophilia-education/factor-activity-levels/high-and-low-activity-levels

Case Presentation

A 66-year-old female with hypertension, type 2 diabetes, and atrial fibrillation presented to the hospital with lower extremity weakness, back pain, and a large right hip hematoma. Imaging revealed diffuse skeletal lytic lesions, concerning for malignancy or plasma cell dyscrasia. Complete blood count showed anemia with Hgb 6.8 and coagulation studies showed prolonged aPTT that did not correct with mixing, leading to further hematologic workup. A plasma cell dyscrasia workup and tumor markers were negative. She was found to have **Factor VIII inhibitor titers of 189 Bethesda units** (normal < 0.5 BU) and **Factor VIII activity of 2%** (normal 50-150%), consistent with acquired hemophilia A. She was started on FIEBA (Factor Eight Inhibitor Bypassing Activity), prednisone, and rituximab. She was discharged on a prednisone taper, rituximab, and atovaquone for Pneumocystis jirovecii (PCP) prophylaxis. Factor VIII levels improved to 58% after immunosuppressive therapy.



https://www.learnhaem.com/courses/coagulation/lessons/normal-haemostasis/topic/the-revised-coagulation-cascade/

Clinical Implications

- AHA should be considered in elderly patients with new onset isolated aPTT prolongation and spontaneous bleeding
- Management of AHA requires both hemostatic control (FEIBA, recombinant factor VIIa NovoSeven, NovoEight, or emicizumab in select cases)[1] and immunosuppression (steroids, rituximab, cyclophosphamide)[3]
- Registry data demonstrates that combination therapy shortens inhibitor eradication time compared to only corticosteroids[1]
- Long-term outcomes and relapse prevention depend on both clinical complexity and psychosocial barriers (non-compliance, steroid toxicity, drug side effects)

Factor Eight Inhibitor Bypassing Activity

- FIEBA is an activated prothrombin complex concentrate (aPCC), → contains a mixture of vitamin K–dependent clotting factors (II, VIIa, IX, and X)
- In AHA, autoantibodies neutralize factor VIII, blocking the intrinsic pathway at the FVIII–FIXa step, so normal clot formation cannot occur
- FEIBA bypasses the need for factor VIII by directly supplying downstream clotting factors, allowing generation of thrombin, fibrin clot formation, and hemostasis

Key points:

- Works independent of FVIII → effective even when inhibitors are present
- Rapid onset of action, usually within hours
- Used for treatment of acute bleeding episodes or perioperatively
- Risks: can predispose to thrombosis if overused, so dosing is carefully monitored

Conclusion

Acquired hemophilia A should be included in the differential for elderly patients with spontaneous bleeding and prolonged aPTT. Plasma cell dyscrasia and autoimmune disease remain important associated conditions. Early recognition, immunosuppressive therapy, and multidisciplinary care are key to improving outcomes.

References

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