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Review Article

Acquired Hemophilia a Presenting as Persistent Post-Traumatic Orofacial Bleeding Following Minor Trauma in a Nonagenarian Woman: A Rare Case Report

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ABSTRACT

Acquired hemophilia A (AHA) is a rare autoimmune bleeding disorder caused by the development of autoantibodies against coagulation factor VIII.[1] It predominantly affects the elderly and may present with life-threatening bleeding despite the absence of a prior bleeding history. We report the case of a 92-year-old woman who presented with persistent bleeding following minor facial trauma after a fall. Initial local measures and suturing failed to achieve hemostasis. Laboratory evaluation demonstrated isolated prolongation of activated partial thromboplastin time (aPTT), low hemoglobin levels requiring repeated blood transfusions, and a markedly elevated factor VIII inhibitor titre detected by Bethesda assay. Imaging revealed bilateral mandibular condylar fracture-dislocation secondary to trauma. The patient was subsequently diagnosed with acquired hemophilia A. This case highlights the importance of considering acquired coagulation factor inhibitors in elderly patients presenting with disproportionate bleeding following minor trauma, particularly when routine hemostatic measures fail. Early recognition and multidisciplinary management are crucial to reduce morbidity and mortality.

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INTRODUCTION

Acquired hemophilia A (AHA) is a rare autoimmune bleeding disorder caused by the development of autoantibodies against coagulation factor VIII, resulting in impaired intrinsic coagulation and potentially severe hemorrhage. The estimated annual incidence is approximately 1–1.5 cases per million population, with a bimodal age distribution and a peak incidence among elderly individuals.[1,2] Unlike congenital hemophilia, affected patients typically have no personal or family history of bleeding disorders.

Clinical manifestations range from spontaneous ecchymoses and soft-tissue hematomas to life-threatening mucosal, gastrointestinal, retroperitoneal, or postoperative bleeding.[1,2] Diagnosis is often delayed because routine coagulation screening may reveal only an isolated prolongation of activated partial thromboplastin time (aPTT), which can be overlooked in the setting of trauma or surgical bleeding.[1] Confirmation requires demonstration of reduced factor VIII activity and detection of factor VIII inhibitors using the Bethesda assay.

Mortality rates ranging from 8% to 22% have been reported, largely due to severe bleeding complications and delayed recognition.[1,2] Early diagnosis and prompt initiation of hemostatic and immunosuppressive therapy are therefore essential to improve outcomes.We report a case of acquired hemophilia A in a 92-year-old woman who presented with persistent bleeding following minor facial trauma associated with bilateral mandibular condylar fracture-dislocation. This case highlights the importance of considering acquired coagulation factor inhibitors in elderly patients presenting with disproportionate bleeding and isolated prolonged aPTT despite appropriate local hemostatic measures, thereby facilitating timely diagnosis and management.

CASE REPORT

A 92-year-old female presented with persistent bleeding following a slip-and-fall injury at home approximately 12 days before admission. The injury resulted in a lip wound with continuous bleeding that was difficult to control. There was no documented personal or family history of bleeding disorders, and no history of previous abnormal bleeding episodes.

At presentation, the patient had ongoing bleeding from the injury site with features suggestive of significant blood loss. She required hospitalization and multidisciplinary assessment.

Initial laboratory investigations revealed:

- Hemoglobin: approximately 6.1 g/dL • PT/INR: within normal limits • Markedly prolonged aPTT: approximately 112.7 seconds • Peripheral smear: dimorphic anemia with mild neutrophilic leukocytosis

Because of isolated prolongation of aPTT, a mixing study was performed. The prolonged aPTT failed to correct after mixing with normal plasma, indicating the presence of a circulating inhibitor.

Further coagulation studies demonstrated: • Reduced factor VIII activity • Positive factor VIII inhibitor assay • Bethesda inhibitor titer: approximately 25.6 Bethesda Units

These findings established the diagnosis of acquired hemophilia A.

TherapeuticIntervention: The patient received supportive management including blood transfusion and local wound care. Multiple specialties, including hematology and surgery, were involved in management. Hematology consultation recommended hemostatic therapy and long-term inhibitor eradication strategies using immunosuppressive treatment according to standard AHA management principles.[5]

Follow-up and Outcomes: Bleeding stabilized during hospitalization, and the patient was discharged with follow-up recommendations and hematology review for definitive long-term management of the inhibitor.

DISCUSSION

Acquired hemophilia A is an uncommon but severe bleeding disorder that should be suspected in elderly patients presenting with unexplained bleeding and isolated prolongation of aPTT. Unlike congenital hemophilia, bleeding into joints is uncommon; instead, mucocutaneous bleeding, soft tissue hemorrhage, and post-traumatic bleeding predominate.

The hallmark laboratory finding is isolated prolonged aPTT with normal PT/INR. Mixing studies are essential in differentiating coagulation factor deficiency from inhibitor-mediated coagulopathy.[1,3] Failure of correction strongly suggests the presence of an inhibitor. Confirmation requires measurement of factor VIII activity and inhibitor quantification using the Bethesda assay.[1]

The inhibitor titer in this patient was high (approximately 25.6 BU), indicating clinically significant inhibitor activity. Such patients are at substantial risk for severe and potentially fatal bleeding. Current international recommendations advocate rapid initiation of hemostatic therapy using bypassing agents or recombinant porcine factor VIII, alongside immunosuppressive therapy aimed at inhibitor eradication.[1] Corticosteroids alone or in combination with cyclophosphamide or rituximab are commonly employed.[1,5]

This case emphasizes the diagnostic value of coagulation studies in elderly patients with persistent bleeding after seemingly minor trauma. Recognition of isolated prolonged aPTT and appropriate referral for hematological evaluation enabled definitive diagnosis of acquired hemophilia A.

CONCLUSION

Acquired hemophilia A is a rare but potentially life-threatening autoimmune bleeding disorder that should be considered in elderly patients presenting with unexplained, persistent, or disproportionate bleeding, particularly in the absence of a personal or family history of bleeding disorders. The presence of an isolated prolonged activated partial thromboplastin time (aPTT) with a normal prothrombin time should prompt further investigation for coagulation factor deficiencies or inhibitors. Mixing studies play a pivotal role in differentiating factor deficiencies from inhibitor-mediated coagulopathies, while factor VIII activity measurement and Bethesda inhibitor assay remain essential for definitive diagnosis.

This case underscores the diagnostic challenge posed by acquired hemophilia A, especially when bleeding occurs following trauma and may initially be attributed solely to local injury. Failure of conventional hemostatic measures and ongoing transfusion requirements should raise suspicion for an underlying coagulation disorder. Early recognition, prompt hematological evaluation, and timely initiation of appropriate hemostatic therapy and immunosuppressive treatment are critical to controlling bleeding, eradicating inhibitors, and reducing morbidity and mortality.[1,4]

Furthermore, this report highlights the importance of multidisciplinary collaboration among emergency physicians, surgeons, hematologists, and transfusion medicine specialists in the management of patients with acquired hemophilia A. Increased awareness of this rare condition may facilitate earlier diagnosis and improve clinical outcomes, particularly in vulnerable elderly populations.[7]

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