Acquired Hemophilia A: A diagnostic challenge across distinct clinical settings

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INTRODUCTION

Acquired hemophilia A (AHA) is a rare but potentially life-threatening autoimmune bleeding disorder caused by the development of inhibitory autoantibodies against coagulation factor VIII. It typically presents with sudden, severe bleeding in patients without prior personal or family history of hemorrhagic disorders.

AHA occurs in various clinical settings, including autoimmune diseases, malignancies, infections, and the postpartum period, although a significant proportion of cases remain idiopathic. Early recognition is critical, as delayed diagnosis may lead to inappropriate management and increased morbidity.

CASES

Patient 1 – Post operative context

A 70-year-old man presented with postoperative bleeding following inguinal hernia repair, associated with melena and anemia, without prior bleeding history.

Laboratory evaluation showed isolated prolonged aPTT with reduced factor VIII levels and a circulating inhibitor, confirming acquired hemophilia A in the context of chronic hepatitis B infection.

Treatment with bypassing agents, corticosteroids, and antiviral therapy led to a favorable outcome.

Patient 2 – Autoimmune context

A 47-year-old woman developed progressive spontaneous ecchymoses. Initial suspicion of deep vein thrombosis led to anticoagulation and clinical worsening.

Further investigations revealed isolated prolonged aPTT, low factor VIII levels, and a circulating inhibitor in an autoimmune context.

Management included discontinuation of anticoagulation and initiation of bypassing therapy and corticosteroids, resulting in clinical improvement.

Patient 3 – Postpartum context

A 33-year-old woman presented with severe postpartum hemorrhagic syndrome complicated by hemoperitoneum and hemorrhagic shock. Physical examination revealed diffuse spontaneous hematomas (figure A, B and C).

Laboratory evaluation showed isolated prolonged aPTT with markedly reduced factor VIII levels and a circulating inhibitor, confirming acquired hemophilia A.

Despite initial management, the patient required intensive care and surgical intervention with evacuation of approximately 3 liters of blood. Treatment included bypassing agents and immunosuppressive therapy with corticosteroids, cyclophosphamide, and rituximab, leading to progressive clinical and biological improvement.

PATIENT	CLINICAL CONTEXT	PRESENTATION	KEY LABORATORY FINDINGS	TREATMENT	OUTCOME
1	Postoperative + HBV	• Melena • Anemia	• ↑ aPTT (ratio ~2.24) • ↓ Factor VIII (8%) • Inhibitor positive (7 Bethesda Units) • PT and platelets normal	• PCC (bypassing agent) • Corticosteroids • Antiviral therapy (tenofovir) • Transfusion support	Favorable
2	Autoimmune (DVT misdiagnosis)	• Progressive ecchymoses • Soft tissue bleeding	• ↑ aPTT • ↓ Factor VIII • Inhibitor positive • PT and platelets normal	• PCC (bypassing agent) • Corticosteroids • Discontinuation of anticoagulation	Favorable
3	Postpartum	• Diffuse hematomas • Hemoperitoneum • Hemorrhagic shock	• ↑ aPTT • ↓ Factor VIII • Inhibitor positive • PT and platelets normal	• rFVIIa (bypassing agent) • Corticosteroids • Cyclophosphamide • Rituximab • Supportive care	Favorable

All cases shared a characteristic profile of acquired hemophilia A, with isolated prolonged aPTT, reduced factor VIII levels, and circulating inhibitors, despite occurring in distinct clinical contexts.

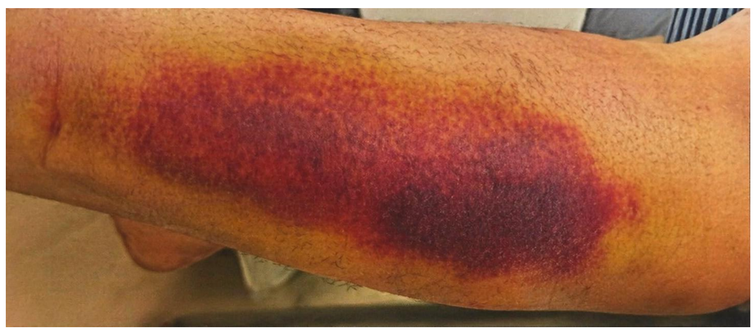

Figure A : Hematoma at the level of the right arm in our patient

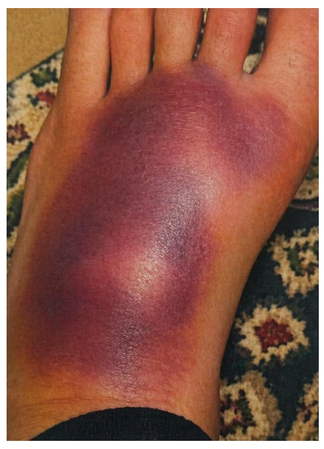

Figure B : Hematoma with edema at the level of the left foot in our patient

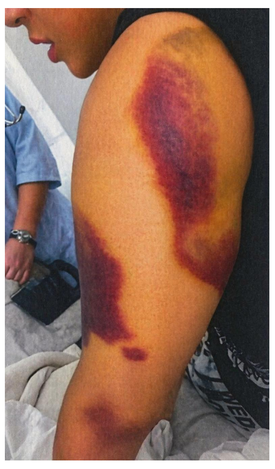

Figure C : Hematoma at the level of the left arm in our patient

DISCUSSION

Acquired hemophilia A (AHA) is a rare autoimmune bleeding disorder caused by the development of autoantibodies directed against factor VIII, leading to impaired coagulation and potentially life-threatening hemorrhage. It typically occurs in patients without prior bleeding history and may be associated with a wide range of conditions, including autoimmune diseases, malignancies, infections, or the postpartum period, although up to 50% of cases remain idiopathic.

Clinically, AHA is characterized by spontaneous and often severe bleeding, predominantly involving soft tissues, muscles, or mucous membranes, rather than hemarthrosis, which is more typical of congenital hemophilia. The diagnosis should be suspected in the presence of isolated prolonged aPTT that does not correct on mixing studies, and confirmed by reduced factor VIII levels with detection of circulating inhibitors.

Our case series illustrates the heterogeneity of clinical contexts and presentations, ranging from postoperative bleeding to autoimmune and postpartum forms, as well as the potential for diagnostic delay or misdiagnosis, which may expose patients to inappropriate treatments such as anticoagulation.

Management relies on a dual approach combining rapid hemostatic control using bypassing agents and eradication of the inhibitor through immunosuppressive therapy, including corticosteroids, cyclophosphamide, or rituximab in refractory cases. Early recognition and prompt treatment are essential to reduce morbidity and mortality.

CONCLUSION

Acquired hemophilia A is a rare but potentially life-threatening bleeding disorder that should be suspected in any patient with unexplained hemorrhage and isolated prolonged aPTT.

Our series highlights the clinical heterogeneity, diagnostic challenges, and risk of mismanagement, emphasizing the importance of early recognition. Prompt initiation of appropriate hemostatic and immunosuppressive therapy is essential to improve patient outcomes.

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