

ACQUIRED HEMOPHILIA A WITH HIGH-TITER INHIBITOR IN A PATIENT WITH CARDIOVASCULAR AND METABOLIC COMORBIDITIES.

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Background

Acquired hemophilia A (AHA) is a rare autoimmune coagulopathy characterized by the development of neutralizing autoantibodies against factor VIII (FVIII). The clinical presentation can be severe, with spontaneous and recurrent bleeding, often requiring multidisciplinary management, particularly in patients with multiple comorbidities.

Case Report

A 74-year-old female patient was referred to our unit for recurrent acute anemia (Hb 3 g/dL) and diffuse mucocutaneous and muscular hematomas, especially in the bilateral subscapular regions with low perfusion. The hemorrhagic picture triggered episodes of sustained ventricular tachyarrhythmia, resulting in activation of a previously implanted single-chamber ICD, placed for dilated hypokinetic cardiomyopathy of hypertensive-valvular origin (moderate mitral regurgitation).

Past medical history included insulin-dependent type II diabetes mellitus with stage III chronic kidney disease, non-progressive steatotic hepatomegaly, euthyroid uninodular hyperplasia, and a long-standing calcified uterine fibroid. FVIII inhibitor testing was performed, revealing FVIII activity at 8% and an inhibitor titer of 8 BU/mL, leading to a diagnosis of acquired hemophilia A.

The patient received intensive hemostatic treatment with recombinant activated factor VII (rFVIIa), totaling 35 vials (5 mg), which resulted in bleeding control and progressive reduction of transfusion needs (15 units of packed red blood cells). Concurrently, corticosteroid immunosuppressive therapy was initiated at 1 mg/kg, with a complete response achieved: inhibitor eradication and normalization of FVIII levels. At discharge, the patient showed hemodynamic stability, no further arrhythmic episodes, resolution of hematomas, and partial functional recovery.

Conclusion

AHA may present acutely and severely, even in patients without overt autoimmune disease. Management requires a multidisciplinary approach, individualized therapy, and close monitoring of comorbidities.

This case highlights the complexity of managing AHA in elderly patients with advanced cardiac disease and chronic metabolic disorders. The use of bypassing agents such as rFVIIa proved effective and safe even in a fragile cardiologic context, with careful monitoring. Prompt initiation of immunosuppressive therapy was crucial, allowing for hematologic recovery without significant infectious or metabolic complications.

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