



DISORDINI EMORRAGICI ACQUISITI: ASPETTI
DIAGNOSTICI E CLINICI

ACQUIRED HEMOPHILIA A IN THE SURGICAL PATIENT: DIAGNOSTIC AND THERAPEUTIC COMPLEXITY

M. Luongo¹, P. Massaro¹, M. Di Mascio¹, P.G. Simeone², F. Santilli², S. Verna¹

¹Immunohematology Service, Asl 02 Abruzzo, Chieti, Italy; ²Department of Medicine and Aging Sciences, and Center for Advanced Studies and Technology, "G. d'Annunzio" University of Chieti-Pescara, Italy

Background. Acquired hemophilia A (AHA) is a rare, potentially life-threatening autoimmune coagulopathy characterized by the presence of autoantibodies directed against factor VIII (FVIII). The disease often arises in complex clinical settings, posing significant diagnostic and therapeutic challenges. Diagnostic delay is common, particularly in patients with concomitant thrombotic risk or those receiving anticoagulant therapy—conditions that may hinder the correct interpretation of coagulation tests.

Case Report. We report the case of an 81-year-old patient with multiple comorbidities (diabetes, ischemic heart disease, a biological prosthetic valve, and peripheral artery disease) on antiplatelet therapy, who underwent emergency surgery for intestinal obstruction complicated by hemoperitoneum and ileal ischemia. Given the high thrombotic risk, heparin prophylaxis was initiated after discontinuation of aspirin. The postoperative course was complicated by diffuse bleeding from surgical sites and subcutaneous tissue, disproportionate to the ongoing antithrombotic therapy, accompanied by severe anemia requiring transfusion. Laboratory tests revealed a prolonged aPTT (65 s), a normal prothrombin time (PT), FVIII activity of 0.7%, and an anti-FVIII inhibitor titer of 5.5 BU, consistent with a diagnosis of AHA. Initial treatment with recombinant activated factor VII (rFVIIa) (subsequently replaced by activated prothrombin complex concentrate), corticosteroids, and cyclophosphamide failed

to produce a significant clinical or laboratory response. The patient exhibited persistent bleeding, anemia, and progressive clinical deterioration, requiring continuous transfusion support. Following a decline in renal function, cyclophosphamide was discontinued. During follow-up, an abdominal CT scan revealed a splenic infarction. Suspecting endocarditis involving the prosthetic valve, blood cultures were performed and were positive for *Candida* spp., indicating fungal sepsis. Given the presence of systemic infection, rituximab was contraindicated, rescue therapy with cyclosporine and intravenous immunoglobulins (IVIG) was initiated resulting in a reduction of the inhibitor (1.19 BU) and an increase in FVIII levels (15%). Despite hematological improvement, the patient died of septic shock.

Conclusions. In patients presenting with bleeding disproportionate to the clinical context, particularly in the postoperative setting, AHA should be strongly considered. Concomitant anticoagulation and an underlying thrombotic risk profile may contribute to diagnostic delays. Therapeutic decisions must be individualized, and aggressive upfront immunosuppressive approaches should be used cautiously in frail patients, as their toxicity and associated infectious risk may limit subsequent treatment options. A multidisciplinary approach is essential to balance hemostatic efficacy with the risks of immunosuppression, as infectious complications represent a critical determinant of prognosis.