

WHEN BLEEDING MIMICS THROMBOSIS: DIAGNOSTIC DELAY AND THROMBOTIC COMPLICATIONS IN ACQUIRED HEMOPHILIA A

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Background. Acquired hemophilia A (AHA) is a rare autoimmune bleeding disorder caused by neutralizing anti-FVIII antibodies. Its recognition may be delayed when acute limb swelling mimics venous thrombosis, exposing patients to potentially harmful anticoagulation. We report a case of AHA associated with systemic lupus erythematosus, initially misinterpreted as upper-limb thrombosis and later complicated by catheter-related femoral thrombosis.

Case Report. A 62-year-old Caucasian man with recently diagnosed systemic lupus erythematosus presented to another emergency department with progressive swelling of the left arm. Suspected deep vein thrombosis led to low-molecular-weight heparin administration before transfer. Initial tests showed severe anemia (Hb 7.8 g/dL), isolated prolonged aPTT (85 s) and normal INR. At our center, CT angiography documented a large left arm/axillary/pectoral hematoma (6 x 5.3 x 11.7 cm) with active arterial contrast extravasation. Coagulation work-up showed FVIII:C 0.2%, normal VWF antigen/activity, a mixing test consistent with an inhibitor and anti-FVIII inhibitor titre of 25.92 BU/mL, confirming AHA. Antiphospholipid antibodies, viral serology and tumor markers were negative. Haemostasis was achieved with activated prothrombin complex concentrate, escalated from 3000 IU to 4000 IU three times daily because of limb progression and then reduced after clinical and hemoglobin

stabilization. Emergency embolization of the bleeding site was also performed. Inhibitor eradication included methylprednisolone 1 mg/kg/day, cyclophosphamide and low-dose rituximab (100 mg weekly for four doses). During hospitalization, after femoral vascular access for embolization, the patient developed painful right leg swelling. Ultrasound and CT angiography showed right femoral deep vein thrombosis with femoral pseudoaneurysm (25 x 16 x 45 mm). Since full-dose anticoagulation was contraindicated by life-threatening bleeding, he received submaximal low-molecular-weight heparin, ultrasound-guided thrombin embolization of the pseudoaneurysm and temporary inferior vena cava filter placement. At discharge, FVIII:C was still <10%, while inhibitor titre had decreased to 1.56 BU/mL. After rituximab completion and steroid tapering, FVIII:C increased to 79% with undetectable inhibitor on March 27, 2026. Anticoagulation was switched to dabigatran and the cava filter was removed uneventfully on April 1, 2026.

Conclusions. This case illustrates the diagnostic and therapeutic challenges of acquired hemophilia A, where delayed recognition may expose patients to inappropriate anticoagulation while subsequent invasive procedures may increase thrombotic risk. Early diagnosis and multidisciplinary management remain crucial to successfully navigate the bleeding-thrombosis balance