

Acquired Hemophilia A manifested as severe bleeding after invasive procedure

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Introduction: Acquired Hemophilia A (AHA) is a rare disease caused by the depletion of coagulation factor VIII due to autoantibodies. Factor VIII inhibitory antibodies neutralize the activities of FVIII to cause bleeding, and it can lead to life-threatening bleeding or death, with a high mortality rate of 9-22%. Here we report two cases of AHA, manifested as severe bleeding after invasive procedure.

Case 1: A 48-year-old female with systemic lupus erythematosus had polypectomy for endometrial polyp. She had severe vaginal bleeding continued for 1 week after the procedure. Laboratory tests revealed prolonged aPTT, not corrected by mixing test, and low factor VIII activity 1%. The Factor VIII antibody was 18 BU/mL. Upon diagnosis, recombinant activated factor VII (rFVIIa) was given every 3 hours as a hemostatic treatment and prednisolone (PL) 1 mg/kg as an immunosuppressive treatment. 5 days after the start of treatment, the bleeding stopped, and the patients was taking reduced dose of PL. **Case 2:** A 36-year-old female was diagnosed with deep vein thrombosis of lower leg. During the course, she complained of chest discomfort, and received coronary angiography. After angiography, left femoral puncture site hematoma developed, and left common femoral artery stent was inserted through right femoral puncture. However, both femoral site bruise aggravated. Laboratory study revealed prolonged aPTT, not corrected by mixing test, and low factor VIII activity 1%. The Factor VIII antibody was 37 BU/mL. The patient was treated with rFVIIa and achieved hemostasis. To eradicate the antibody, PL was used initially and PL combined with cyclophosphamide was used as second line therapy. Antibody disappeared but relapsed without bleeding symptom after 3 months later. Rituximab (375 mg/m² q 1 week, 4 doses) was used and complete response was established.

Conclusion: We report 2 cases of AHA, both first manifested as severe bleeding after invasive procedure. Before every invasive procedure, coagulation profiled should be checked, and prolonged aPTT with normal PT should be always further evaluated before procedure to avoid procedure related hemorrhagic events.

