

ACQUIRED VON WILLEBRAND SYNDROME IN AN ELDERLY PATIENT WITH EARLY-STAGE B-CLL

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Background. Acquired von Willebrand syndrome (AvWS) is a rare bleeding disorder commonly associated with lymphoproliferative, myeloproliferative, cardiovascular, or autoimmune diseases and may mimic inherited von Willebrand disease. Recognition of this condition is particularly relevant in elderly patients presenting with new-onset bleeding in the absence of a family history of congenital bleeding disorders.

Case Report. In August 2020, an 88-year-old man was admitted for extensive cutaneous bleeding and muscular hematomas. His history included B-CLL Rai stage A diagnosed in 2004 without signs of progression, benign prostatic hyperplasia, and hypertension. Baseline blood tests showed hemoglobin 12.2 g/dL, white blood cells $19.77 \times 10^9/L$ (lymphocyte predominant), platelets $219 \times 10^9/L$, normal creatinine and LDH, PT ratio 1.0, prolonged aPTT ratio 1.21, antithrombin 89%, and D-dimer 250 ng/mL; coagulation tests in 2015 had been normal. Further hemostatic work-up revealed FVIII:C 29.5%, vWF:RCo 4.5%, vWF:Ag 17.1%, vWF:RCo/vWF:Ag ratio of 0.26; no FVIII inhibitor was detected. Family screening was negative for bleeding abnormalities.

Additional evaluation (ANA, CT scan, serum protein electrophoresis, transthoracic echocardiography) excluded autoimmune, malignant or valvular causes. Because of late onset, negative family screening, previously normal coagulation tests, and underlying B-CLL, a diagnosis of AvWS was established. Acute management included tranexamic acid and recombinant activated FVII 90 mcg/kg every 4 hours with gradual discontinuation after one week, intravenous methylprednisolone followed by oral prednisone tapering, and supportive care. In September 2020, intravenous immunoglobulins every 4 weeks were started and are still ongoing. The patient has experienced no further bleeding manifestations.

Conclusions. AvWS should be considered in elderly patients with new bleeding symptoms and laboratory evidence of combined FVIII/VWF reduction, especially in the presence of lymphoproliferative disease. Prompt hemostatic control and an etiologic diagnostic work-up are essential, while IVIG may represent an effective maintenance option in selected patients.