Acquired von Willebrand disease (AvWD) refers to defects in von Willebrand factor (vWF) concentration, structure, or function that are not inherited directly, but are consequences of other medical disorders. Laboratory findings in AvWD are similar to those in von Willebrand disease and may include decreased values for vWF:Ag, vWF:RCo or FVIII. AvWD is usually caused by one of these mechanisms: autoimmune clearance or inhibition of vWF, increased shear-induced proteolysis of vWF or increased binding of vWF to platelets or other cell surfaces, which can consume large vWF multimers. An inverse relationship exists between the platelet count and vWF multimer size, probably because increased encounters with platelets promote increased cleavage of vWF by ADAMTS13. This mechanism probably accounts for AvWD associated with myeloproliferative disorders, such as chronic myeloid leukemia (CML).

We report you the case of a 55-year-old woman diagnosed with CML – accelerated phase, without any history of significant bleeding. After the bone marrow biopsy, she had an excessive local bleeding with a muscular hematoma of 16/13cm. An abnormality of clotting was suspected and a series of other analysis were performed (D-Dimer, C-protein, S-protein, PDF, FVIII, vWF:Ag, lupus anticoagulant, aggregation tests). This way we've established the diagnosis of AvWD associated with CML. We've immediately started the treatment of the leukemia (initially with hydroxyurea, than gradually the replacement with imatinib) and the patient also received RBC concentrates, FFP, CRYO, desmopressin, with slow relief of the symptoms.

AvWD occurs in a variety of conditions, but other clinical features may direct attention away from this potential cause of bleeding. More studies are needed to determine the incidence of AvWD and to define its contribution to bleeding in the many diseases and conditions with which it is associated.