

SPONTANEOUS HEMATOMA IN A PATIENT WITH POLYCYTHEMIA VERA: A CASE OF ACQUIRED VON WILLEBRAND DISEASE?

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Background. Polycythemia vera (PV) is a chronic myeloproliferative neoplasm associated with both thrombotic and hemorrhagic complications. Bleeding events may occasionally be related to acquired von Willebrand disease (AvWD). We report a case of spontaneous rectus sheath hematoma in a patient with JAK2-positive PV, raising suspicion of concomitant AvWD.

Case Report. A 60-year-old woman affected by JAK2-positive polycythemia vera, receiving treatment with Ruxolitinib and low-dose Aspirin, presented to the Emergency Department on May 1st, 2025, complaining of acute abdominal pain following a coughing episode. Abdominal ultrasound revealed a large rectus sheath hematoma measuring approximately 13 cm. Contrast-enhanced abdominal CT scan demonstrated active bleeding from a branch of the epigastric artery. The patient subsequently underwent right iliac axis angiography with successful embolization of the ipsilateral inferior epigastric artery. Hemostatic investigations performed at our center demonstrated normal factor VIII levels, ristocetin cofactor activity at the lower limit of normality, very reduced collagen-binding activity (4%). However, platelet function analyzer testing (PFA/ADP and PFA/EPI) was within normal range, as was the analysis of high-molecular-weight von Willebrand factor multimers. Notably, colla-

gen-binding activity normalized after five months of follow-up.

Conclusions. This case highlights the diagnostic challenge of spontaneous bleeding in patients with polycythemia vera and underlines the importance of investigating acquired von Willebrand disease in the setting of unusual hemorrhagic complications. Although laboratory findings were not fully consistent with AvWD, the altered von Willebrand factor profile raises questions regarding subclinical hemostatic abnormalities in patients with myeloproliferative neoplasms. In the literature, only one case of AvWD caused by an antibody directed against the glycoprotein Ib-binding domain of von Willebrand factor has been described [1]. Moreover, the hemorrhagic event itself may have been favored by additional contributing factors, including concomitant pharmacological treatment. Early recognition and multidisciplinary evaluation may significantly impact therapeutic management and bleeding risk assessment.

References

1. P J van Genderen et al, Acquired von Willebrand disease caused by an autoantibody selectively inhibiting the binding of von Willebrand factor to collagen, *Blood*. 1994 Nov 15;84(10):3378-84.