

ACQUIRED VON WILLEBRAD SYNDROME IN PATIENTS WITH MYELOPROLIFERATIVE SYNDROME: LITERATURE REVIEW AND SINGLE-CENTER EXPERIENCE.

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Background and aims: Acquired von Willebrand Syndrome (AvWS) is a rare bleeding disorder, exhibiting clinical and laboratory features similar to hereditary von Willebrand Disease (vWD). Unlike vWD, which stems from genetic defects in von Willebrand factor (vWF), AvWS occurs primarily in adults without a history of bleeding diatheses. AvWS is related with cardiovascular diseases, solid and hematologic malignancies and autoimmune diseases.

This study aims to review the latest data on the pathophysiology, clinical presentation, laboratory findings, and treatment of AvWS, focusing on its occurrence in myeloproliferative syndromes. We will also discuss cases from the Hemostasis and Thrombosis Center of AOU Renato Dulbecco in Catanzaro.

Methods: A literature review was conducted using major biomedical databases, focusing on AvWS in myeloproliferative syndromes. In our center, 100 patients with myeloproliferative syndromes underwent diagnostic tests, including bone marrow biopsy and von Willebrand disease panels. Treatment followed standard protocols, with aspirin prescribed for thrombotic prevention.

Results: The association between AvWS and MPNs involves mechanisms like vWF sequestration by platelets, excessive clearance, proteolysis by ADAMTS13, and autoantibody formation. The diagnosis requires comprehensive clinical history, laboratory tests including vWF multimer analysis. The treatment of AvWS primarily focuses on reducing platelet count, often using cytoreductive agents like hydroxyurea or ruxolitinib, along with hemostatic treatments (DDAVP, vWF/FVIII concentrates) in selected cases. The use of antithrombotic therapies in MPN patients with AvWD requires caution due to the potentially high bleeding risk. Four patients were diagnosed with AvWS: one with polycythemia vera and three with essential thrombocythemia.

Conclusions: This study highlights the complex relationship between MPNs and AvWD, influenced by excessive platelet production, vWF structural changes, and increased proteolysis by ADAMTS 13. The monocentric experience confirmed that regular monitoring of vWF levels is essential for identifying risk patients and preventing severe bleeding.

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