Von Willebrand's Disease: Keeping Your Coronaries Clean or A Bloody Mess?

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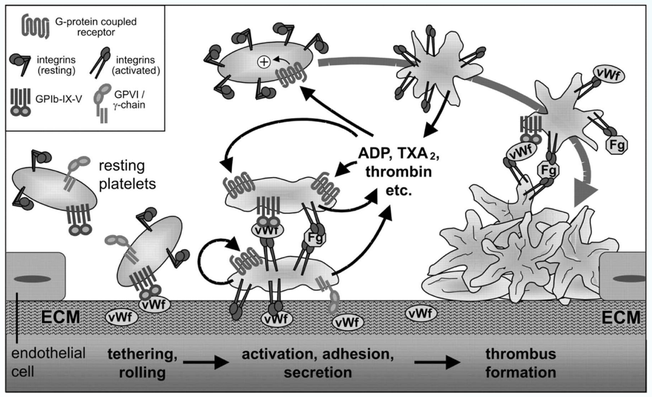
Learning Objectives

1. Does von Willebrand's Disease reduce risk for acute coronary syndrome?
2. How do we treat a von Willebrand's Disease patient with acute coronary syndrome?

Introduction

Von Willebrand's Disease (vWD) is the most common heritable bleeding disorder in the world. The disease is caused by a multitude of mutations that may lead to diminished levels or function of von Willebrand factor (vWF), Without sufficient vWF, platelets are unable to sufficiently bind endothelial components when vessel damage occurs. Due to its integral role in platelet function and platelet plug formation, it is thought that vWD may provide some level of protection from acute coronary syndrome. This case demonstrates a young male with a history of vWD presenting with potential acute coronary syndrome.



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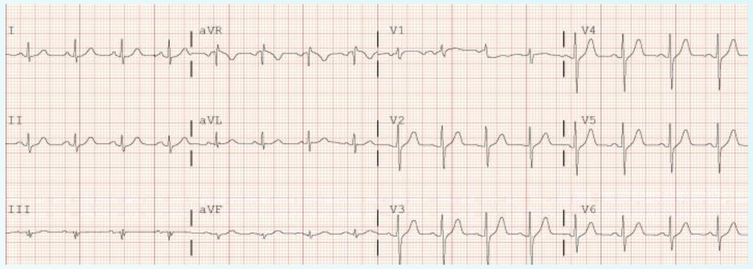
Patient Presentation

HPI: A 40-year-old gentleman with a history of vWD presented to a local Emergency Room following multiple episodes of anginal chest pain and dyspnea over the course of twelve hours. In the ED, initial EKG showed sinus tachycardia without ST segment or T wave changes. Lab work was significant for a troponin elevation to 6.5, consistent with an NSTEMI. Due to potentially significant bleeding risk, the case was discussed with Hematology who recommended administering aspirin but withholding heparin. The patient was transferred to our facility for cardiac evaluation.

PHYSICAL EXAM:
Cardiac: RRR; no murmurs, rubs, gallops; 2+ pulses
Pulmonary: CTA; no wheezes, rhonchi, or rales

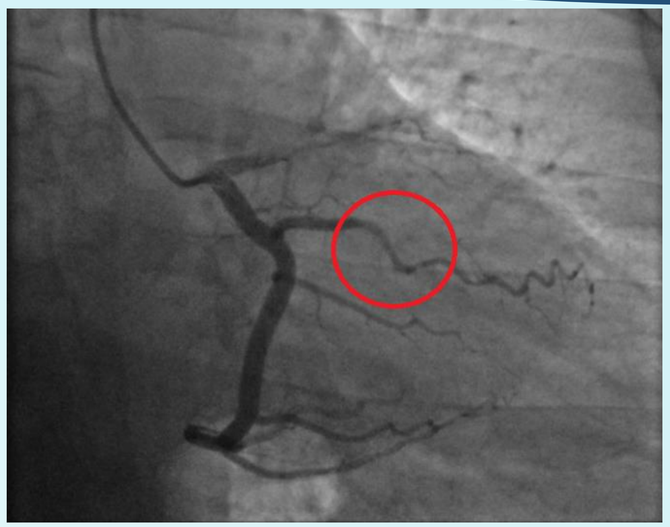
LABORATORY DATA:
CBC – normal
BMR – AST 41
Troponin - 5.091
Coagulation Panel
- INR: 1.0



Clinical Course

At arrival to our facility, the patient was no longer having active chest pain. Repeat EKG showed unchanged findings from the outside hospital. Repeat labs showed a down-trending troponin (5.091) and mildly elevated AST (41), but otherwise was unremarkable. He was taken for percutaneous cardiac intervention where a hazy mid-narrowing in the 1st obtuse marginal vessel was found; suggestive of spontaneous vessel dissection. Due to the tortuous and fragile nature of dissected vessels, no stents were placed at this time due to the patient's hemodynamic stability.

Following the procedure, hematology recommended only use of single anti-platelet therapy due to bleeding risk thus the patient was started of clopidogrel. His post-procedural hospitalization was complicated by the formation of a hematoma around the radial catheterization site. He however remained hemodynamically stable and did not require blood transfusion. Following stabilization of the hematoma, he was discharged home 3 days later with clopidogrel, statin, beta blocker therapy and intranasal desmopressin.



Take Home Points

Does von Willebrand's Disease reduce risk for acute coronary syndrome?
- In two small cross-sectional studies, there was a slight reduction in atherosclerotic plaque formation in vWD and Hemophilia A compared to controls. (1)
- A meta-analysis revealed that patient's with higher levels of vWF were at great risk for developing MI (Odds ratio: 1.5) (1)

How do we treat a von Willebrand's Disease patient with acute coronary syndrome?
- If pursuing PCI and dual-antiplatelet therapy (DAPT), may administer vWF/Factor VIII or desmopressin pre- and post-operatively to reduce bleed risk. (3)
- Recommend use of bare metal stents over drug-eluting stents due to shortened DAPT duration (30 days). (3)
- Utilizing radial artery approach during PCI for easier tamponade of bleeds. (1)
- Discharging patient with intranasal desmopressin for post-discharge bleeding.