PathMD™: Board Review Letter

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Case #1 The image for this question shown on the website represents initial platelet adhesion to the vessel wall and aggregation. What structure is represented by the red box in the image?

A. GPIb receptor

Answer: A. GPIb receptor on the platelet is responsible for initial adhesion of the platelet to the vessel wall via vWF as a bridge, which binds to exposed collagen.

Case #2 The image for this question shown on the website represents initial platelet adhesion to the vessel wall and aggregation. What structure is represented by the red box in the image?

B. GPIIb/IIIa receptor

Answer: B. GPIIb/IIIa receptor mediates aggregation of platelets with fibrinogen serving as the bridge between the GPIIb/IIIa receptors.

Questions

1. Moderate hemophilia A is characterized by which of the following approximated factor VIII levels:

   C. 1-5%

Answer: C. Severe: <1%, Moderate: 1-5%, and Mild: >5%

2. Adhesion of platelets is mediated through all of the following EXCEPT:

   A. GPIIb/IIIa

Answer: A. GPIIb/IIIa receptor on platelets is responsible for aggregation. The GPIIb/IIIa receptor binds with fibrinogen to create a platelet plug. GPIb, collagen, and von Willebrand's factor all work in concert together to cause adhesion of the platelet to the vessel wall. When the endothelium of the vessel is disrupted, collagen is exposed which binds to von Willebrand's factor. Von Willebrand's factor serves as a bridge between the collagen and the GPIb receptor on the platelet.

3. The PFA-100 analyzer has replaced the bleeding time at many institutions. This test utilizes two tubes for analysis. One tube contains collagen and ADP, while the other tube has collagen epinephrine. In a patient who has a prolonged bleeding time, which of the following would have a normal clotting time in the collagen and ADP tube but be prolonged and the collagen epinephrine tube?

   D. Aspirin effect

Answer: D. Aspirin results in a thromboxane deficiency. In the PFA-100 analyzer, a thromboxane deficiency is overcome by the abundance of ADP. Therefore this test can be helpful in separating storage pool defects, which include aspirin, from other possible at realities. That being said, bleeding times and the PFA-100 are still not very sensitive or specific tests.

4. Which of the following would be least helpful in treating a patient with hemophilia A:

    C. FFP

Answer: C. FFP contains very little factor VIII needed to increase the patient's levels. Cryoprecipitate is rich in vWF and Factor VIII (remember they travel together). DDAVP releases factor VIII and vWF from endothelial cells.

5. A patient underwent cardiac bypass surgery and was having an uneventful post-operative course. On POD #7, the patient developed an ischemic leg. The intern asks your help in diagnosis, and also informs you that his platelet count has recently dropped. The most likely etiology is:

    B. Heparin induced thrombocytopenia

Answer: B. Heparin induced thrombocytopenia (HIT) usually develops approximately 5-10 after starting treatment with heparin (time period can be shorter for patients with previous exposure). In cases of HIT with significant risk of thrombosis the platelet count usually drops >50%. HIT results from antibodies against heparin bound to PF4 on platelets. The platelet count usually recovers in 2-5 days after discontinuing heparin. (Goodnight, pages 425-431)

6. Which of the following tests would be most helpful in diagnosing the patient described in question 5?

    C. Heparin antibody

Answer: C.

7. A patient who is known to have von Willebrand's disease (vWD) has a normal vWF antigen and decreased levels of Factor VIII. What is the most likely vWD subtype in this patient?

    E. vWD, type 2N

Answer: E. vWD type 2N (Normandy) is characterized by normal amounts of vWF, which interact with platelets in a normal fashion. They have a mutation in the region that binds with Factor VIII, which results in decreased affinity. Factor VIII is degraded in the plasma approximately five time faster than when protected by vWF. Platelet type, pseudo-von Willebrand's disease looks identical to vWD type 2B, except the mutation is a gain of function in the GPIb receptor where as vWD type 2B is a gain of function in the vWF. (Kitchens, pages 91-100)

8. A patient is suspected of having Bernard-Soulier disease. If the patient has this disease, they would show response to all of the following during platelet testing EXCEPT:

    D. Ristocetin

Answer: D. Bernard-Soulier disease is defined by deficiency of the GpIb receptor on the platelet. GpIb is responsible for platelet adhesion, which is stimulated only by ristocetin in platelet testing.

References:

Disorders of Hemostasis and Thrombosis. Goodnight SH, Hathaway WE. Second Edition. 2001.

Consultative Hemostasis and Thrombosis. Kitchens, CS, et al. 2002.

Notes for question set:[1]

[1] PathMD strives for the highest quality and accuracy. However, the *PathMD: Board Review Letter* is for review purposes and not meant for clinical decision making. It should not be used in place of review of primary reference texts and the current medical literature. If inaccuracies are identified, please notify us so that a correction may be published. (info@PathMD.com)

