Glanzmann Thrombasthenia

Glanzmann’s Thrombasthenia is a genetic or acquired defect in the GPIIb/IIIa platelet receptor that normally binds fibrinogen and facilitates platelet aggregation. This qualitative platelet defect manifests with symptoms such as mucosal bleeding and easy bruising, while lab tests show an increased bleeding time but a normal platelet count. These patients are advised against taking NSAIDs, which further prevent platelet aggregation and may exacerbate bleeding.

PLAY PICMONIC

Pathophysiology

Autosomal Recessive or Acquired as an Autoimmune Disorder

Recessive-chocolate and Auto-in-moon

Glanzmann’s thrombasthenia is most often an autosomal recessive disease, meaning that both parents must each pass on one mutated allele for the offspring to be affected. Rarely, patients can acquire the disease if they form autoantibodies to their GPIIb/IIIa receptors.

Deficiency of Glycoprotein IIb/IIIa (Gp IIb/IIIa) Receptor

Broken Glider-protein with (2) Tutu Bee and (3) Tree Apples

The GPIIb/IIIa receptors anchor platelets to each other by binding fibrinogen on neighboring platelets. Since platelets are unable to clump together, platelet plugs cannot form, and primary hemostasis does not begin. In contrast, Bernard-Soulier syndrome involves the deficiency of the GPIb receptor, which affects the binding of von Willebrand factor to platelets.

Defective Platelet-to-Platelet Aggregation

Broken Plates can’t Aggregate

Without GPIIb/IIIa receptors on their surface to bind fibrinogen, adjacent platelets are unable to bind to one another and form a platelet plug as part of primary hemostasis. Drugs such as abciximab used for anticoagulation during angioplasty take advantage of this mechanism by blocking GpIIb/IIIa receptors.

Symptoms

Mucous Membrane Bleeding (Gingival Bleeding)

Mucous Dripping and Bleeding

Patients with platelet disorders may exhibit mucosal bleeding, which may manifest as epistaxis, hemoptysis, menorrhagia, or gingival and GI bleeds.

Easy Bruising

Easy-button Bruising

Patients with platelet disorders may exhibit easy bruising, petechiae, purpura, and ecchymoses.

Increased Bleeding Time

Up-arrow Blood Clock

Because platelets have difficulty aggregating and forming a platelet plug, patients have a prolonged bleeding time. This test is rarely performed today but is still tested on exams.

Normal Platelet Count

Normal Plates

Affected patients have a normal platelet count, in contrast to Bernard-Soulier syndrome, in which patients have sparse, enlarged platelets.

Considerations

Avoid NSAIDs

Avoid-sign N-sad

NSAIDs non-selectively inhibit cyclooxygenase (COX), an enzyme that produces thromboxane A2, a protein that promotes platelet plug formation. Patients with Glanzmann’s thrombasthenia should not take NSAIDs, as their platelets already are unable to aggregate properly.