DISTURBANCES IN THE HEMOSTATIC SYSTEM IN HEMORRHAGIC VASCULITIS

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Abstract

In this article you can read about disturbances in the hemostatic system in hemorrhagic vasculitis.

Key words: primary hemostasis, hemorrhagic vasculitis, system, disturbances.

Hemostasis is the physiological process by which a bleeding stops. Its final result is a thrombus (blood clot), which consists of blood cells and fibrin strands. Hemostasis involves the following mechanisms:

Primary hemostasis

Vascular hemostasis: transient vasoconstriction and vWF activation following endothelial injury

Platelet hemostasis: adhesion, activation, and aggregation of platelets, which results in the formation of a platelet plug (white thrombus)

Secondary hemostasis: activation of the coagulation cascade, which results in the formation of a fibrin clot (red thrombus)

Primary hemostasis

Definition: : processes involved in the formation of a platelet plug (white thrombus) following endothelial injury

Vascular hemostasis

Endothelial injury results in:

Neural stimulation reflexes and endothelin release → transient vasoconstriction, leading to:

Reduced blood flow

Platelet accumulation at the vessel walls

Exposure of subendothelial collagen → circulating von Willebrand factor binds to the exposed collagen

Von Willebrand factor (vWF): plasma protein that is synthesized by and stored in endothelial cells (in Weibel-Palade bodies) and platelets (in α-granules)

Mediates platelet adhesion and aggregation

Binds factor VIII (and thereby prevents its degradation)

Platelet hemostasis

Platelet adhesion: platelets bind to vWF via platelet GpIb receptor at the endothelial injury site

Ristocetin normally activates vWF to bind to glycoprotein Ib

Platelet activation: After binding to vWF, platelets change their shape and release mediators that lead to activation of more platelets (positive feedback). ; These mediators include:

Adenosine diphosphate (ADP): promotes adhesion of platelets to endothelium

Thromboxane A2 (TXA2): activates additional platelets and promotes vasoconstriction

Calcium: required for secondary hemostasis

Platelet-activating factor (PAF): a phospholipid mediator that is produced by platelets and inflammatory cells (e.g., neutrophils, monocytes, macrophages), involved in platelet aggregation and activation and local inflammatory response

Platelet aggregation

Mediated by GpIIb/IIIa-receptor and fibrinogen → formation of a white thrombus composed of platelets and fibrinogen

A white thrombus is transient, unstable, and easily dislodged. It stabilizes through the process of secondary hemostasis.

Preview thumbnail for image: Primary hemostasisPreview thumbnail for image: Shape of inactive and active plateletChalk Talk: Primary hemostasis 1Chalk Talk: Primary hemostasis.

Conclusion

Bleeding disorders are a group of heterogeneous conditions characterized by defects in hemostasis that lead to an increased susceptibility to bleeding (also known as hemorrhagic diathesis). They are classified into disorders of primary hemostasis (when caused by a platelet abnormality), disorders of secondary hemostasis (when caused by defects in the extrinsic and/or intrinsic pathway of the coagulation cascade), and hyperfibrinolysis (when there is increased clot degradation). Although clinical features may overlap, mucocutaneous bleeding (e.g., epistaxis, petechiae, gastrointestinal bleeding) is associated with disorders of primary hemostasis, and bleeding into potential spaces (e.g., hemarthrosis, muscular bleeding) is characteristic of disorders of secondary hemostasis. The diagnostic workup of a bleeding disorder begins with a detailed clinical assessment, the CBC, and a coagulation panel. This typically allows the disorder to be classified as one of primary or secondary hemostasis. Specialized studies are then required to determine the specific etiology so that treatment can be initiated. Treatment may include transfusion of blood products, replacement of specific coagulation factors, or administration of adjuvant medications (e.g., tranexamic acid or desmopressin).

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