Platelet and White Blood Cell Disorders.

Amr Qudeimat

4/11/18

Platelet Disorders

Platelet function

Wound healing requirements

Platelet count methods.

Automated methods shortcomings

Platelet count regulation.

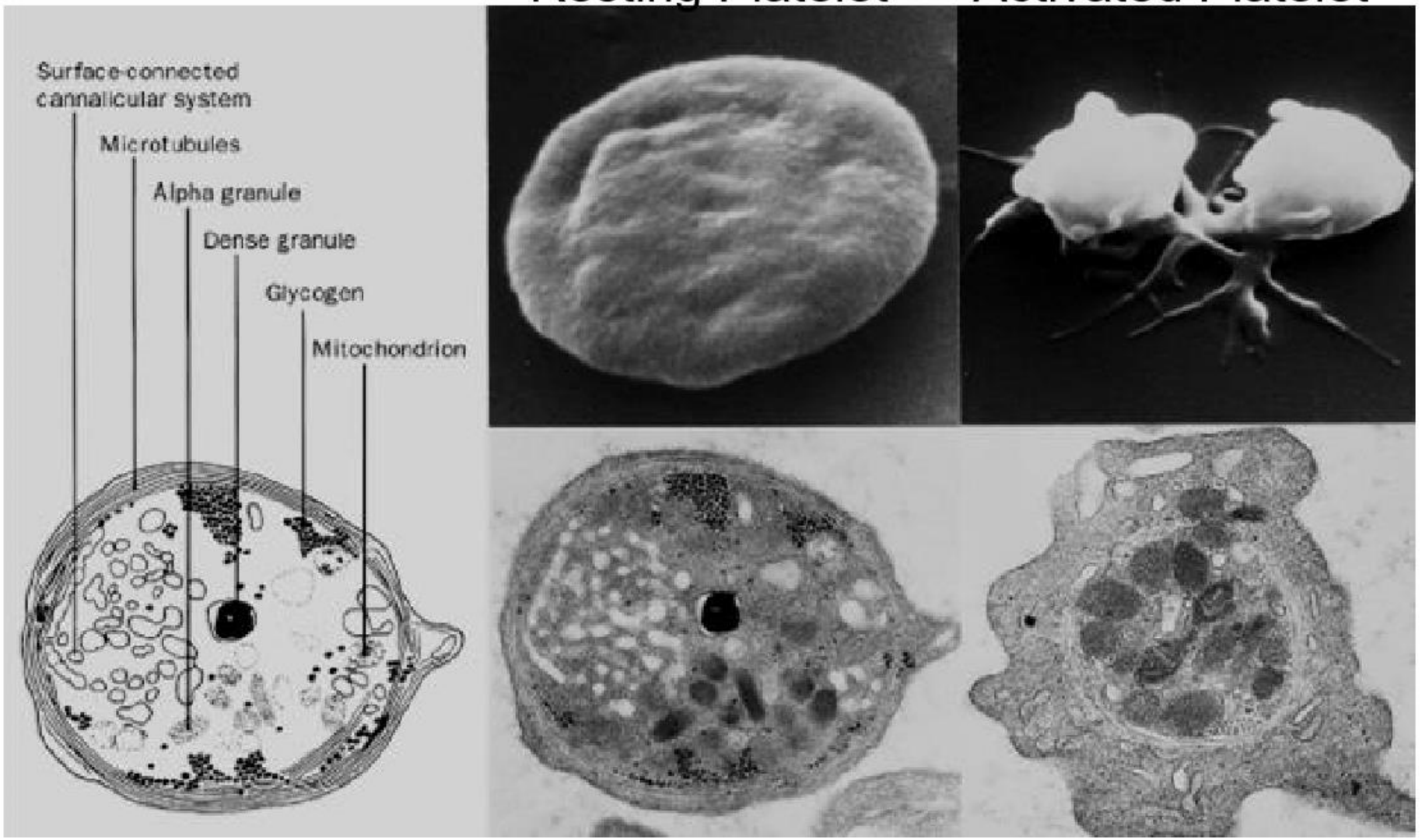
Platelet Structure

- Small disks.
- No nuclei as they don’t need them.
- Mitochondria, lysosomes, peroxisomes, alpha granules (contain VWF, fibrinogen, PF4, PDGF) and dense granules (contain ADP, ATP, serotonin, calcium).

Platelet utilization

- Platelet adhesion: attaching to subendothelial vWF/ collagen via platelet GP1b-IX-V and GPVI.
- Activation: shape changes to expose GPIIb/IIIa receptors.
- Aggregation: cross linking of platelet activated GPIIb/IIIa by fibrinogen/ vWF
- Propagation of coagulation: coagulation factor complexes/ enzymes attach to the to the activated platelet surface.

Resting Platelet Activated Platelet



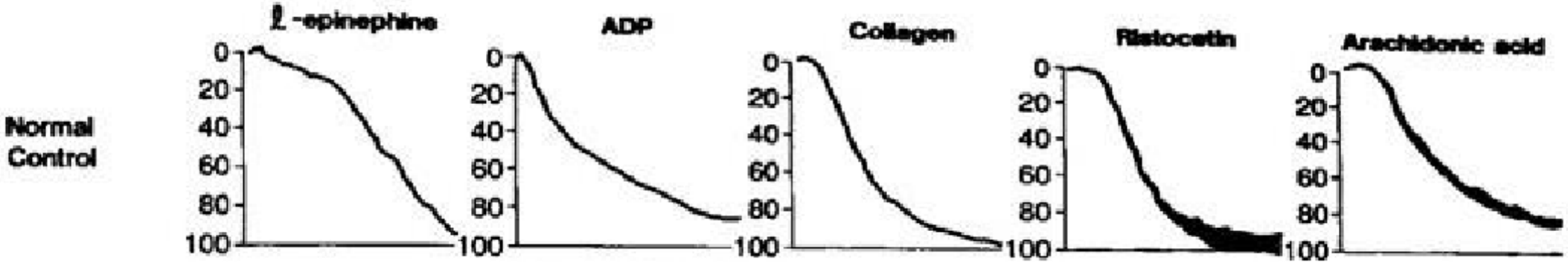
Platelet function testing

- Bleeding time: not used anymore
- PFA-100
- Aggregation studies.

PFA-100 Results

Diagnosis	Col/Epi result	Col/ADP result
Normal Patient	Normal (<187 sec)	Not performed
Aspirin effect	Prolonged	Normal (<114 sec)
Possible platelet defect or Von Willebrand Disease	Prolonged	Prolonged

Platelet Aggregation: Normal Tracing



Platelet related bleeding

Tends to happen immediately after the trauma.

Petechiae are more indicative or platelet problems than are bigger bleeds

Family history can be very helpful as many of the defects may be genetic.

Quantitative platelet disorders

- Pseudothrombocytopenia.
- Primary bone marrow problem.
- Loss of external drive/ inhibition.
- Pooling.
- Increased utilization.
- Increased destruction/ shortened life span.

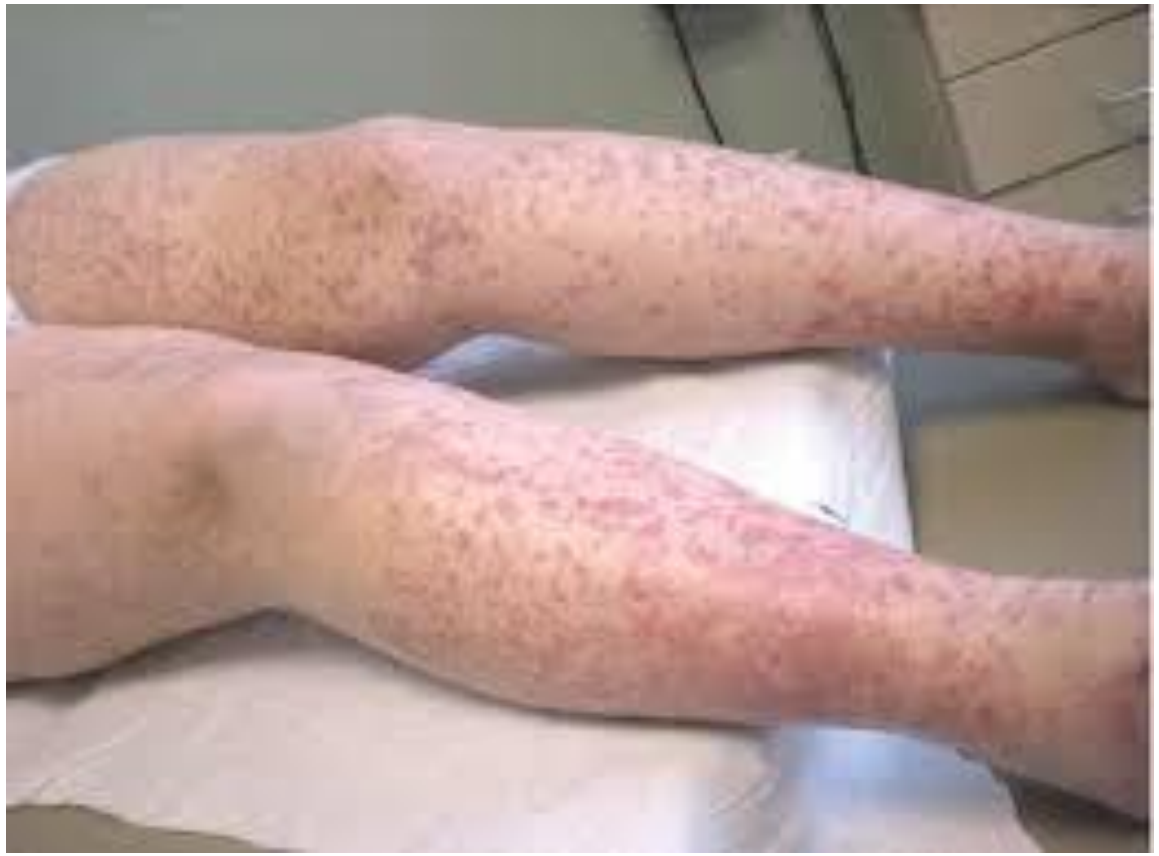
Shortened platelet life span

Non-immune:

- DIC.
- TTP.
- HUS.
- Kasabach-Merritt syndrome.

Shortened platelet life span

- Immune:
- ITP Mechanism, presentation, natural history, treatment and outcome.
- Heparin- induced thrombocytopenia/ drug induced
- Immune thrombocytopenia in neonates: Autoimmune vs alloimmune.



Congenital causes.

- Wiskott-Aldrich syndrome.
- X-linked thrombocytopenia.
- Thrombocytopenia Absent Radii (TAR) syndrome.
- Congenital Amegakaryocytic thrombocytopenia.

Qualitative platelet defects

Glanzmann’s Thrombasthenia: platelet membrane fibrinogen receptor (GPIIb/IIIa).

Bernard-Soulier syndrome: absent or abnormal platelet surface receptor for vWF (GP Ib/IX).

Gray platelet syndrome.

Hermansky-Pudlak syndrome: pulmonary fibrosis, OC albinism and absent dense granules on EM

Disorders of white blood cells

- Neutropenia secondary to infection.
- Drug induced neutropenia.
- Autoimmune neutropenia .
- Chronic benign neutropenia of childhood.
- Alloimmune neutropenia.
- Chronic granulomatous disease

Disorders of white blood cells

- Kostmann’s syndrome.
- Cyclic neutropenia.
- Schwachman-Diamond syndrome.
- Chronic idiopathic neutropenia.
- Leukocyte adhesion deficiency.
- Chediak-Higashi syndrome