Hematology

- Histology
- Biochemistry
- Pathology
- Pharmacology
- Physiology
- Microbiology

- Dr. name : **Saleem**
- lecture number :
- Done BY : **Amani Atallah**

- Handout
- Sheet **7**
- Slide

By: Anwer Abu Shanab

Bleeding Disorders & ABO Blood Groups

A case of bleeding is usually associated with one of these disorders:

1- Vascular disorder (in the blood vessels themselves).

2- Deficiency of platelets (decreased count). #the most common

3- Platelets function disorder.

4- Coagulation factors deficiency (abnormality in blood clotting factors). #the 2nd most common

5- Excessive functioning of the fibrolytic system.

---------- In details-----

1- Vascular disorders;

- The problem (or defect) is either in the blood vessel itself, or in the perivascular connective tissue around it.
- Characterized by: Easily Bruised vessels [Purpura], and Spontaneous Bleeding.
- Usually not sever if the problem is only in the blood vessel.
- Could be inherited/Acquired;
  - Inherited: Appears at childhood (mild), becomes moderate-sever in adulthood and old age.
  - Acquired: types →

    a. Senile Purpura (purpura of old age).

    b. Purpura associated with Infections (chronic infections).

    c. Scurvy Disease (Vit. C deficiency, 'vitamin C is involved in most processes').

    d. Steroid Purpura.

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2- Platelet Deficiency (decreased count);

- It's called "Thrombocytopenia"
- Characterized by: Spontaneous Skin Purpura, Mucosal Hemorrhage, and Prolonged Bleeding especially after trauma.
- Platelets are important for maintaining the normal integrity of the blood vessel. They are deficient in cases of (main causes of thrombocytopenia":
  - Failure of platelets production due to [Drugs, Chemicals, Viral infection].
  - Part of general Bone Marrow Failure [Aplastic anemia, Leukemia, Megaloblastic anemia]
  - Increased destruction of platelets [Heparin, Disseminated Intravascular Coagulation (DIC)]
  - Abnormal Distribution of Platelets [Splenomegaly: enlargement of spleen thus it captures a lot of platelets]
  - Dilutional Loss [Massive Blood Transfusions]

➔ "Thrombocytopenic Purpura": it's the purpura that occurs because of low platelet count characterized by ruptured vessels, easily bruising, and multiple cutaneous skin rashes.

3- Disordered (Abnormal) Platelet Function;

- AKA "Thrombocytopathia"
- Characterized by: Skin Purpura, Mucosal Hemorrhage, Prolonged BT.
- Inherited/Acquired;
  - Inherited: A problem in the release of platelets' substances, or a deficiency in glycoprotein 1 and 4 or Factor8 (vWF) dificiency or deficiency in the production of thromboxane A2, or a failure of platelet aggregation (factor 8-related Ag).
  - Acquired: Aspirin Therapy (which delays the production of Tx-A2) is the most common cause.

➔ "Thrombustynic Purpura": normal platelet count, but abnormal function of circulating platelets.

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4- Coagulation Disorders;

- 2nd most common cause of bleeding.
- Note the described factors+disorders in the table in the slides.
- Haemophilia A, Haemophilia B, von Willebrand Diseases are uncommon, but the others are rare!
- In most, inheritance is somatic; however, some are X-linked.
- These patients should avoid dental extractions and circumcisions unless under proper medical care, because they have a high tendency for bleeding.
- There's usually a correlation between the patient' symptoms and the severity of the disease.
- Factor12 deficiency is not associated with abnormal bleeding.
- Factor11 is activated directly by platelets, and if deficient produces mild symptoms.
- Factor13 is important in stabilizing the fibrin threads; along with thrombin and Ca. Factor13 deficiency produces severe bleeding.
- **Haemophilia A:**
  - The most common among the uncommon. Incidence 1:10000.
  - Factor8-c is deficient; Factor8-related Ag is the only coagulation dependent.
  - Sex-linked (still 30-35% of patients don't have a family history) → Appears in Males. Females are only carrier (a female cannot be diseased; 2 abnormal genes (Homozygosity): Fatal).
- **Von Willebrand disease:**
  - Factor8-related Ag is deficient, and this results in rapid destruction of Factor8-c → defected adhesion+coagulation.
    - Remember that:
    # Factor8 (vWF): for Adhesion
    # Factor8-related Ag: for Aggregation
    # Factor8-c: for Clotting
- **Haemophilia B:**
  - Similar symptoms to A, but less common.
  - Factor9 is deficient.
  - Sex-linked

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5- Excessive functioning of the fibrolytic system;

- Fibrinolysis is a physiological response, but excessive fibrinolysis is pathological.
- Ca is usually eliminated by EDTA or oxalate to decrease its effect on thrombosis.
- Anticoagulant substances are used as medications to treat thrombosis or delay the production of its associated substances. Usually they're classified into 3 classes;
    1. Coumadin -like anticoagulants (Warfarin):
        - Plant origin.
        - Acts slowly (usually after 1-2days)
        - Given orally only.
        - Delay/decrease the production of Vit.K dependent factors (in vivo only).
        - Decrease the production of "Thrombokinase" (which activates factor10).
        - Thrombokinase formation will be delayed if the blood is put on smooth surfaces.
    2. Heparin:
        - Animal origin.
        - Acts rapidly.
        - Given IM or IV
        - Very important anticoagulant. Inhibit all intrinsic factors (10-12) thus decreasing thrombus formation.
        - Functions in vivo and vitro.
    3. Hirudin:
        - A chemical taken from leech "العلق".
        - Functions on thrombin & fibrinogen.
    4. Stirring: (mechanically to remove the thrombus.)

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ABO Blood Groups

- Different Systems are used for blood grouping (classic, Rh, minor), but the ABO system is the main one (Classic one). Different antigens (more than 100) are expressed during early fetal life on the surface of RBCs and remain unchanged. Out of these antigens, we have 15 well-defined blood group systems; however 2 of them are the major systems of grouping).
- 4 Main groups: A, B, AB, O [according to the Ags present on the RBC surface]

1. In group A: there are A antigens (agglutinins; so called due to their interactions with the corresponding Abs in plasma if found) + Antibodies against B (anti-B or beta).
   [If you transfer blood of group B, to a patient of group A, agglutination occurs due to the interactions between B-Ags (in the transfused blood) and B-Abs (in the patient of A group).] : "Incompatible"
2. In group B: B antigens + Anti-A (alpha) antibodies in the plasma.
3. In group AB: Both A Ags + B Ags on the surface of RBCs, NO Abs in the plasma.
4. In O group: NO Ags, Both anti-A Abs + anti-B Abs are present.

- These Antigens are genetically determined (appear in the early fetal life and remain unchanged), but the Antibodies are the ones acquired and produced naturally (after food ingestion [meats], which contain the Ags of the produced Abs) during the first 2-8 months of life. [Abs are not found in neonates.]
  Sometimes, the Antibodies are not produced at all throughout the whole life, unless the person is exposed to blood transfusion from a person with incompatible blood group.
- These Antigens are found in other tissues; Salivary glands, Pancreas, Kidneys, Liver, Lungs, Testes, Semen, Amniotic fluid.
- A phenotype "A" of a person, doesn't tell whether the Genotype is "AA, homozygous" or "AO, heterozygous". (Goes for phenotype "B" too.)
- A phenotype "AB" of indicates the co-dominance of both A and B genes.

  # Other minor blood grouping systems include: MM, MN, NN, PP, etc; these may be associated with problems of producing the Ag with its Ab resulting in agglutination of blood.

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Rh Blood grouping

- One of the important systems of grouping.
- Depends on the presence (D+) or absence (D-) of the antigen "Rhesus" on the surface of RBC in addition to other antigens.
- In Europe, 85% of population: D+, 15%: D-.
- There are NO ANTIBODIES in the plasma of D+ people.
- There are neither antibodies nor antigens in the D- people.
- When the blood is transfused from a D+ person to a D- patient, the D- blood will start making antibodies against D antigens.
- It's believed that there are more than one antigen responsible for making the Rh grouping; Dd,Cc,Ee; but the most recognizable is D!

Racial distribution of blood groups (table)

- Jordanians + Arabs in general are similar to the people of China and India.
- The data is collected in 1943, but still similar to the data of new studies.

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- Amani Atallah