DISEASES INVOLVING BLOOD PLATELETS -I

DEPARTMENT OF ORAL PATHOLOGY

LEARNING OBJECTIVES

- At the end of the lecture student should be able to
- Describe definition, types,classification, etiology, clinical features,lab. Findings & treatment of purpura

- Describe clinical features & lab.Findings of thrombotic thrombocytopenic purpura

- Describe clinical features & lab.Findings of Wiscott aldrich syndrome

INTRODUCTION

- Platelets, also known as thrombocytes are the smallest of blood cells varying in diamteter from 2-4μm.

- They are spherical or oval discoid structures, look pinhead under oil immersion.

- Blood smear stained with Leishman stain stains platelet mauve-pink.

- Formation of platelets-

- Produced in the bone marrow.

- The pluripotent stem cell destined to form the platelets is converted into colony forming unit called Meg-CFU.

- Meg-CFU is converted into Megakaryoblast.

- Megakaryoblast converted to Promegakaryocyte.
- Promegakaryocyte converted to megakaryocyte .
- Megakaryocyte converted to platelets.
- Formation of platelets from stem cell takes about 10 days.
- Lifespan varies from 8-12 days.

- Functions –
- When activated,they perform following functions-
- Role in hemostasis.
- Role in clot formation.
- Role in clot retraction.
- Role in repair of injured blood vessel and defence mechanism.

PURPURA

Characterized by decreased number of circulating blood platelets.

Defination : Purplish discoloration of the skin and the mucous membrane due to the spontaneous extravasations of blood and itself is a symptom rather than disease. Normal count is 1,50,000-4,50,000/mm^3 of blood.Average being 2.5lac per cubic milimeter.

Classification :

- Nonthrombocytopenic
  - Primary
- Thrombocytopenic
  - Secondary

- Nonthrombocytopenic purpura-
- This type of purpura is not mediated through changes in the blood platelets but rather through damage to capillary endothelium that may result in their increased permeability.
- Thrombocytopenic purpura-
- A hematologic disorder characterized by markedly decreased number of circulating blood platelets.

Causes of nonthrombocytopenic purpura-

Drug induced damage to the capillary wall as seen in patients with prolonged treatment with corticosteroids,penicillin,sulphur drugs and aspirin.

Deficiency of vitamin C leading to less stable capillary basement membrane.

Damage to the capillary wall by antibodies.

- ***PRIMARY THROMBOCYTOPENIA (Werlhof's disease, purpura hemorrhagica and idiopathicpurpura)***
- *Autoimmune disorder* in which a person becomes immunized and develop the antibodies against his own blood platelets

- It appear due to absence of platelet stimulating factor.
- Reduced platelet production
- Increased platelet destruction
- Sequestration in a spleen

Etiological factors for secondary thrombocytopenia

1. Condition associated with the reduction of the platelet production

a. Hypoplasia or aplasia of megakaryocytes

- Ionizing radiation
- Drugs & chemicals
- Congenital hypoplastic anemia

• b. Infiltration of marrow by abnormal cells

- Leukemia
- Multiple myeloma

c. Megaloblastic anaemia

d. Metabolic disorder like hypothyroidism

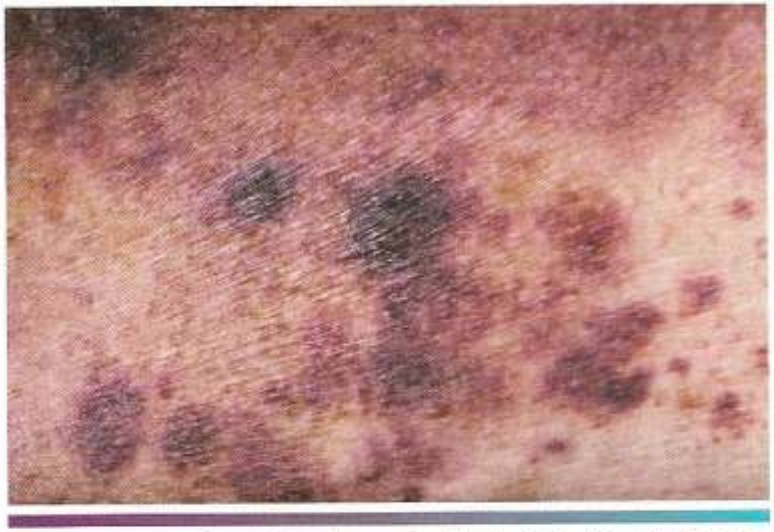
e. Infection

Conditions associated with a reduction of platelet lifespan due to

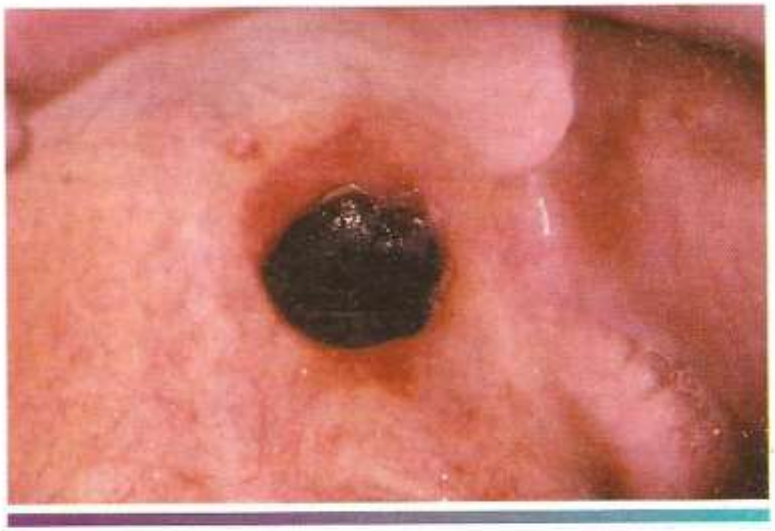
- Certain drugs intake like sedatives,antipyretic etc.
- Infection
- Hemolytic anaemia.

Clinical features

- Spontanous appearance of purpuric or hemorrhagic lesions.
- Timy red pinpoint petchiae to large purplish ecchymoses & even massive haematoma.
- Epistaxis or bleeding from nose – common manifestation of a disease.



Thrombocytopenia. The bruising (purpura) seen on this patient's forearm is a result of reduced platelet count secondary to myelodysplasia, a preleukemic bone marrow disorder.



Thrombocytopenia. This dark palatal lesion represents a hematoma caused by a lack of normal coagulation, characteristic of thrombocytopenia.

Oral manifestations

- Severe and often profuse gingival haemorrhage
- Petechiae on palate
- Oral surgical procedures particularly tooth extraction contraindicated

Lab findings

- Thrombocytes count < 60,000/mm^3
- Bleeding time > 1 hr.
- Coagulation time is normal
- Capillary fragility increase
- Torniquet test strongly positive

Gingival biopsy

- Gingival biopsy shows fibrin deposits in small vessel

Treatment

- No specific treatment
- Symptomatic relief from the bed rest and blood transfusion.
- Corticosteroids
- Prognosis → Fairly good

THROMBOTIC THROMBOCYTOPENIC PURPURA (Moschcowitz disease)

- Uncommon type of thrombocytopenic purpura.
- A life threatening immunologically mediated multisystem disorder described by Eli Moschowitz in 1924.

C/F-Young adults, more common in females.

- Characterized by thrombocytopenia, hemolytic anemia, transitory neurological dysfunction and renal failure.

H/P-Microthrombi in arterioles,venules,capillaries in all tissues and organs.

Presence of occlusive subintimal deposits of PAS positive material at arteriocapillary junctions is seen in biopsy of gingival tissue

- **L/F -**Blood examination : Thrombocytopenia and anemia.
- Peripheral smear- Fragmented RBCs (schistocytes)
- Prothrombin time and activated partial thromboplastin time are within normal limits.

Treatment-Corticosteroid,Platelate aggregation inhibitors, splenectomy and exchange transfusions.

- WISKOTT-ALDRICH SYNDROME- It is X-linked genetic defect in a protein termed Wiskott-Aldrich Syndrome Protein. (WASp).
- It resides in Xp11.22-23.
- Associated with deficiency of IgM.
- **C/F-** Thrombocytopenic Purpura,eczema beginning on the face.
- Susceptibility to infection.

- Petechiae purpuric rash or ecchymoses of the skin may be early signs of the disease.
- Patients manifest boils, otitis media, bloody diarrhea, and respiratory infection.
- Increased susceptibility to infection as patient is unable to form antibodies against polysaccharide-containing organisms.
- Malignant lymphoma.

O/M-Spontaneous bleeding from gingiva.

- **Palatal petechiae.**
- **L/F:** Prolonged bleeding time as the number of platelets is reduced between 18000-80000per cubic milimeter.
- Anisocytosis –the platelets appear smaller than the normal.

Treatment-

- Antibiotic and platelet transfusion
- Bone marrow transplantation .

SUMMARY

- Definition, types,classification, etiology, clinical features,lab. Findings & treatment of purpura
- Clinical features & lab.Findings of thrombotic thrombocytopenic purpura
- Clinical features & lab.Findings of wiscott aldrich syndrome

BIBLIOGRAPHY

- Shafer's-6th edition- Oral Pathology
- Sembulingam- Physiology

THANK YOU