HEMATOLOGY

HEMOLYTIC ANEMIA:

diverse grp of disorders characterized by an ↑in the Rate of RBC destruction

- Morphological features of RBC destruction: Microspherocytes, elliptocytes, RBC fragments etc.
- erythroid hyperplasia of Reticulocytosis – hallmark of hemolytic Anemia
- Expansion of Marrow spaces → Thalassemia & sickle cell Anemia.
- ↑LDH, ↑unconjugated bilirubin, ↓haptoglobin, ↑urobilinogen, urine hemosiderin.

CLASSIFICATION

extravascular & Intravascular hemolysis.

In most cases.

thermal, (osmotic cell)

→ EXTRAVASCULAR HEMOLYSIS: caused by factor extrinsic to RBC. It is due to defects that ↑ the destruction of Red cell by Phagocytes. This occurs in spleen.

Features:

→ Hyperbilirubinemia & jaundice: ↑ due degradation of Hb in macrophages.

→ splenomegaly: due work hyperplasia.

→ pigment stones: long standing HA can ↑ Risk of cholelithiasis bilirubin Rich gallstone.

→ INTRAVASCULAR HEMOLYSIS: RBCs are destroyed while circulating. It is characterized by injury to Red cells that are severe that they Rupture within the circulation.

Features

- Hemoglobinemia, Hemoglobinuria, hemosiderinuria: Hb is Released into the circulation (Hemoglobinemia)

- Free Hb combine with plasma protein called Haptoglobin. lead to ↑ in serum haptoglobin level. extend in urine hemoglobinuria.

→ hemosiderin is found in the urine hemosiderinuria:

Intrinsic

→ Hereditary → RBC membrane Abnormalities. ⟨ spherocytosis / elliptocytosis.

→ Enzyme defiency: G6PD defiency, pyruvate kinase, hexokinase defiency.

→ Disorder of Hb synthesis → Thalassemia of sickle cell Anemia

Extrinsic

→ Immune hemolytic Anemias.

→ Fragmentation syndrome.

- **Loss of iron in Intravascular hemolysis**

Lab finding of HA

PERIPHERAL BLOOD:

- Marked ↑ in Reticulocyte count:

↓

They appear as polychromatophilic Red cell

- Nucleated Red cells → mostly late normoblast.
- morphology - spherocyte, sickle cell, target cell, acanthocyte Macrocytic Poikilocytic etc.
- WBC → ↑ in immature neutrophil in the circulating blood. presence of metamyelocyte, myelocyte
- Thrombocytosis

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BONE MARROW

Usually not necessary.

→ cellularity is ↑ed., erythroid hyperplasia.

→ ↑ no. of erythroid precursor (normoblast) in the marrow.

→ M:E Ratio is ↓ with a Reversal Ranging from 1:1 to 1.6.

HEREDITARY SPHEROCYTOSIS:

— Rare HA Resulting from inherited defect in the Red cell membrane. Leads to formation of spherocytosis. Thus RBC are highly susceptible to sequestration & destruction in the spleen.

— caused by various mutation (common) → band3, protein 4.2 Ankyrin or spectrin

↓

They stabilize the lipid Bilayer of Red cell.

→ spherical RBC - As there is weak interaction b/w the membrane skeleton of intrinsic Red cell membrane proteins. This destabilize the lipid bilayer of Red cell.

↓

As the RBC Ages, they fragment into circulation.

As a Result small Amount of

NON