

ITP

IMMUNE THROMBOCYTOPENIC PURPURA:

most common form of thrombocytopenia

Acute chronic

Acute Thrombocytopenia Purpura:

→ self limited disease - sudden onset of petechiae, gum bleeding, epistaxis.

→ occurs in children (2 to 4 years)
(usually 1 to 3 weeks after viral infection: (measles, Rubella, EBV)

→ M = F

Chronic Thrombocytopenia

→ more common

→ F:M = 3:1 b/w 20 to 40 years.

PATHOGENESIS:

Autoimmune disorder → chronic ITP is Autoimmune due to (Type II hypersensitive) #:

→ AutoAntibodies Mediated Agonist platelet destruction.

Against platelet membrane glycoprotein IIb/IIIa.
or IIb/IIIa complex.

Activation of complement

syn

These Antibodies are IgG type.



These IgG Anti Act as opsonins & coat platelet
Removed by spleen (destruction) by C3b

(no splenomegaly) → Destruction of platelet by phagocytosis (Mac)

Clinical features

- Insidious onset
- Present with petechiae, oral ulcers, epistaxis, gum bleeding, menorrhagia.

HLI → splenectomy of Immune suppression.

LAB DIAGNOSIS:

- Platelet count → Markedly Reduced
- Hb ↓ sed

PS → Thrombocytopenia.

→ Abnormally large sized platelet (giant platelets)

RBC → microcytic hypochromic Anemia. due ↓ RBC

due to large Release of young platelets

Bone Marrow

- Hypercellular
- Megakaryopoiesis → ↑ in number

Immature predominate → large not lobulated & less granules hypoblastosis

- Storage iron ↓ due to chronic bleeding
- lymphoid hyperplasia.

BT → Prolonged

CT → normal

PT & APTT → normal

Tourniquet test → +ve

Test for platelet Auto Antibodies → +ve

Spleen → normal size

EMPH SYMPTOMS → Autoimmune Hemolytic Anemia + positive Coombs test + PTP

HLI → Retrospective

They include TTP & HUS.

TP is 0

4) pentad of fever, thrombocytopenia, HANA, neurological defect & Renal failure

c) HUS has all except neurological defect features

In both condition NADH occurs.

Following endometrial Triplex:

Glycoprotein Ia/IIa bind with collagen

1" can bind with VWF

VWF are formed as large multimers which are disintegrated by ADAMTS13 / VWF metalloproteinases

Absence of ADAMTS13 (vWF) from high speed thrombi within the vessels

Accumulate in the Plasma $\rightarrow$ spontaneous platelet
Activation & Aggregation

→ In Addition presence of superimposed endothelial dysfunction cause thrombus formation within vessel

ЦАНА -

PATHOMECHANISMS OF TYPICAL HUS:

Infectious gastroenteritis



Shigella like toxin



Endothelial dysfunction



Platelet Activation



Defect in complement factor → Activate Alternate → Platelet Activation
Factor H & I deficiency pathway

→ As RBC pass through this thrombi → Fragmented RBCs -

Peripheral smears

→ schistocytes, erythroid Regeneration