

Subject:

HLS-pathology

Topic:

Lecture 5

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الجامعة



4 FACTORS RESPONSIBLE ABOUT STOP BLEEDING

1-BLOOD VESSELS

vasculitic infection

connective tissue disease

blood vessels related bleeding

chronic steroid intake->
weaken the blood vessels-> bleeding

vitamin c deficiency->
important for structure of
collagen in blood vessels

systemic amyloidosis->
amyloid infiltrate any organ

2-PLATELETS

platelets related bleeding: 1- thrombocytopenia 2-dysfunctioning platelets

Immune Thrombocytopenia purpura:
IgG AB bind to
platelet membrane glycoprotein IIb/IIIa ->
engulfed by macrophages in spleen->
((chronic ITP-> which affect middle age)->
f>m
- Acute ITP : affect children after viral infection (self limited)

Heparin induced thrombocytopenia:
IgG AB bind to
factor 4 on platelets ->
thrombosis+thrombocytopenia

Thrombotic microangiopathies:1-TTP 2-HUS
similar symptoms for both diseases:
fever, hemolytic anemia, thrombocytopenia
neurologic defect(just in TTP),
renal failure(dominance for HUS)
- small circulation is filled with
platelets rich microthrombi
blood film: schistocyte
thrombocytopenia

HUS: common in children
E.coli in the gut-> shiga toxin
-> reaches kidney -> endothelial
damage -> thrombosis

TTP: deficiency in ADAMTS13(plasma protein)->
important for VWF->
no mature VWF->
precursor of VWF (which is large)->
bind large number of platelets->
aggregation

glanzmanthrombasthenia : 1-autosomal recessive 2- acquired (AI)
-platelets do not bind to fibrinogen->
because of deficiency in platelets glycoprotein IIb-IIIa (CD41/CD61 complex)
->no aggregation of platelets -> bleeding

Bernard soulier syndrome:
deficiency in platelets membrane Ib(CD42b) ->
no binding to VWF ->
no adherence of platelets -> bleeding

4-ENDOTHELIUM : wide endothelial damage->release of tissue factor(prothrombotic agent)
->causing disseminated intravascular coagulation-> rapid consumption of clotting factor
-> exceed replacement -> life threatening bleeding
peripheral blood shows schistocyte , anemia ,thrombocytopenia

3-CLOTTING FACTORS: clotting factor related bleeding: PT test PTT test Mixing study (to differentiate between true deficiency and inhibitor antibody

Hemophilia B:
christmas disease
X linked, deficiency in factor 9
less common than
Hemophilia A
clinically similar to
Hemophilia B

Hemophilia A:
Xlinked inheritance, reduced factor 8
-Mild deficiency :excessive bleeding
after trauma
-severe,life threatening-> if level
of factor 8 is less than 1%
No skin petechiae(bleeding in
deep tissue)

Von Willebrand Disease:
1- autosomal dominant
2-most inherited
bleeding disorder
3- VWF in plasma
carrying factor 8
4- normally when endothelial is damaged
VWF bind to platelets -> platelets blog
((so VWD results in :1- non functional platelets
2-deficiency in factor 8

Type 1 VWD:
decreased level of
serum VWF

Type IIA:
absent of high molecular
weight multimers of VWF
((VWF precursor is absent))

Type IIB:
high molecular weight
multimers of VWF have
very short half life
, hyperactivation
-> consuming platelets
-> mild chronic
thrombocytopenia