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Q1. Write a note on Hodgkin lymphoma?

Answer1 : Hodgkin lymphoma :

The stages of Hodgkin lymphoma are:

- **Stage I** The cancer has only affected one lymph node region or organ.
- **Stage : II** Two or more lymph nodes are affected. The cancer is either above or below the diaphragm.
- **Stage III** : The cancer may affect lymph nodes on both sides of the diaphragm.

  Stage IV: Cancer cells have moved beyond the lymph system into other tissues and organs, such as the liver, lungs and other extranodal sites .

  - The stage number in all case is followed by the letter A or B and indicating the absence A or presences B of one or more of the following .
  - Unexplained fever about the 38°C , night sweats , or loss of more than 10% of body weight within 6 month
  - Localized extranodal extension from a mass of nodes does not advance the stage

    but is indicated by the subscript E .

Hodgkin Disease and clinical presentation :

Signs and symptoms	% of patients
Lymphadenopathy	90
Mediastinal mass	60
B' symptom	30
Fever ,weight loss, night sweats	
Hepatosplenomegaly	25

- ● Most commonly involved lymph nodes are the ccervical and supraclavicular in 75%

- Bone marrow is involved in 5% of patients.

Prognosis :

- Dagree of splenic involvement: > 5 nodules poor prognosis
- Age : > 50 year unfavorable .

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Q2.WHAT IS HEMOSTASIS , ALSO EXPLAIN STEPS AND CLOTTING FACTORS?

Answer 2 : HEMOSTASIS:

- The term of hemostasis means prevention of blood loss .
- Hemostasis is the arrest of blood flow and control of hemorrhage from an injured blood vessels

OR

- It the process by which bleeding at any site as the arrested by formation of hemostatic plug .

There are steps of hemostasis :

1. **Vasoconstriction :**
   - The muscles of the damaged blood vessels contract to reduce the amount of blood flowing through it by reducing size of the blood vessels .
   - Hormonal component :

     Circulating epinephrine and activision of sympathetic nervous system .

     Interact with cell surface adrenergic receptors

2. **Platelets plug formation :**

   Collagen fibers protruding from the site of the injury catch platelets and from a plug . platelets contract and shrink the wound .

- Here platelets perform 2 function .

1) Release other tow chemical like (thromboxane and ADP adenosine diphosphate)

2) fill the gap (injured areas).

Thromboxane and ADP help in platelets aggregations

3. Coagulation of blood :

Through out of life blood in fluid state but when removed or shed or collected in containers become jelly like - clot

- **Fluidity _** necessary for circulation.
- **Coagulation-** protective from excessive bleeding.
- Major homeostatic function responsible for preventing and termination of bleeding
- The process by which a blood clot is formed.
- Coagulation system consist of
- 13 clotting fators that help in different coagulation pathways.

Clothing Focters :

1 fibrinogen

2 Prothrombin

3 Thromboplastin (tissue factor)

4 Calium

5 Labile facto

6 Presence not proved

7 Stable factor

8 Antihemophilic

9 Christmas

10 Staurt power factor

11 Plasma thromboplastein or antecedent

12 Hagemen

13 Fibrin stablizing factors

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Q3.Explain Hemophilia its types, symptoms, and lab diagnosis ?

Answer 3 : Hemophilia : .

- Hemophilia A and B are similar in both clinical and pathological features,

the difference beingin the deficient factor.

- Both are sex-linked recessive disorders resulting in inherited deficiency

of the clotting factor or synthesis of a defective clotting factor. Males are affected and females are carriers..

TYPES OF HEMOPHILIA :

The two most common types of hemophilia are

1 **Hemophilia A**

2 **Hemophilia B**

1) Hemophilia A :

- Hemophilia A is the most common hereditary x linked recessive disease with a reduction.

● In the amount or activity of factors 8 about 30 % of hemophilia have no family history and may be due to acquired mutation .

2) Hemophilia B :

- Both factor 8 and 9 together activate factor 10 in Coagulation cascade.
- Assays of factors 9 should be done diagnosis Christmas disease .
- These server factors 9 deficiency is clinical indistinguishable from hemophilia A .

The three main forms of hemophilia include the following:

- Hemophilia A: Caused by a lack of the blood clotting factor VIII; approximately 85% of hemophiliacs have type A disease.
- Hemophilia B: Caused by a deficiency of factor IX.
- Hemophilia C: Some doctors use this term to refer to a lack of clotting factor XI.

Symptoms of hemophilia :

Signs and symptoms of spontaneous bleeding include:

- Unexplained and excessive bleeding from cuts or injuries, or after surgery or dental work
- Many large or deep bruises
- Unusual bleeding after vaccinations
- Pain, swelling or tightness in your joints
- Blood in your urine or stool
- Nosebleeds without a known cause
- In infants, unexplained irritability
- Double vision
- Neck pain

Lab diagnosis :

Diagnosis includes screening tests and clotting factor tests. Screening tests are blood tests that show if the blood is clotting properly. Clotting factor tests, also called factor assays, are required to diagnose a bleeding disorder. This blood test shows the type of hemophilia and the severity.

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Q4 .Describe Von Wille Brand disease?

Answer 4 : Von Wille Brand disease :

VON Willebrand disease (VWD) is a blood disorder in which the blood does not clot properly. Blood contains many proteins that help the body stop bleeding. One of these proteins is called von Willebrand factor (VWF). People with VWD either have a low level of VWF in their blood or the VWF protein doesn't work the way it should.

Introduction : Von Willebrand disease (vWD) was first described in 1926 by a Finnish physician named Dr. Erik von Willebrand.

- Von Willebrand disease is estimated to affect 1 in 100 individuals.
- Most researchers agree that von Willebrand disease is the most common genetic bleeding disorder.
- Glycoprotein and composed of 2050 amino acid
- It is a autosomal dominant inheritance disease pattern
- Males and females are affected equally

Function of VWF:

Functions : mainly have two roles

- Von Willebrand factor's primary function is binding to particular factor VIII and Factor VIII is bound to vWF while inactive in circulation; factor VIII degrades rapidly when not bound to vWF. Factor VIII is released from vWF by the action of thrombin
- It is important in platelet adhesion to wound sites. vWF binds to collagen, e.g., when it is exposed in endothelial cells due to damage occurring to the blood vessel.

Clinical Features :

- Most cases are of mild bleeding
- Spontaneous bleeding from mucous membranes (e.g. epistaxis)
- Excessive bleeding from wounds
- Menses bleeding increase
- In severe cases, similar to hemophilia A.

Physiology of VWF:

♧ Location: The majority of vWF is circulating in the blood plasma.

- Synthesis: vWF is synthesized as a monomer that is subsequently made into multimers that are secreted alpha granules of platelets and endothelial cells of blood vessels
- Store: mainly in weibal palate bodies of endothelial cells
- Receptors: A1 and C1 domain for plts, A3 domain for collagen and D3 for FVIII
- Cytogenetic Location: 12p13.3, which is the short (p) arm of chromosome 12 at position 13.3

❖ Also called coagulation factor VIII or F8VWF..

Pathophysiology :

There are two forms of VWD:

1 Hereditary vWD

- Quantitative
- Type I
- Type III

➢ Qualitative

- Type II
- Platelets type

2 Acquired vWD

Antibodies against vwf

LAB DIAGNOSIS :

- ❖ CBC
- ❖ Hemoglobin N/
- ❖ Hematocrit N/
- ❖ Platelet count Normal and Low in type 2b
- ❖ Prothrombin time Normal
- ❖ Activated partial thromboplastin time increase
- ■ Fibrinogen Normal

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Q5.Explain Hemolytic uremic syndrome and its types ?

Answer5 : Hemolytic uremic sy syndrome :

Hemolytic uremic syndrome (HUS) is a condition that results from

the abnormal premature destruction of red blood cells.

- Hemolytic uremic syndrome (HUS) is a condition that can occur when the small blood vessels in your kidneys become damaged and inflamed. This damage can cause clots to form in the vessels. The clots clog the filtering system in the kidneys and lead to kidney failure, which could be life-threatening.

Introduction :

- Most cases of hemolytic uremic syndrome develop in children after two to 14 days of diarrhea — often bloody — due to infection with a certain strain of Escherichia coli (E. coli).

■Adults also may develop hemolytic uremic syndrome after an E. coli infection, but the cause also may be certain medications, other types of infections, pregnancy or it may be unknown..

There are main two types of HUS:

Types of HUS :

_Classify into 2 main categories ,depending on weather it is associated with shiga- like toxin or not

Typically HUS:

- Typically HUS following diarrheal infection causes often by E .coli OH157 : H7 infection release shiga toxin produced E coli .shigella penumococcal infection HIV typically other viral or bacteria infection .

- **Topically HUS :**
- Caused by exposure to certain medication (eg ciclosporin, tacrolimus) genetic mutations in the complement pathways (4) and systemic conditions . include lupus cancer and pregnancy .

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*** *The END* ***