PATHOLOGY OF BLOOD AND LYMPHATIC SYSTEM–6

Dr. Tariq Al-Adaily, MD د. طارق العديلي

Associate Professor

Department of Pathology

The University of Jordan

Email: TNALADILY@ju.edu.jo

School of Medicine

WHITE BLOOD CELL DISORDERS

- Disorders include deficiency (leukopenia) and proliferation
- Leukocytosis: increased number of WBC in peripheral blood (any cause). If benign, it is called reactive leukocytosis
- Leukemia: increased number of WBC in peripheral blood secondary to neoplastic disease
- Leukocytosis is more common than leukopenia
- Reactive leukocytosis is more common than leukemia

NEUTROPENIA/ AGRANULOCYTOSIS

- Patients become susceptible to infections (namely bacterial and fungal)
- If neutrophil count drops below 500 cells/uL → spontaneous infection
- Decreased production: aplastic anemia, myelophthisic anemia, myelodysplastic syndrome, advanced megaloblastic anemia, chemotherapy, drugs (anti-epileptic, anti-hyperthyroidism)
- Increased destruction: immune mediated, splenomegaly, overwhelming bacterial, fungal or rickettsial infections

REACTIVE LEUKOCYTOSIS

- Neutrophilia: infections, inflammation (necrosis)
- Lymphocytosis: vial infections, Bordetella pertussis infection, chronic infections (TB, brucellosis)
- Monocytosis: chronic infections, rheumatologic diseases, inflammatory bowel disease
- Eosinophilia: asthma, allergic diseases, drug sensitivity, parasitic infections, Hodgkin lymphoma
- Basophilia: rare, seen in myeloproliferative neoplasms

REACTIVE LYMPHADENITIS

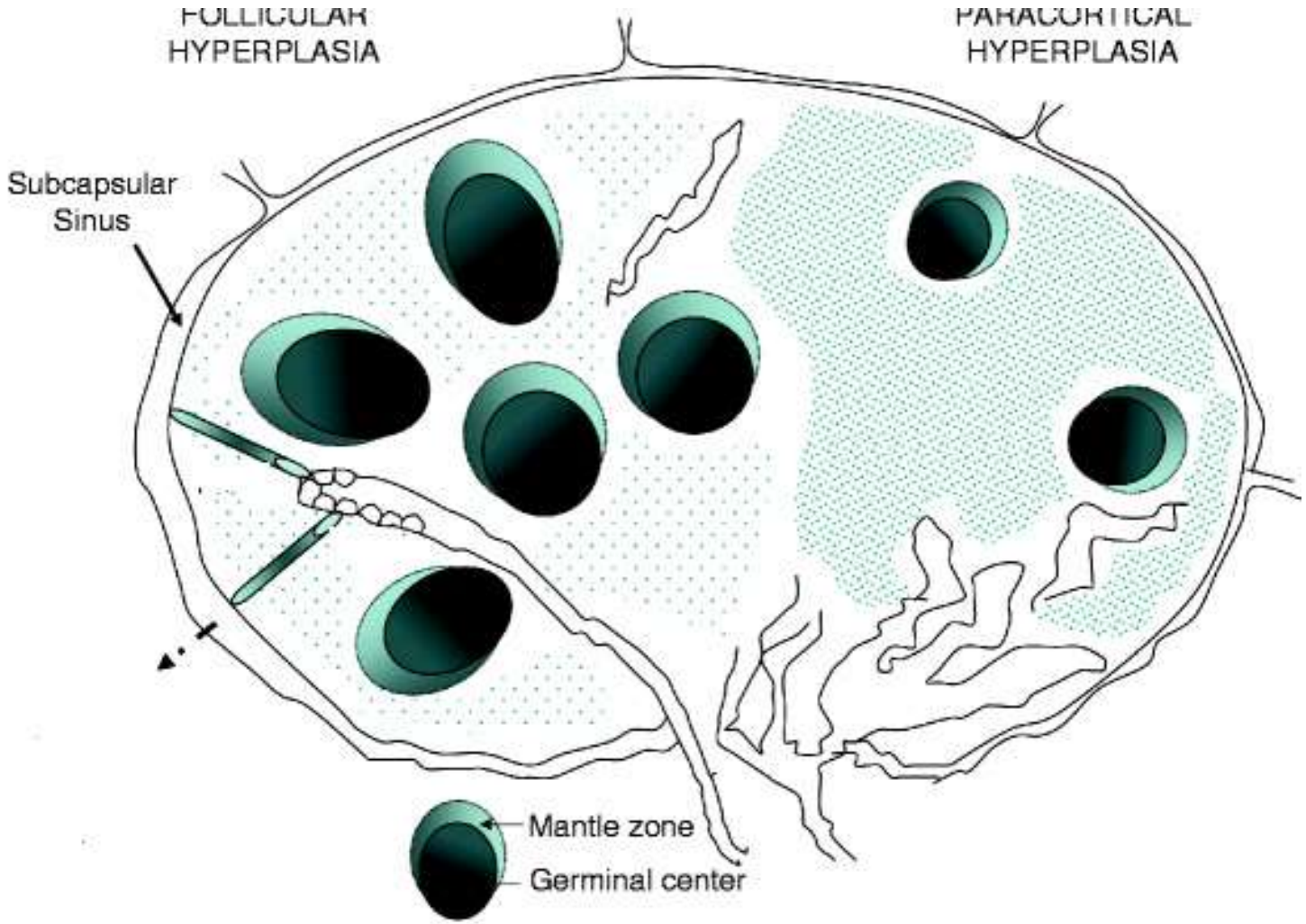
- Antigenic stimulation in lymph nodes
- Causes lymph node enlargement (lymphadenopathy)
- Can be localized or generalized

ACUTE NON-SPECIFIC LYMPHADENITIS

- Swollen, enlarged and painful lymph nodes
- Overlying skin is red and may develop a sinus tract
- The germinal centers in the lymph node are enlarged, infiltrated by neutrophils. With severe infection, liquefactive necrosis develop and may enlarge to form an abscess.

CHRONIC NON-SPECIFIC LYMPHADENITIS

- Chronic enlargement of lymph node, painless
- Follicular hyperplasia: chronic proliferation of B-lymphocytes, seen in rheumatologic diseases, toxoplasmosis and HIV infection
- Paracortical hyperplasia: proliferation of T-lymphocytes, seen in viral infections (example EBV), after vaccination and drug reaction
- Sinus histiocytosis: proliferation of macrophages in lymph node sinuses, seen in adjacent cancer


FOLLICULAR
HYPERPLASIA
PARACORTICAL
HYPERPLASIA
Subcapsular
Sinus
Mantle zone
Germinal center

CAT-SCRATCH DISEASE

- Bartonella henselae
- Transmitted from cats (bite, scratch, infected saliva)
- Most commonly in children
- Causes acute lymphadenitis in neck/axilla area
- Symptoms appear after two weeks of infection
- Bacteria causes liquefactive necrosis and necrotizing granulomas in lymph nodes
- Mostly self-limited in 2-4 months, rarely can disseminate into visceral organs

HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS

- HLH is uncommon disease
- Viral infection or other inflammatory agents activate macrophages (histiocytes) throughout body to engulf normal blood cells and their precursors in bone marrow
- Patients have defective genes related to the function of cytotoxic T-cells and natural killer cells, thus they are engaged with their target (virus-infected cells) for a long period and release excess interferon-γ that activates macrophages
- Activated macrophages release TNF and IL-6 that causes systemic symptoms of inflammation (systemic inflammatory response syndrome "SIRS")

HLH-TYPES

- 1) Infants and young children
- Homozygous defects in gene PRF1 that encodes perforin
- An essential enzyme in cytotoxic T-lymphocytes and natural killer cells

HLH-TYPES

- 2) Adolescents and adults
- X-linked lymphoproliferative disorder (males)
- Defective Signaling lymphocyte activation molecule (SLAM)-associated protein
- Inefficient killing of EBV-infected B-lymphocyte

HLH-TYPES

- 3) May be associated with systemic inflammatory disorders such as rheumatologic diseases
- Patients have heterozygous genetic defects in genes required for cytotoxic T-cells

HLH-TYPES

- 4) T-cell lymphomas
- Malignant T-cells produce aberrant cytokines leading to dysregulation of normal cytotoxic T-cells

SYMPTOMS

- Fever, splenomegaly and pancytopenia
- High ferritin
- High triglyceridemia
- High serum IL-2
- Low level of blood cytotoxic T-cells and natural killer cells
- BM: numerous macrophages engulfing RBCs, platelets and granulocytes