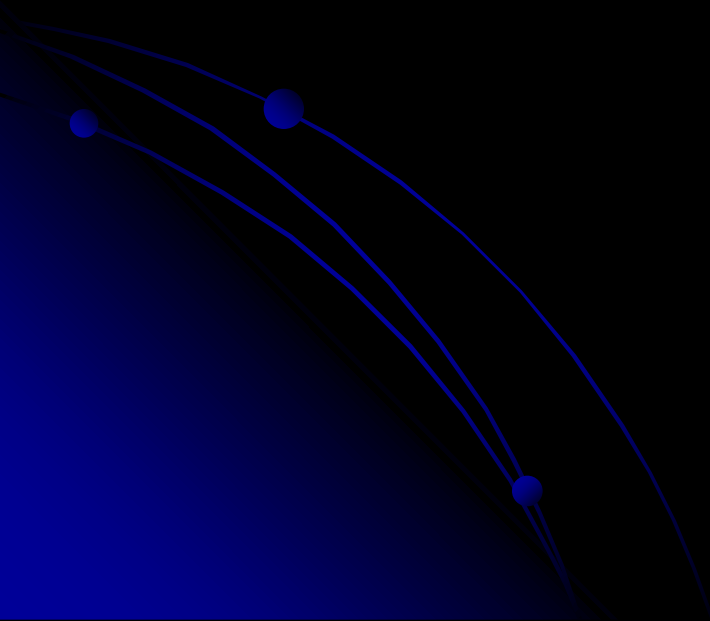
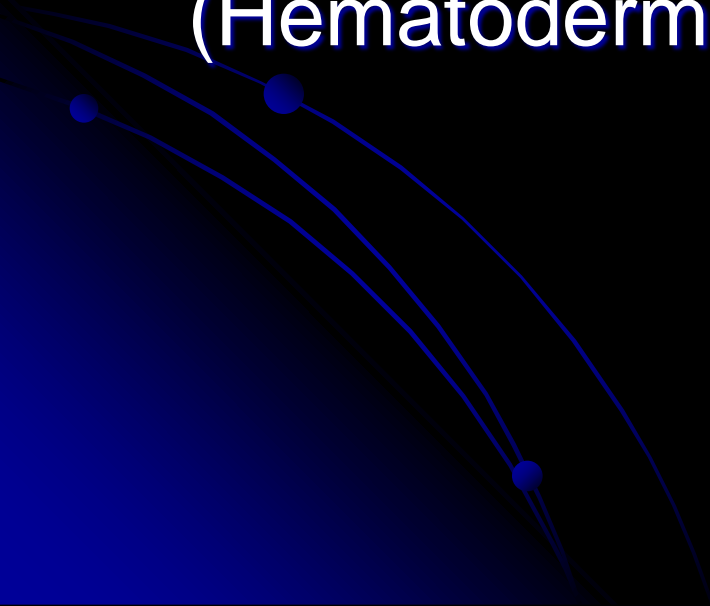


# Blastic Plasmacytoid Dendritic Cell Neoplasm

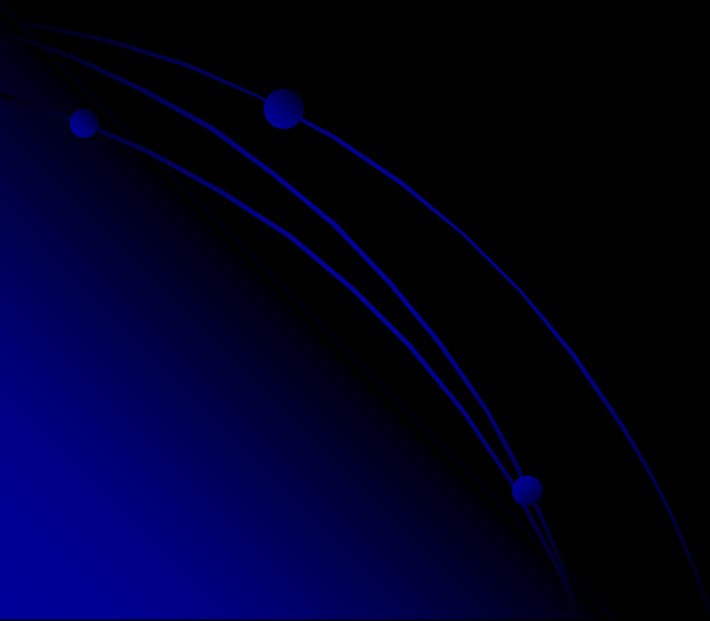


# Definition

- Lymphoblast-like cells
  - Plasmacytoid dendritic cell lineage
  - Identical to primary cutaneous CD4+, CD56+ hematolymphoid neoplasm (Hematodermic Lymphoma)
- 

# Definition

- Derivation from plasmacytoid dendritic cells (DC2 cells)

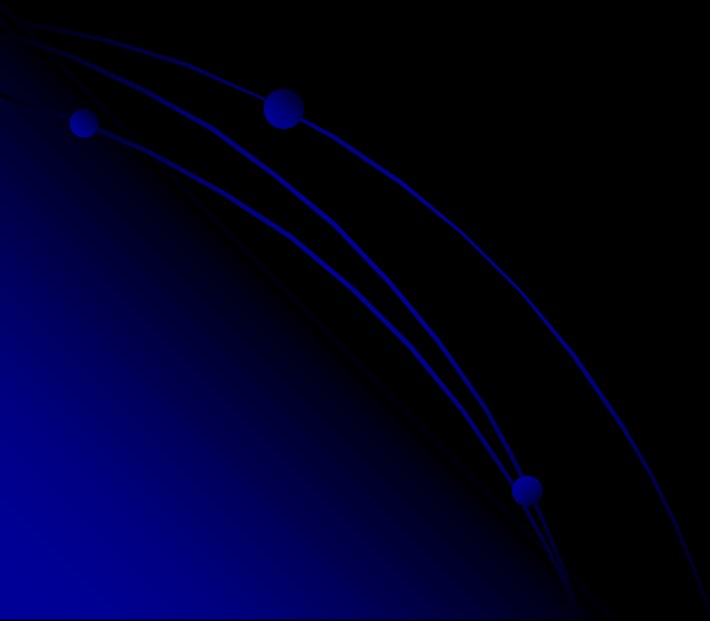


# Synonyms

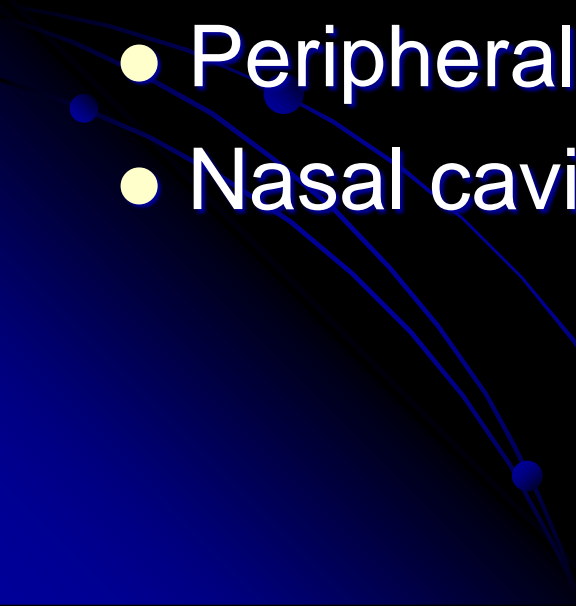
- None in the Lukes-Collins, Kiel, Working Formulation, or REAL classification
- Lymphoblastoid variant of NK-cell lymphoma
- Monomorphic NK-cell lymphoma
- Blastic NK cell lymphoma
- CD4+/CD56+ hematolymphoid neoplasm
- Hematodermic Lymphoma

# Epidemiology

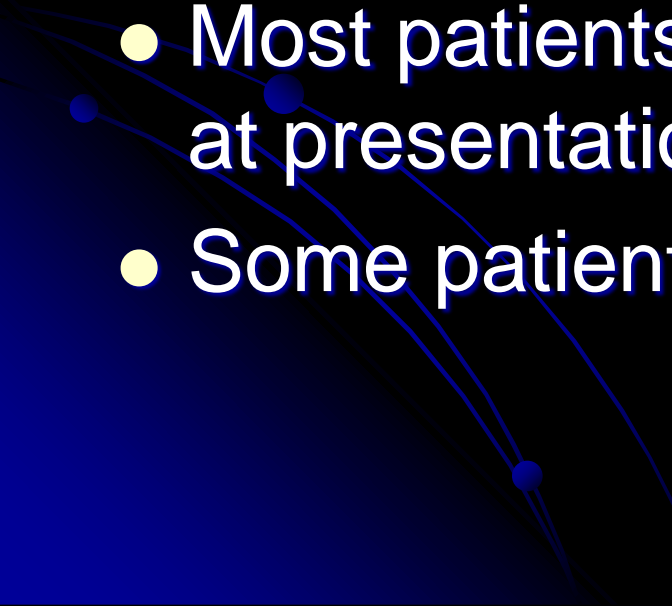
- Rare
- Any age, but mostly middle-aged or elderly



# Sites of Involvement

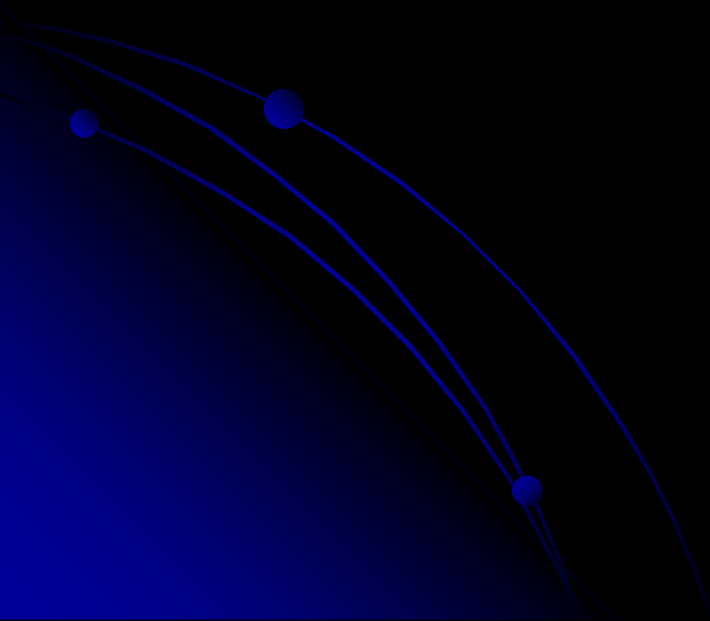
- Multiple sites
  - Skin predilection
  - Lymph nodes
  - Bone marrow
  - Peripheral blood
  - Nasal cavity (rare)
- 

# Clinical Features

- Skin lesions main presentation
  - Lymphadenopathy
  - Most patients have at least minimal bone marrow involvement
  - Most patients have disseminated disease at presentation
  - Some patients develop AML
- 


# Etiology

- Plasmacytoid dendritic cell (DC2)
- No EBV association





# Histology

- Diffuse infiltrate of medium-sized cells with fine nuclear chromatin (lymphoblastic or myeloblastic-like)
  - Single filing
  - No coagulative necrosis
  - No angiocentric lesions
  - Sometimes Homer-Wright rosettes
- 

# Immunophenotype

- CD56+, CD4+, CD43+, CD123+, TCL1+
- CD4 and CD43 generally positive
- TdT+ and CD34+ in some cases
- Surface CD3 negative
- CD2, CD7, CD3 $\epsilon$ , cytotoxic molecules generally negative
- CD68 generally negative

# Blastic Plasmacytoid Dendritic Cell Neoplasm: Diagnosis

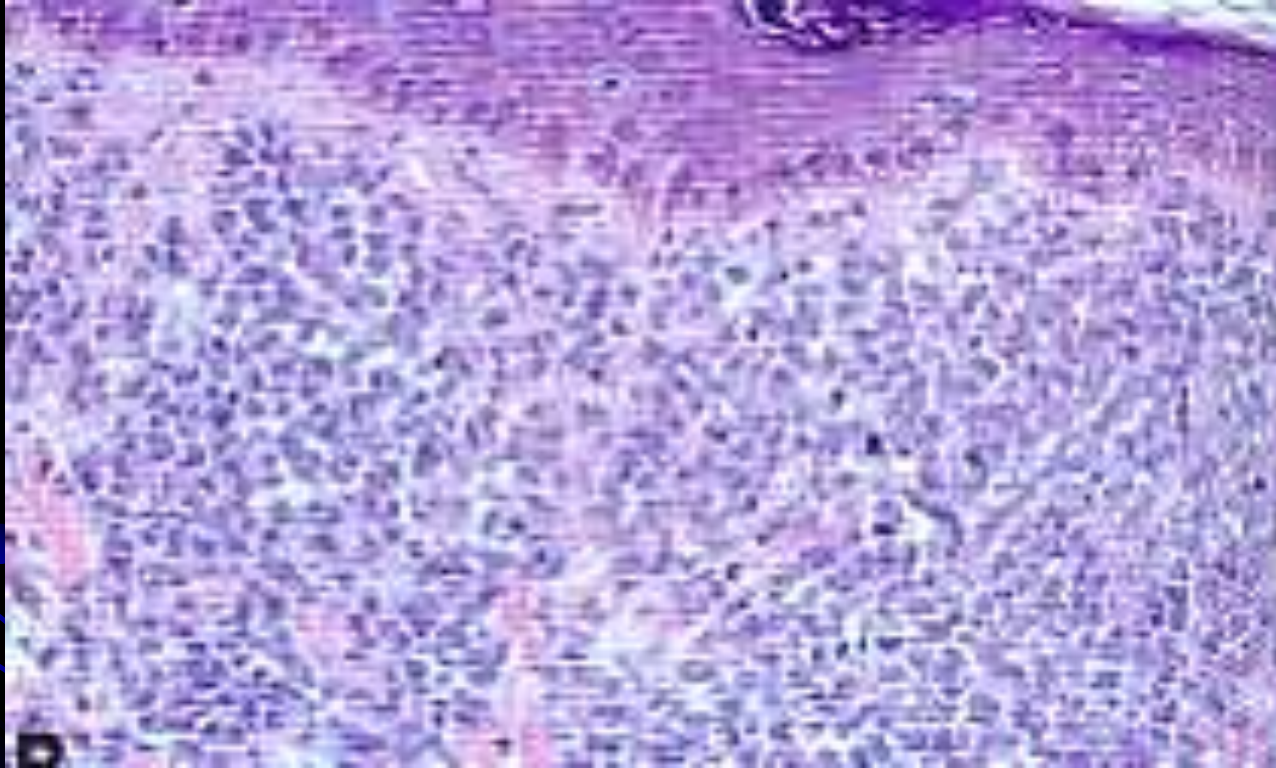
- CD56+ also in some AMLs and precursor T-cell lymphoblastic leukemia/lymphoma
- Diagnosis made only if myeloid and T cell lineages can be ruled out
- CD3 -, CD33 -, MPO -
- Absence of T-cell receptor gene rearrangements

# Blastic Plasmacytoid Dendritic Cell Neoplasm



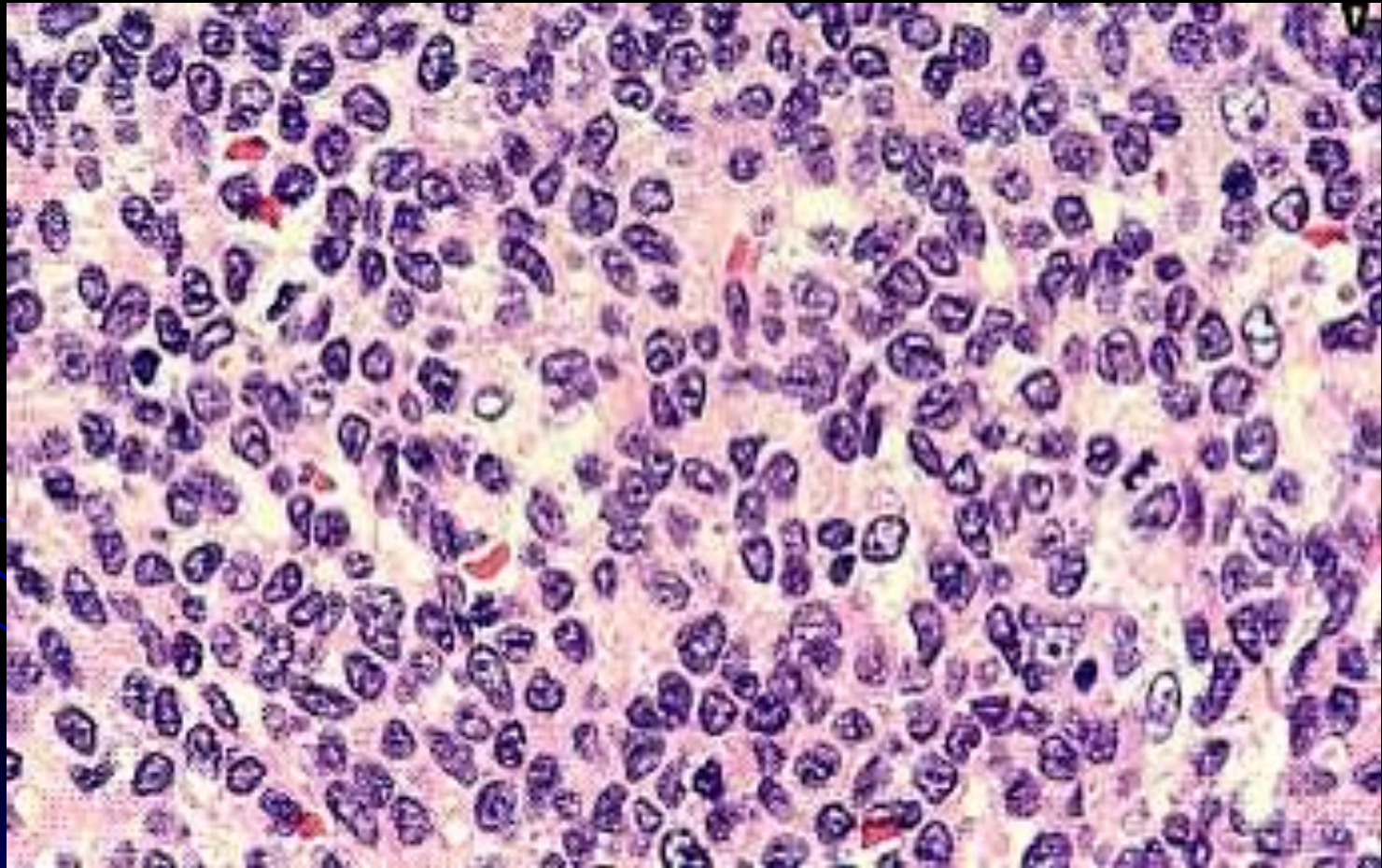
Skin tumor

# Blastic Plasmacytoid Dendritic Cell Neoplasm

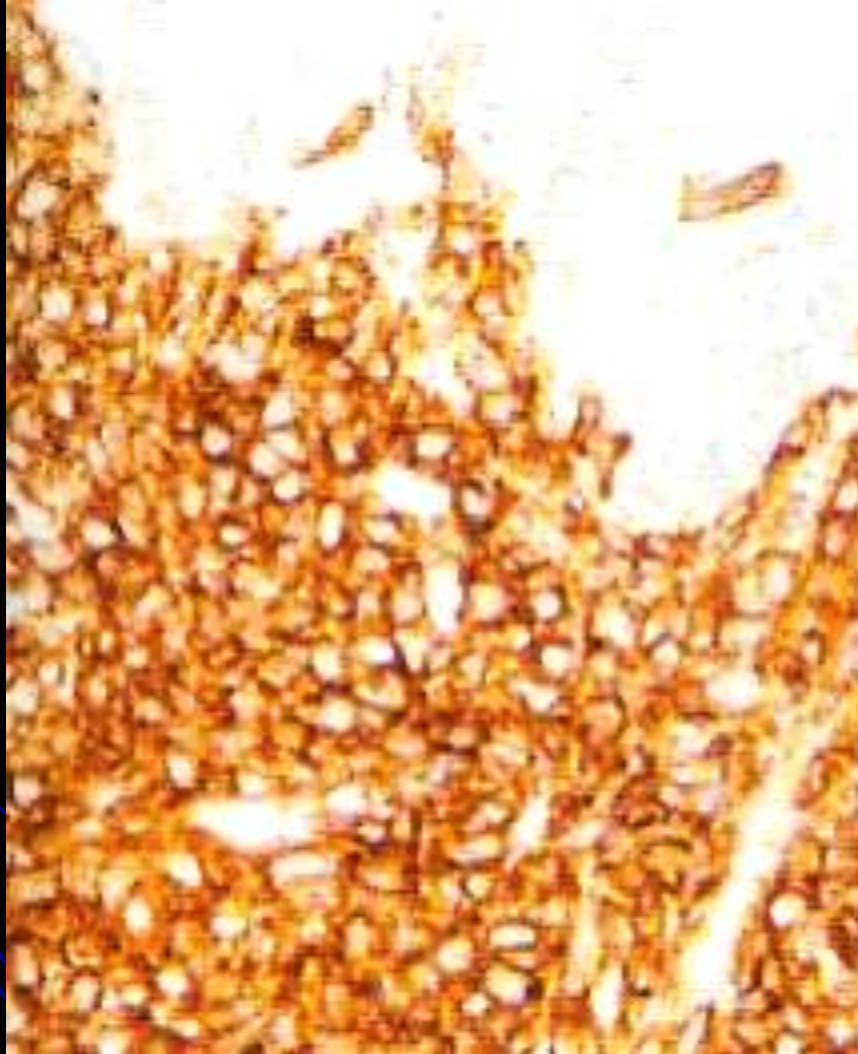


Diffuse dermal infiltration

# Blastic Plasmacytoid Dendritic Cell Neoplasm

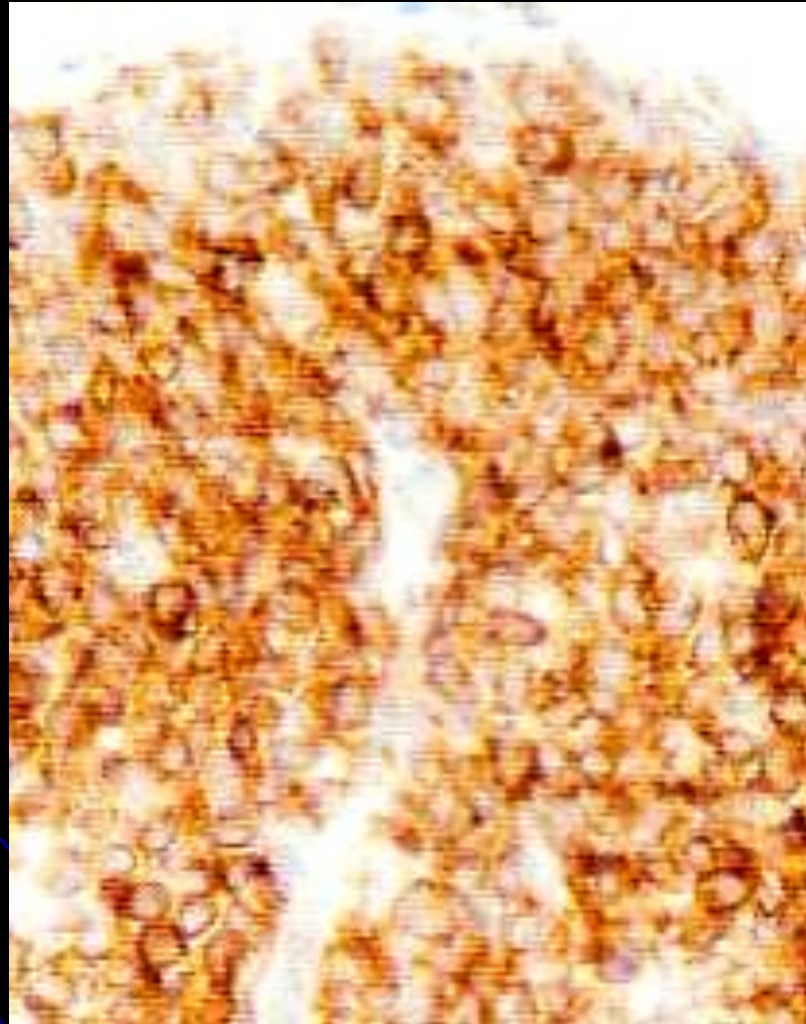


# Blastic Plasmacytoid Dendritic Cell Neoplasm



**CD43**

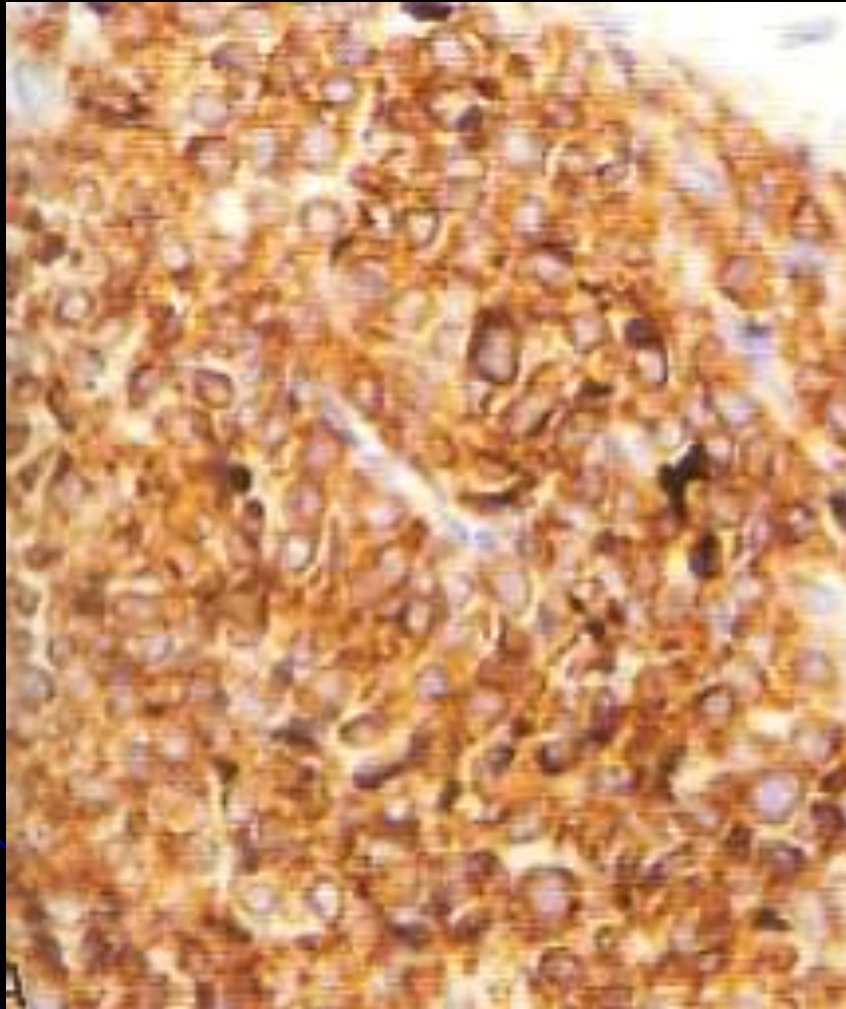
# Blastic Plasmacytoid Dendritic Cell Neoplasm



**CD56**

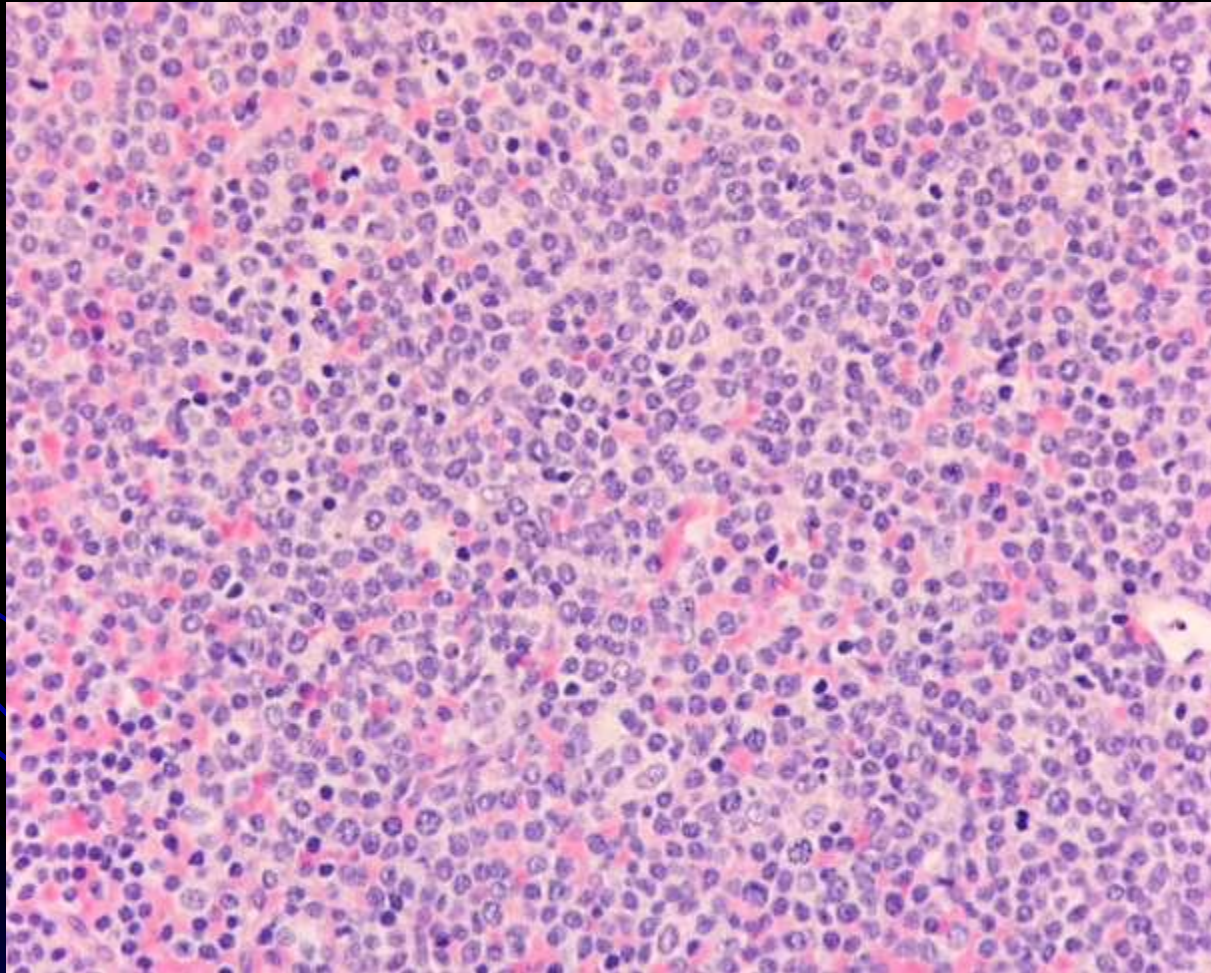


# Blastic Plasmacytoid Dendritic Cell Neoplasm

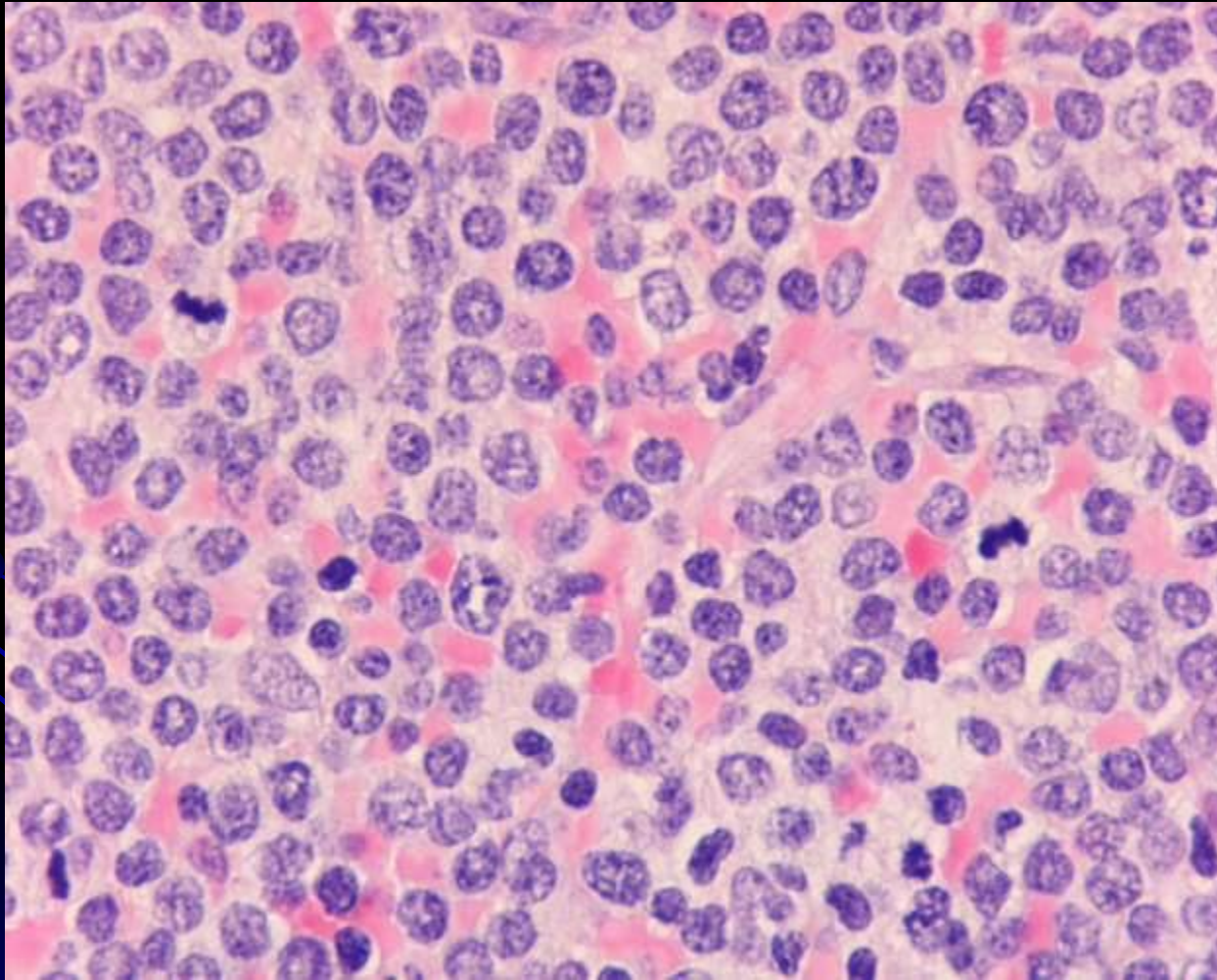


**CD4**

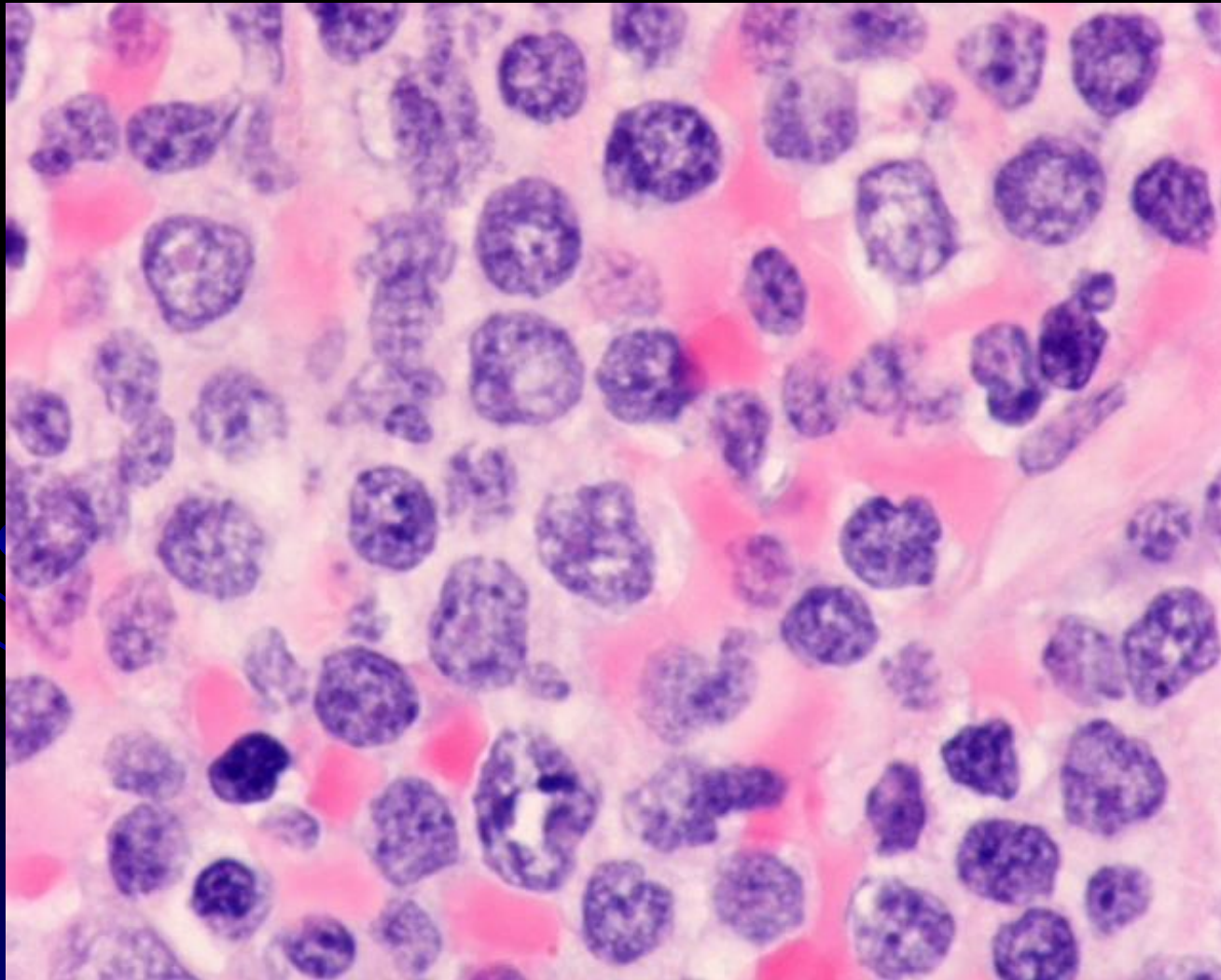
# Blastic Plasmacytoid Dendritic Cell Neoplasm



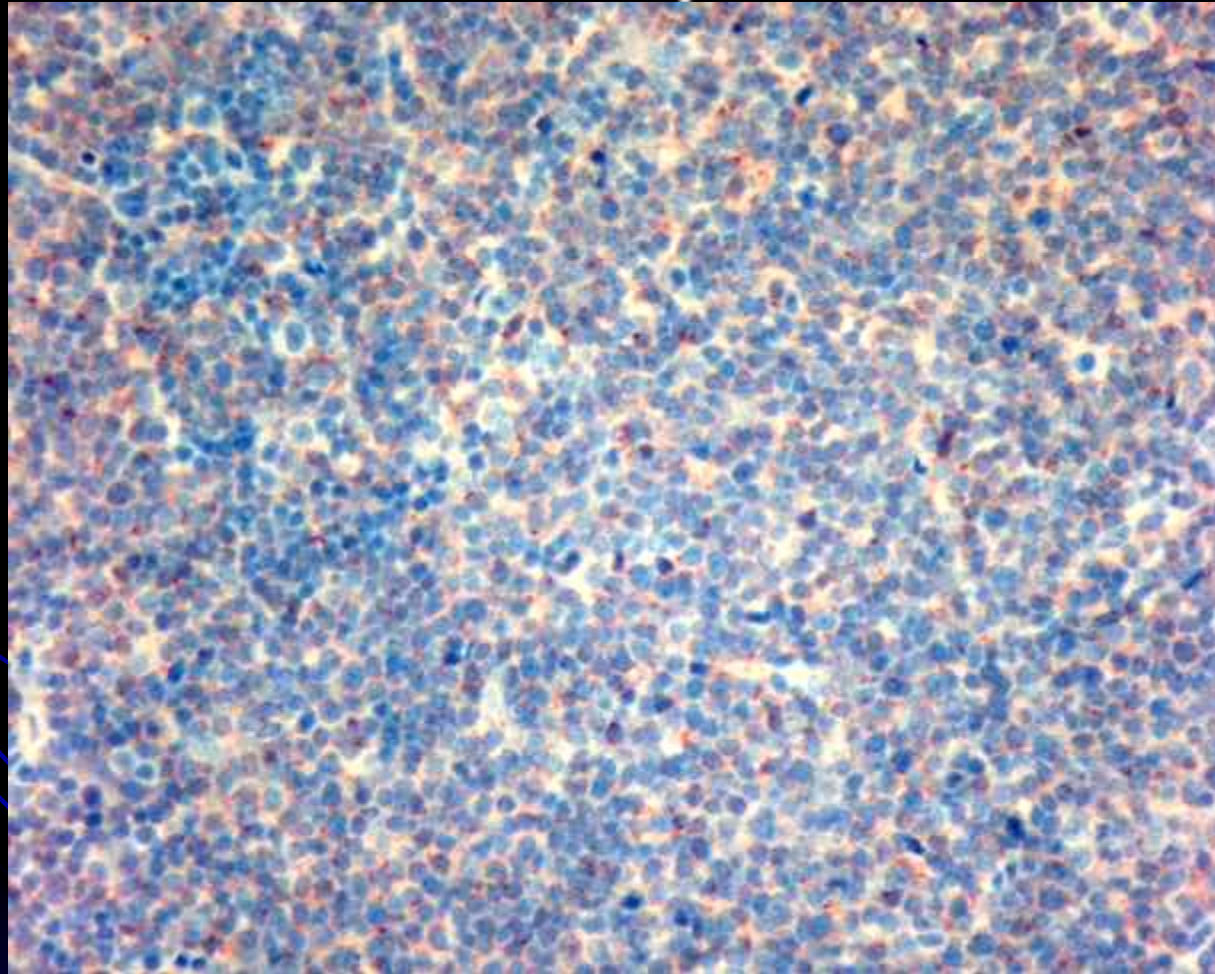
# Blastic Plasmacytoid Dendritic Cell Neoplasm



# Blastic Plasmacytoid Dendritic Cell Neoplasm

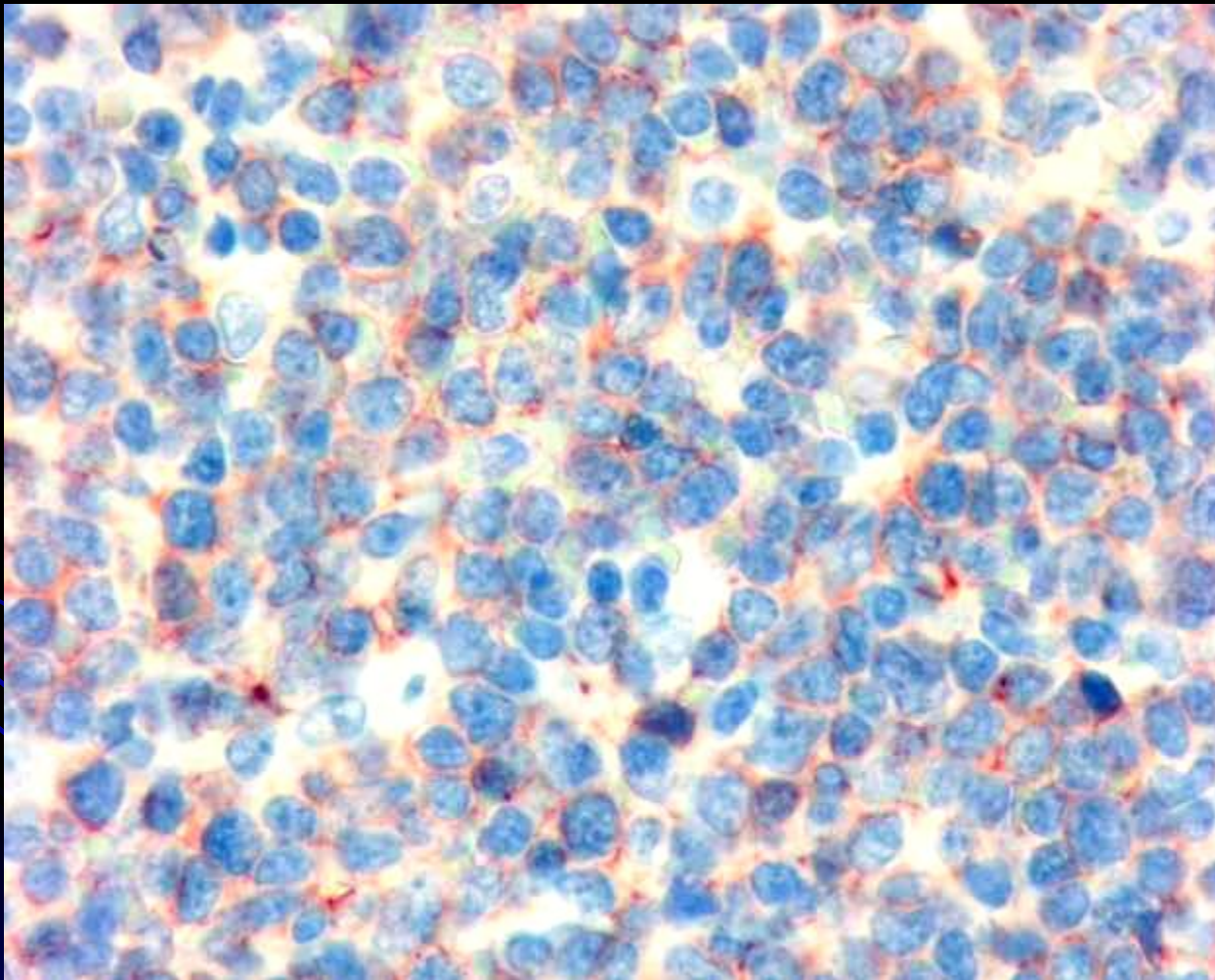


# Blastic Plasmacytoid Dendritic Cell Neoplasm



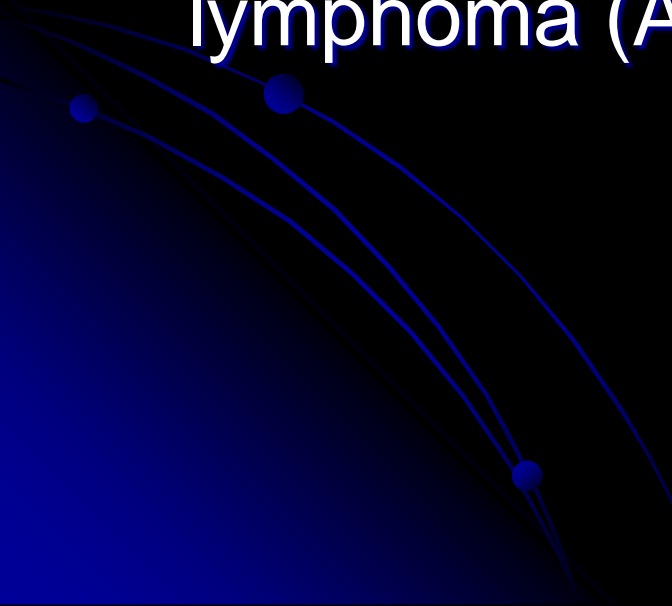
*CD56*

# Blastic Plasmacytoid Dendritic Cell Neoplasm



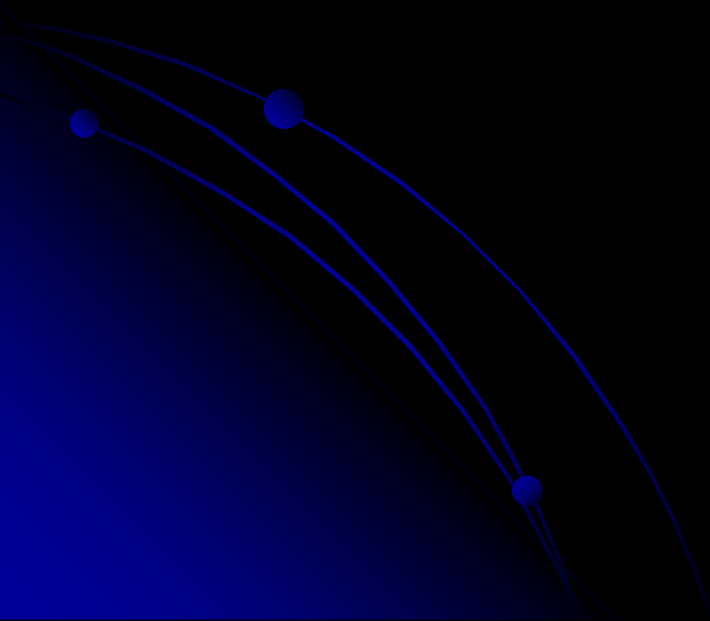
CD56

# Differential Diagnosis

- Mycosis fungoides
  - Extranodal T/NK-cell lymphoma, extra-nasal type
  - Primary cutaneous anaplastic large cell lymphoma (ALCL)
- 

# Genetics

- No specific chromosome abnormalities
- Germline T-cell receptor genes





# Prognosis

- Aggressive, poor response to therapy
  - Partial responses with “acute leukemia-like” therapy
  - Better prognosis in patients with localized skin lesions
- 