

Peripheral T-cell Lymphoma, unspecified

Peripheral T-Cell Lymphoma Unspecified

- T-cell lymphomas that don't meet the criteria for the more specific types
- About 50% of the T-cell lymphomas
- Mostly adults, but may occur in children
- Usually nodal, but may be extranodal
- Usually advanced stage at diagnosis

Peripheral T-Cell Lymphoma Unspecified

- Patients present with lymphadenopathy
- Constitutional symptoms often present
- Paraneoplastic features: eosinophilia, pruritus, hemophagocytic syndrome
- Aggressive clinical course
 - Patients respond poorly to treatment
 - Relapses are frequent
 - Overall 5 year survival 20-30%

Peripheral T-Cell Lymphoma



Scaly plaque



Large tumors

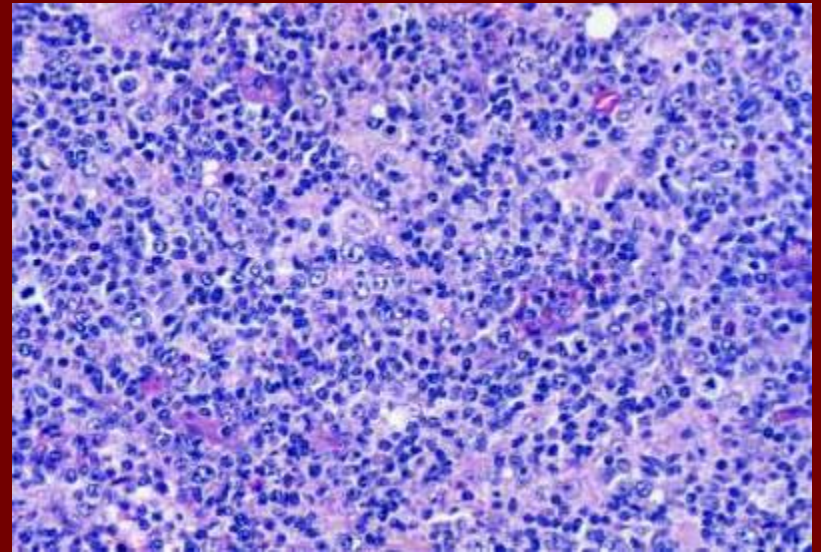
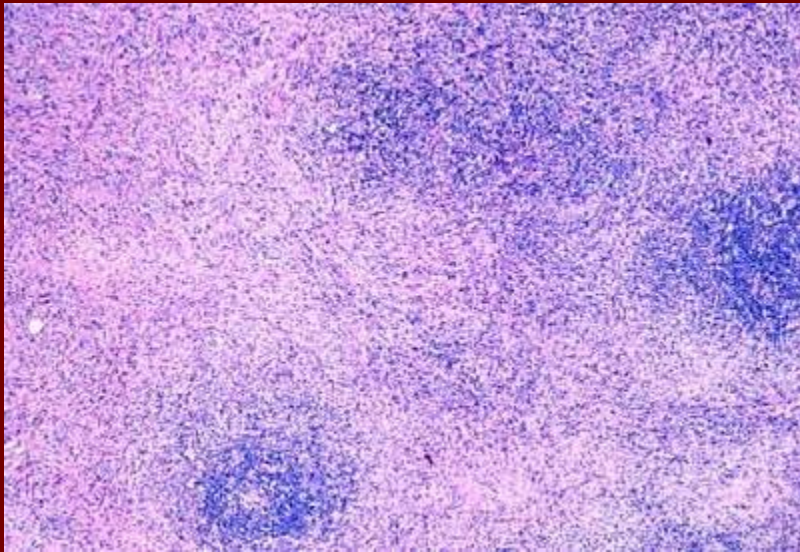
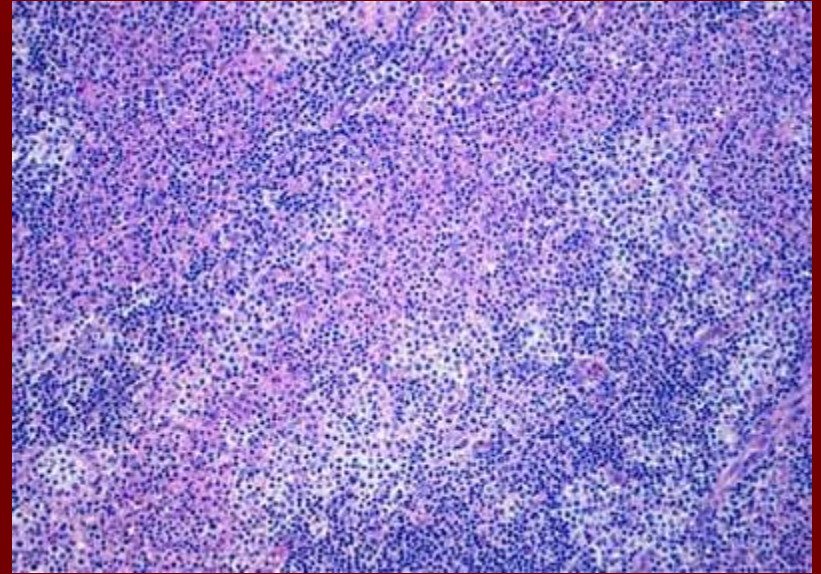
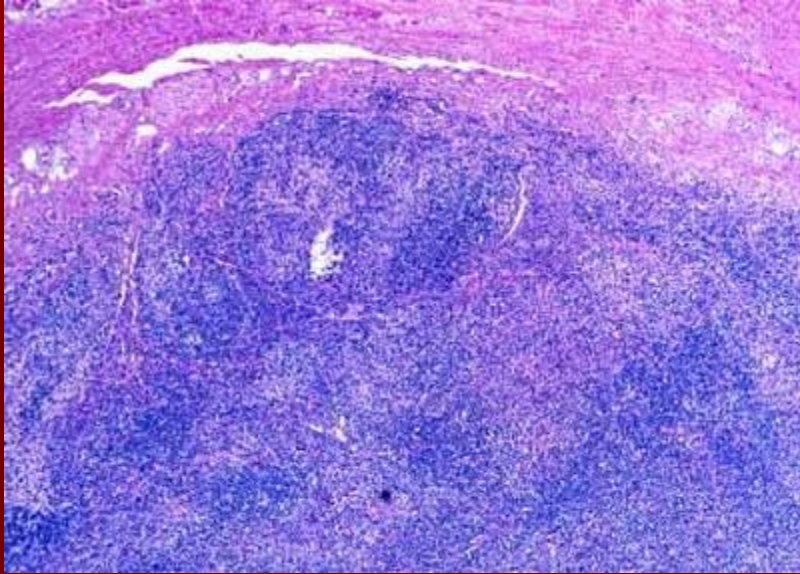
Peripheral T-Cell Lymphoma Unspecified

- Diffuse infiltration with effacement of lymph node architecture
- Broad cytologic spectrum: usually predominance of medium-sized or large cells with irregular nuclei
- Clear cells and Reed-Sternberg-like cells
- High endothelial venules increased
- Polymorphous inflammatory background

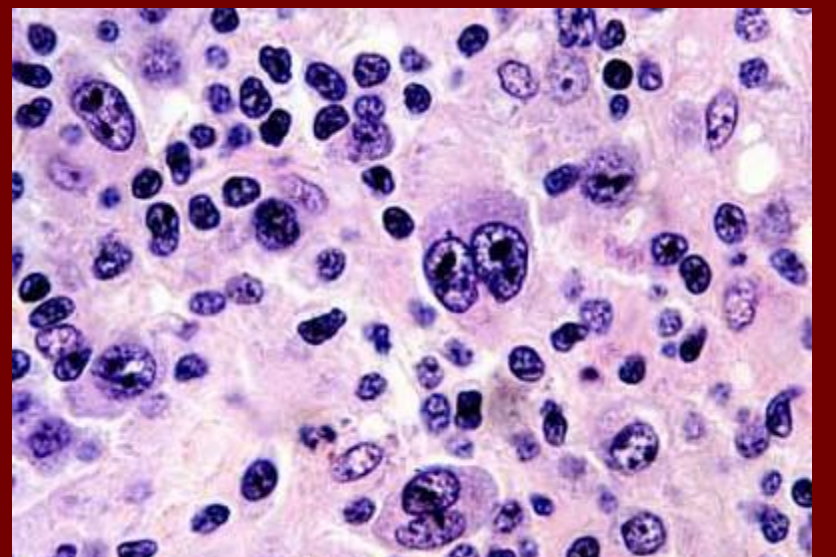
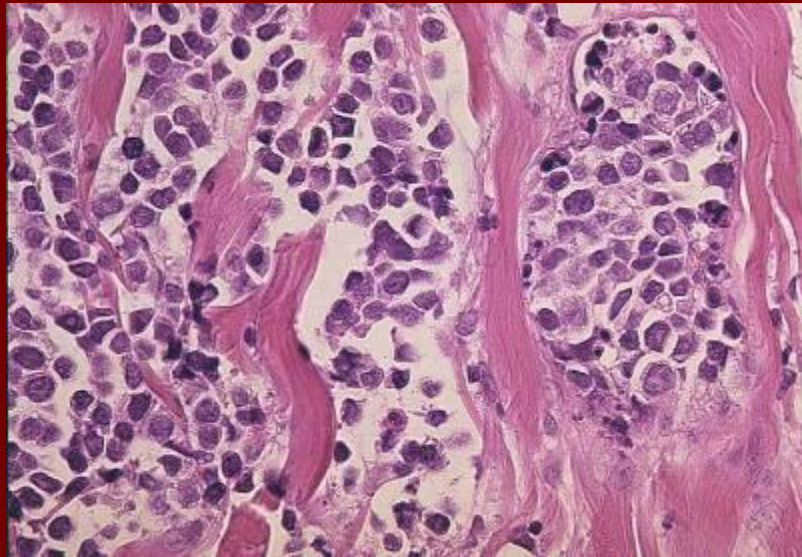
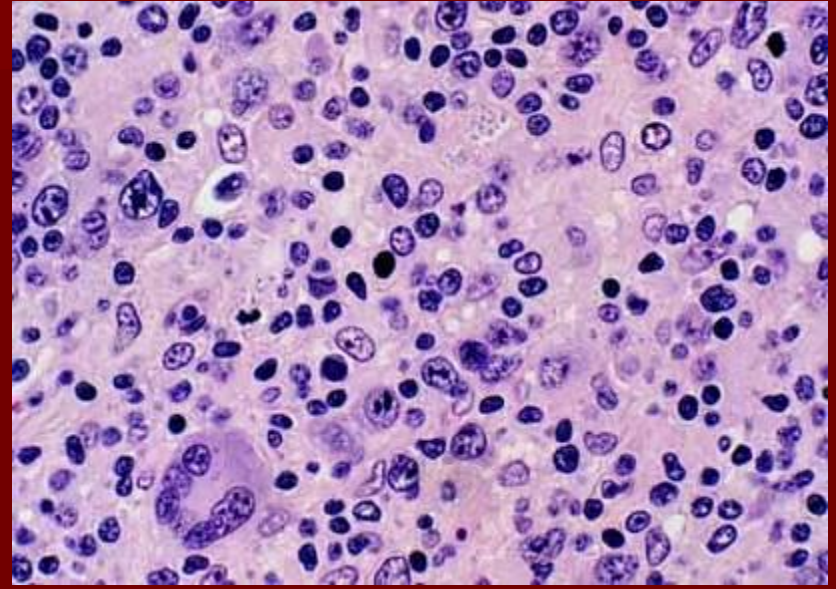
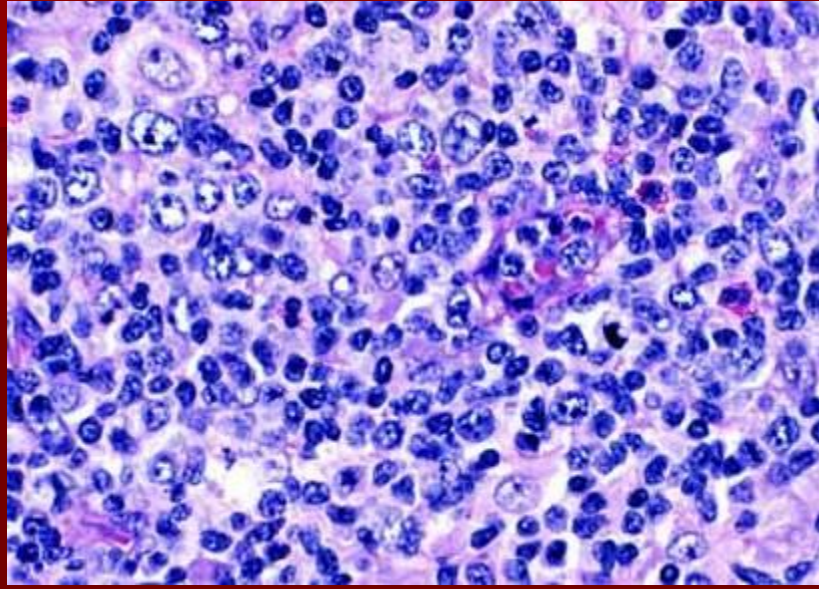
Peripheral T-Cell Lymphoma Unspecified

- T-zone variant
 - Interfollicular growth pattern with preserved or even hyperplastic follicles
 - Tumor cells predominantly small or medium-sized without nuclear pleomorphism
- Lymphoepithelial variant (Lennert lymphoma)
 - Diffuse or interfollicular
 - Numerous small clusters of epithelioid histiocytes

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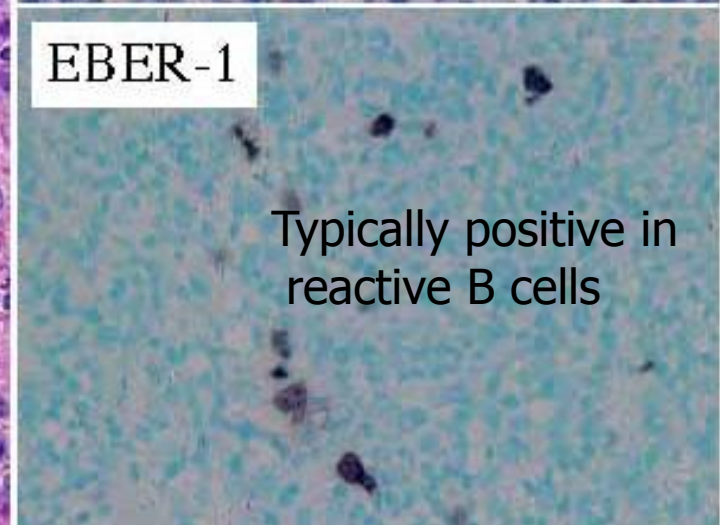
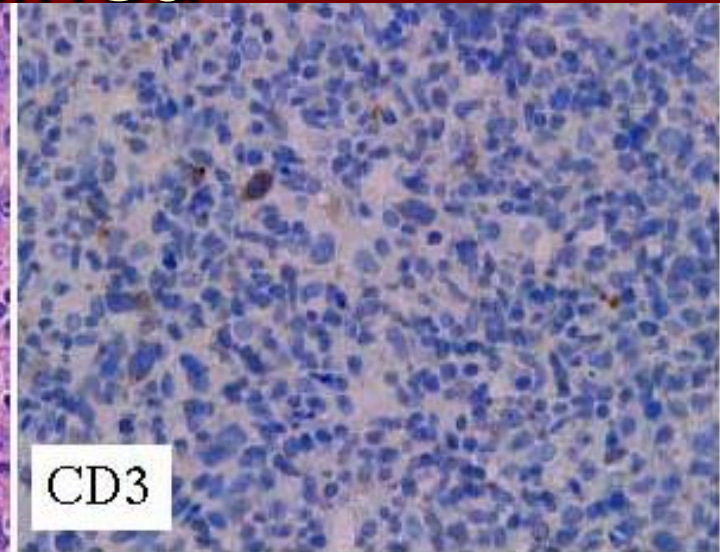
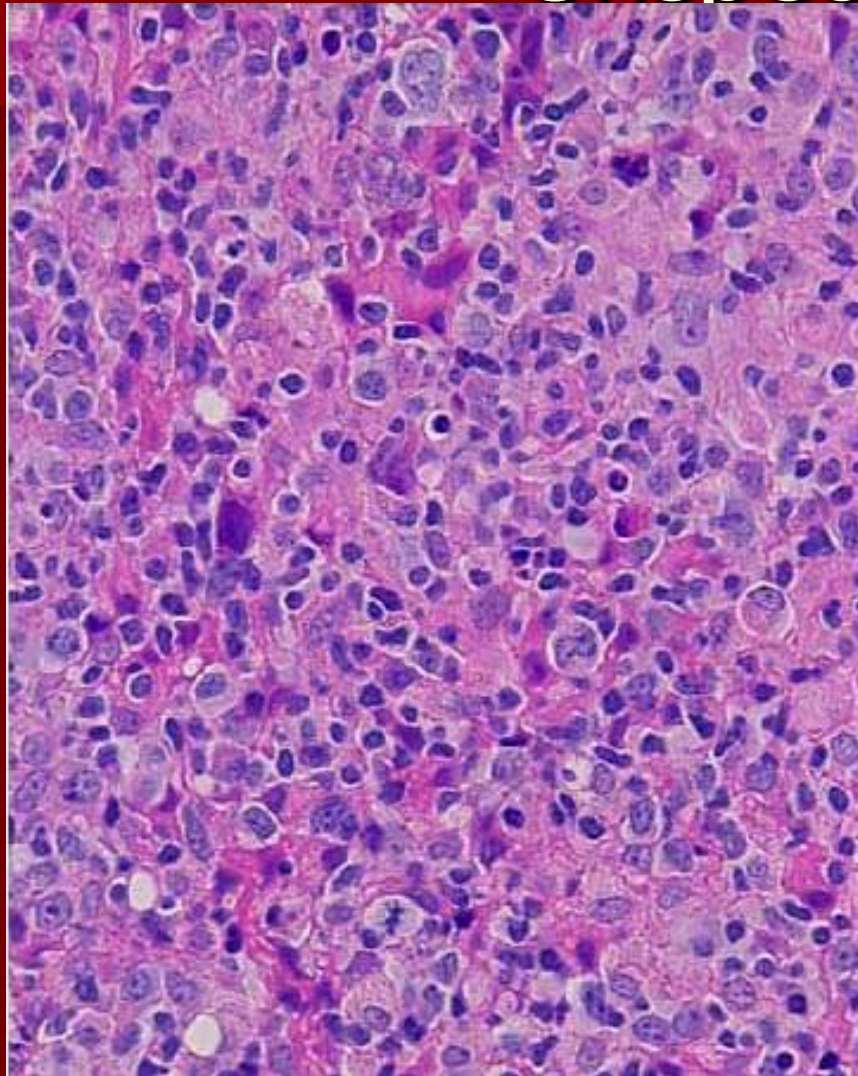
Immunophenotype

- T-cell associated antigens (CD3, CD5, CD7)
- Often show loss of normal antigen expression
- Most nodal cases are CD4+, CD8-
- CD30 may be positive, but not cytotoxic granule associated proteins
- Some cases may express CD56, usually extranodal with cytotoxic T-cell phenotype

■ Genetics

- TCR genes clonally rearranged in most cases

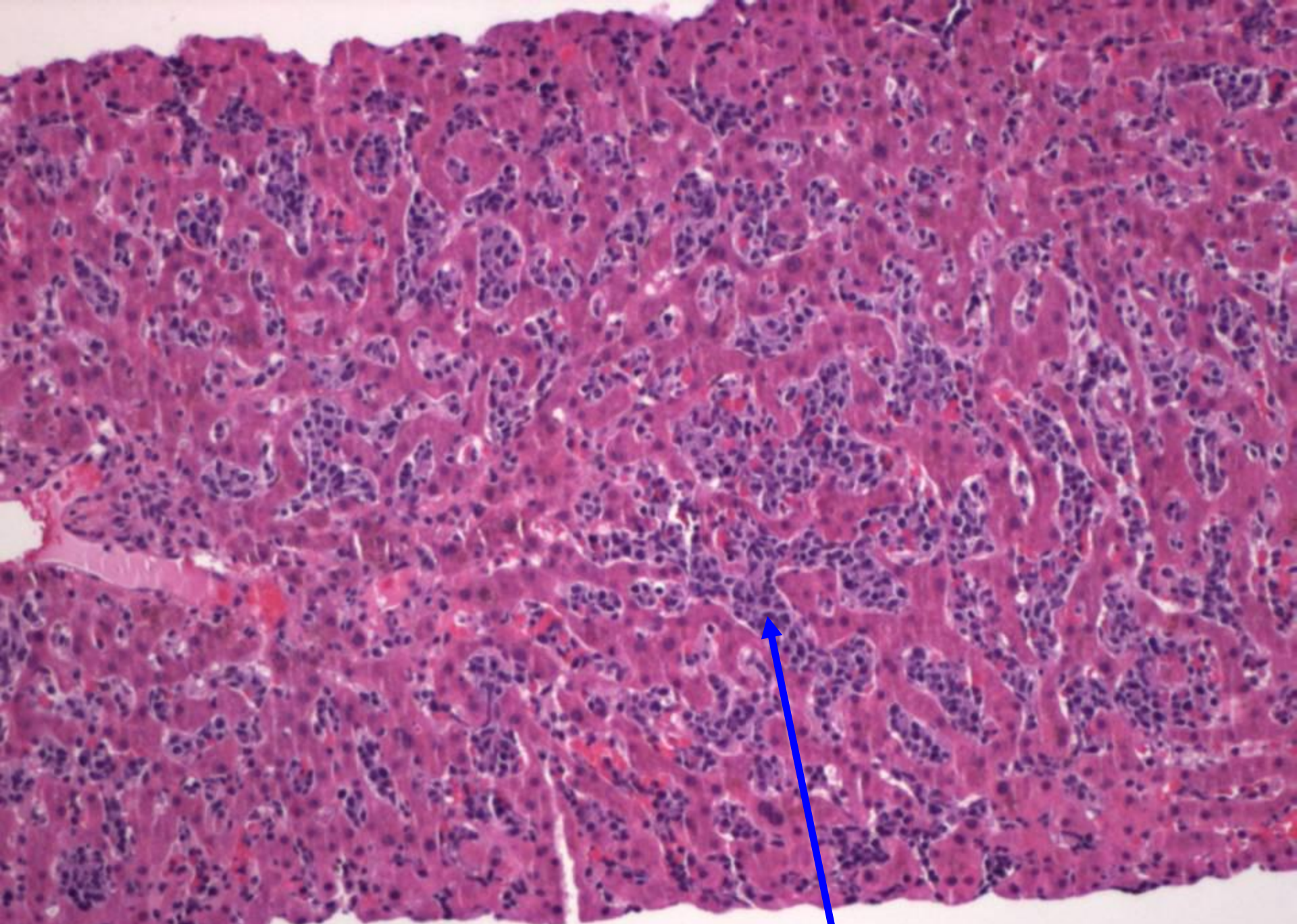
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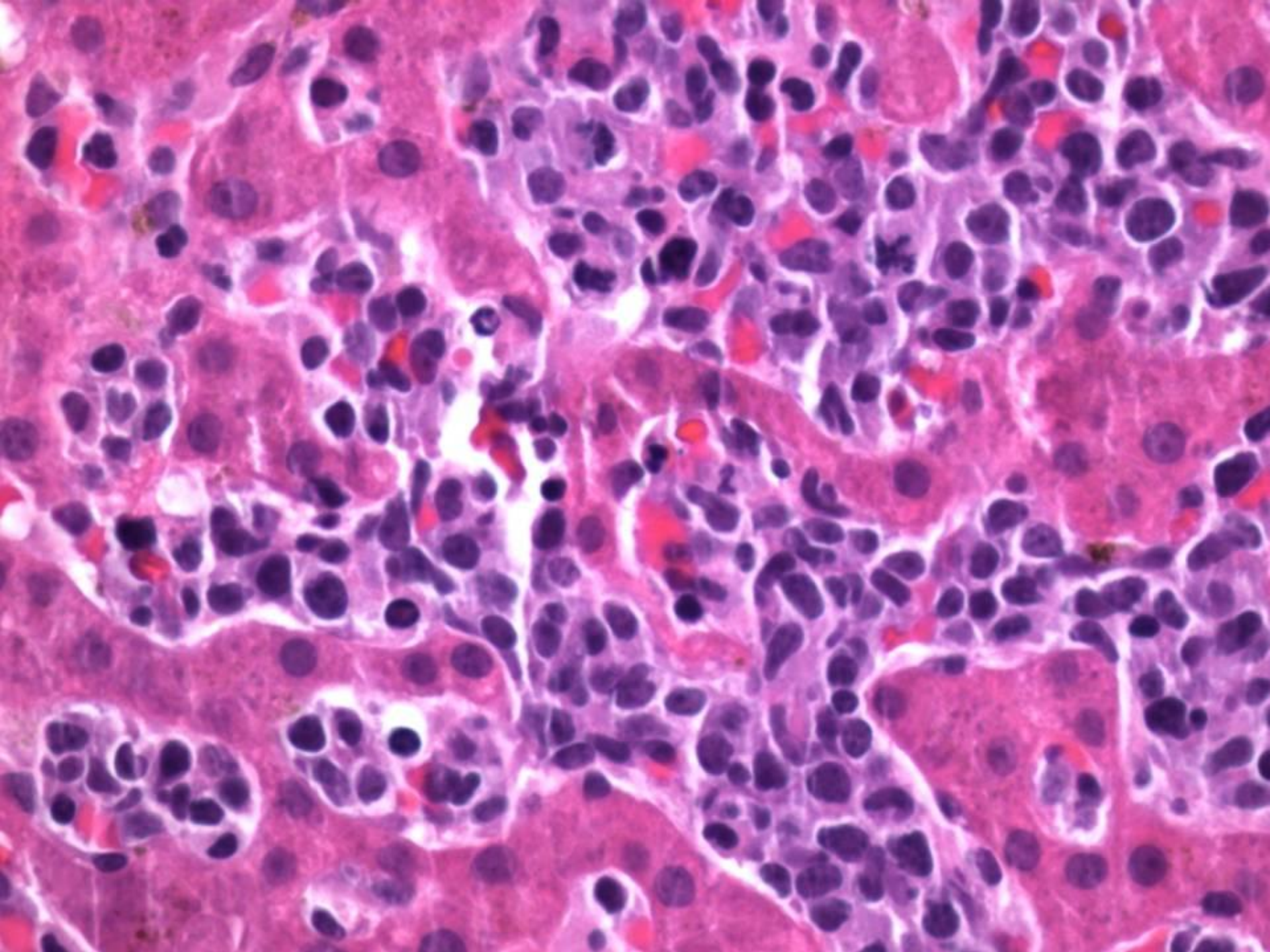
Hepatosplenic T-cell Lymphoma

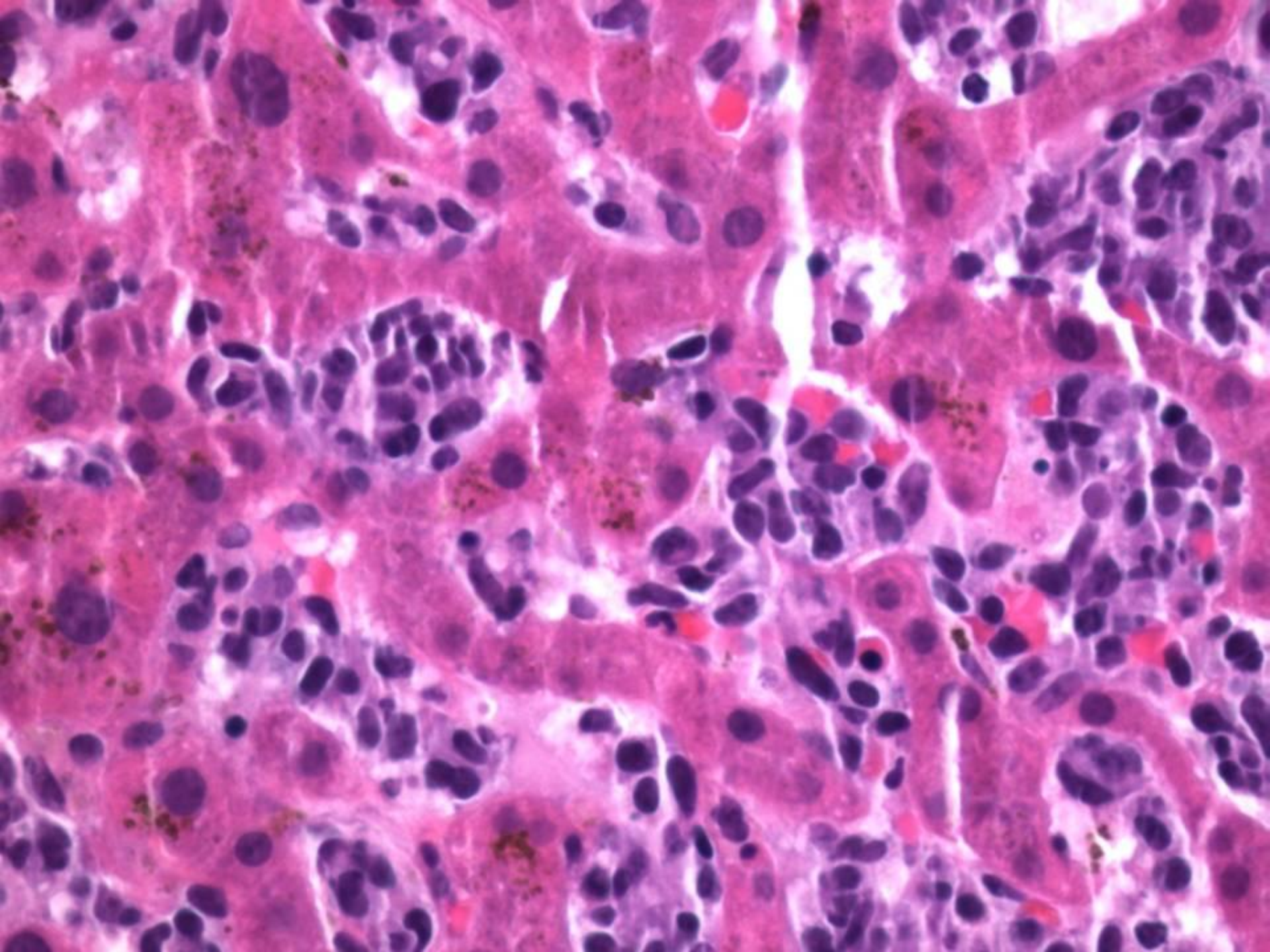
Hepatosplenic T-cell Lymphoma

- Extranodal and systemic lymphoma usually of cytotoxic T-cells of the $\gamma\delta$ type
- Marked sinusoidal infiltration seen in the spleen, liver and bone marrow
- Patients present with marked hepatosplenomegaly but no lymphadenopathy
- Bone marrow almost always involved
- More common in immunosuppressed pts



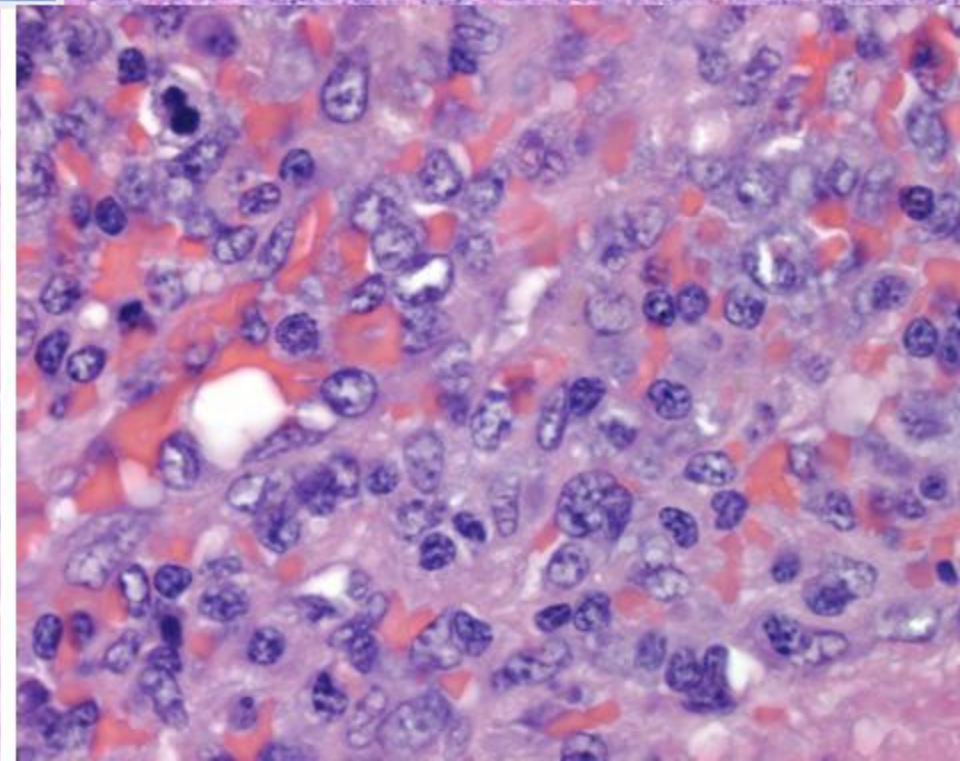
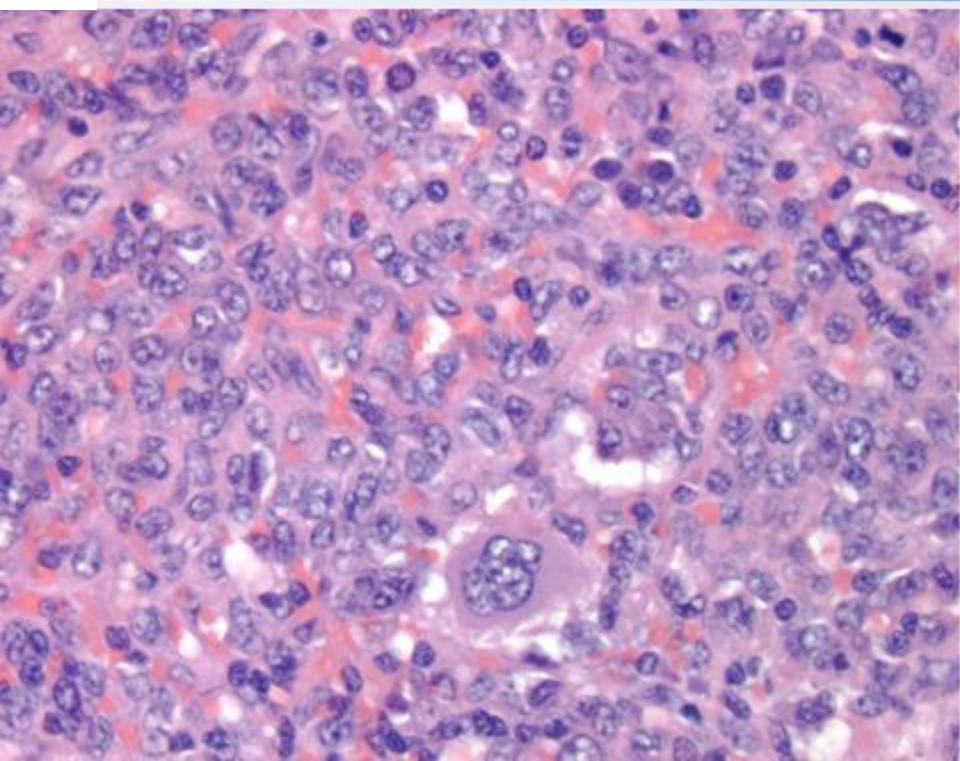
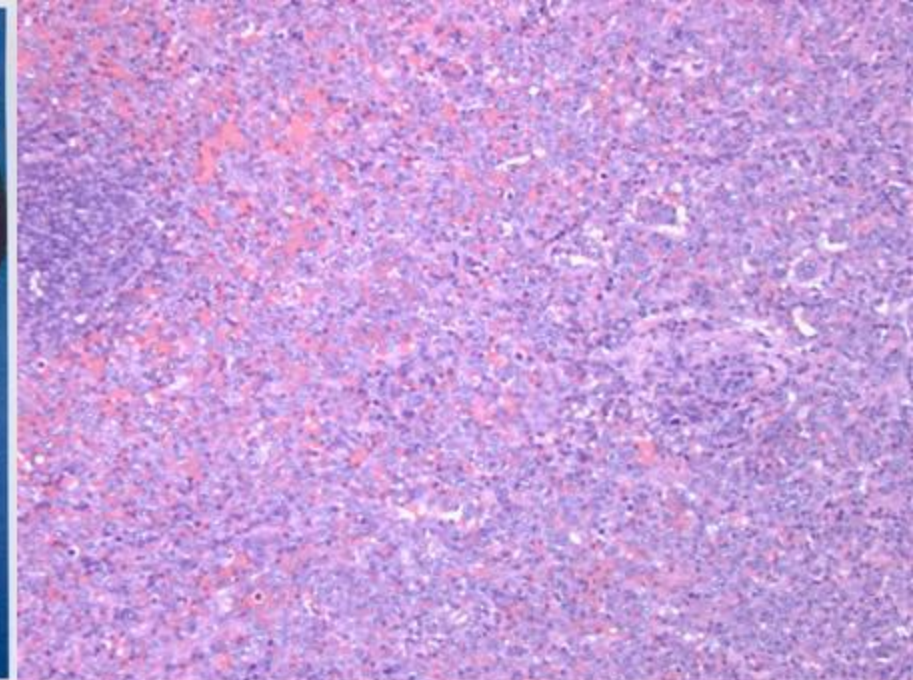
Sinusoidal infiltrate



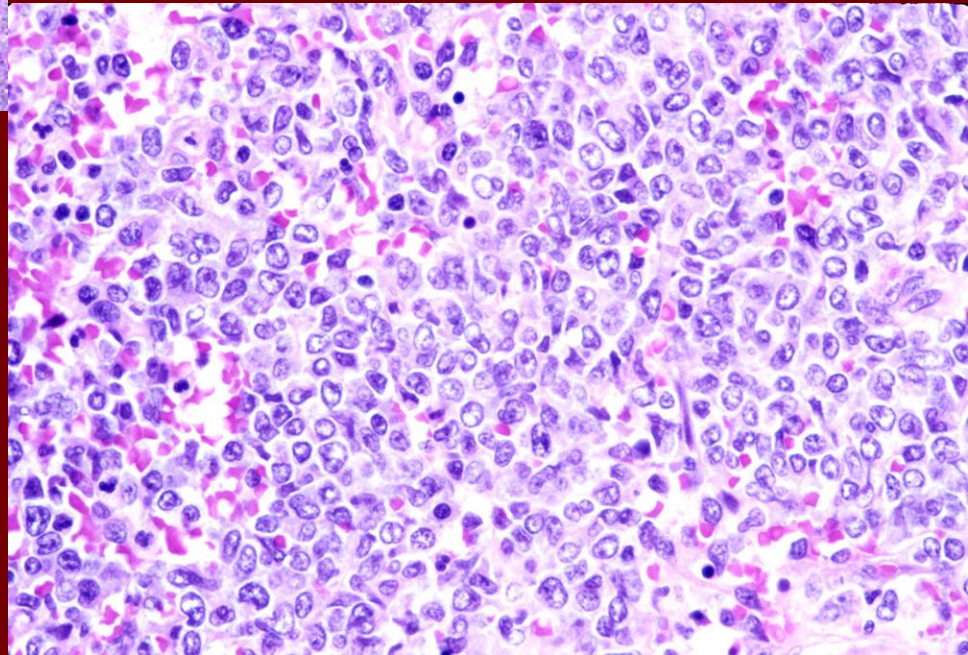
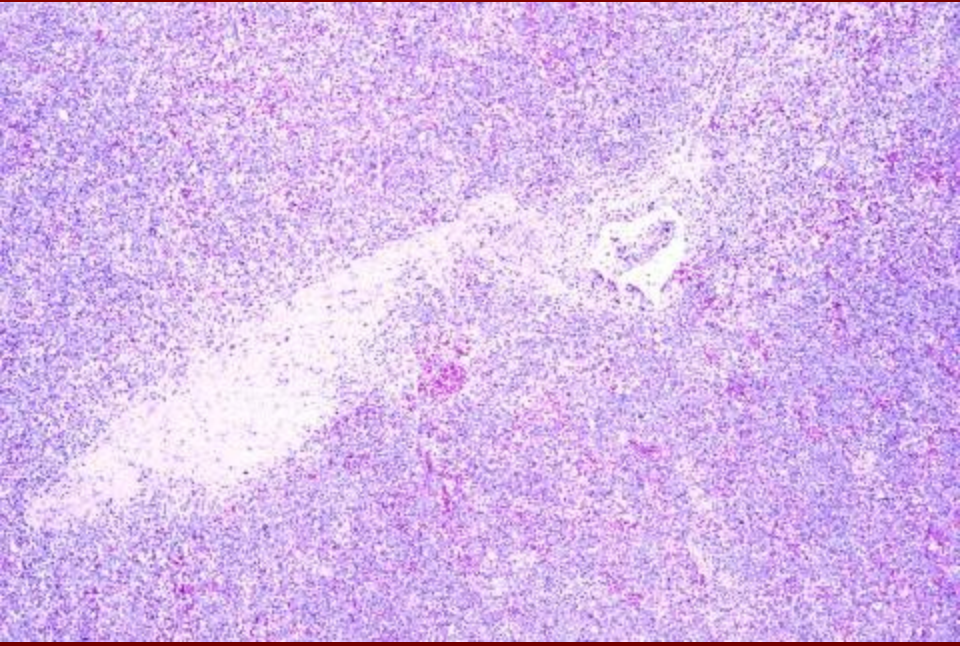




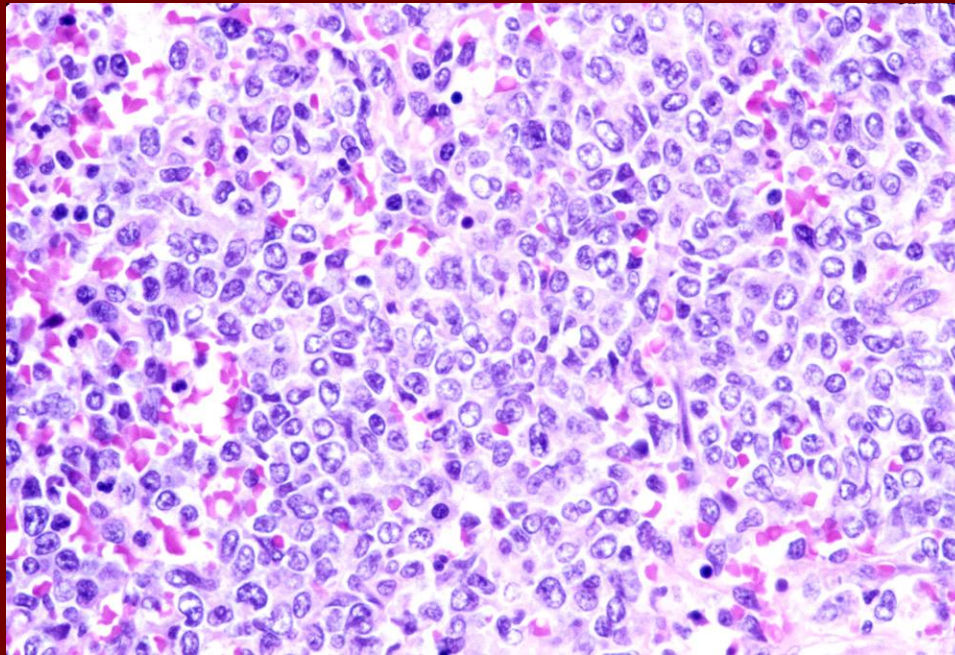
59y/o woman with fatigue.



Hepatosplenic T-cell Lymphoma



Hepatosplenic T-cell Lymphoma



Hepatosplenic T-cell Lymphoma

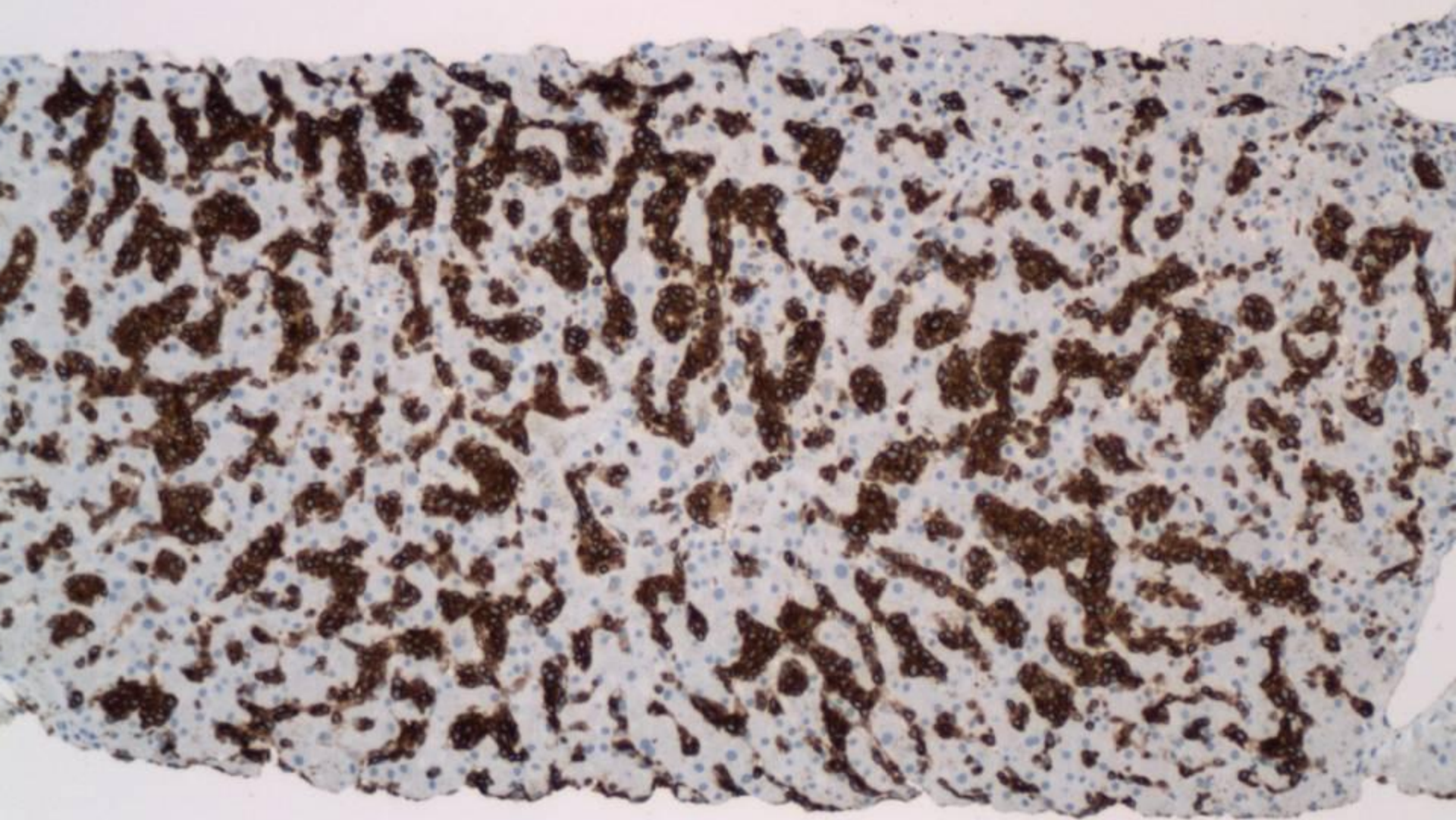
■ Immunophenotype

- CD3+, CD4-, CD8-, CD5-
- TCR δ 1+, TCR $\alpha\beta$ -
- Positive for cytotoxic protein TIA-1

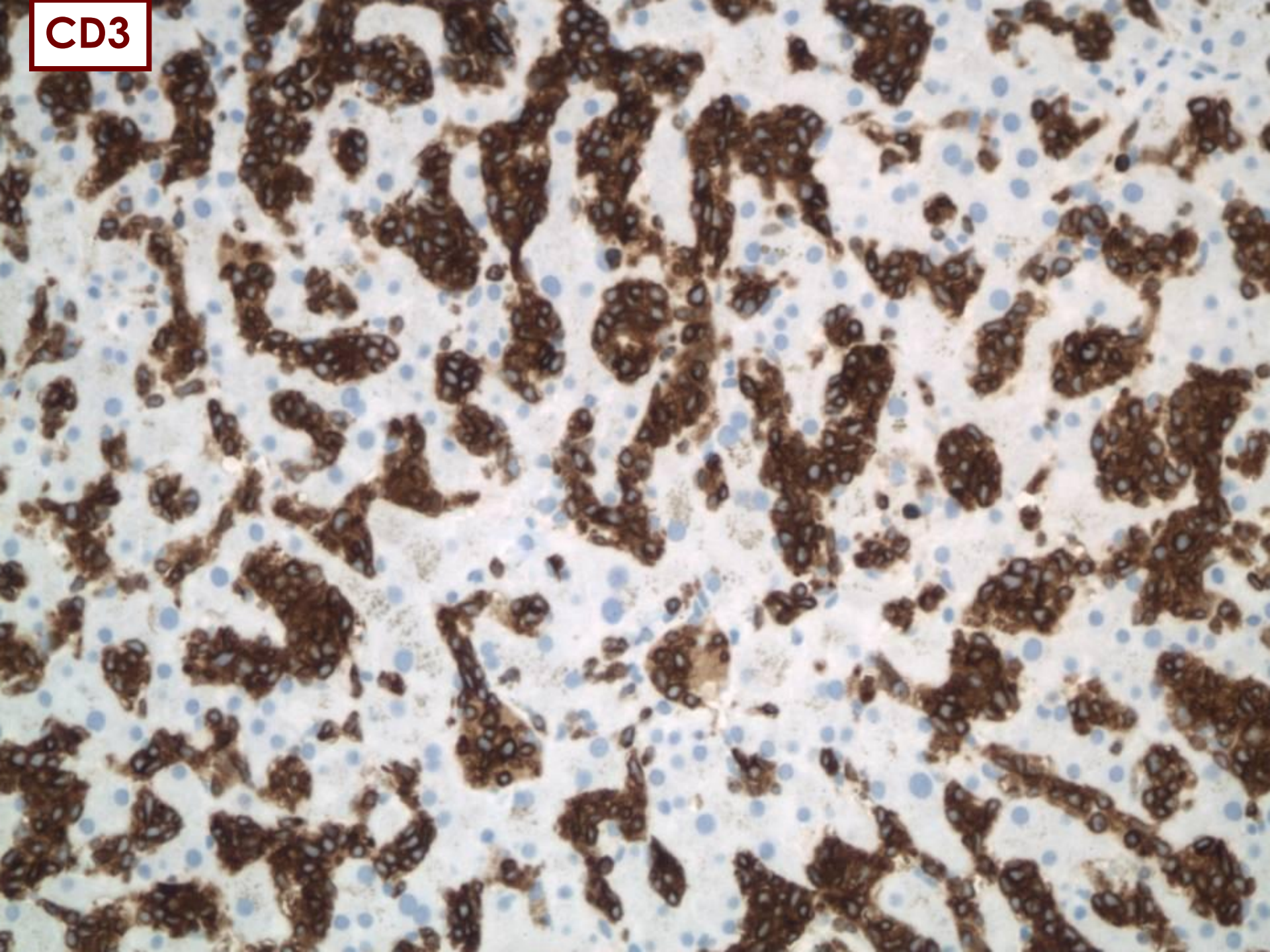
■ Genetics

- TCR γ gene rearrangement
- Isochromosome 7q in all cases studied
- Sometimes other abnormalities such as trisomy 8

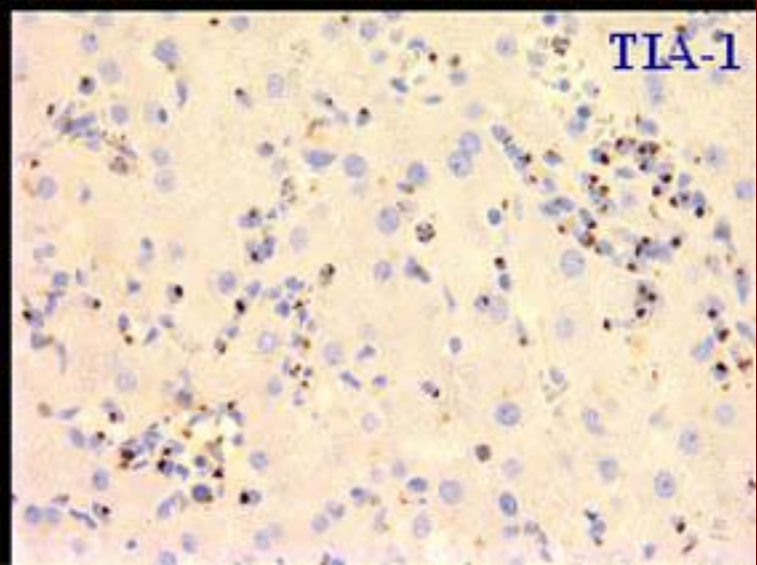
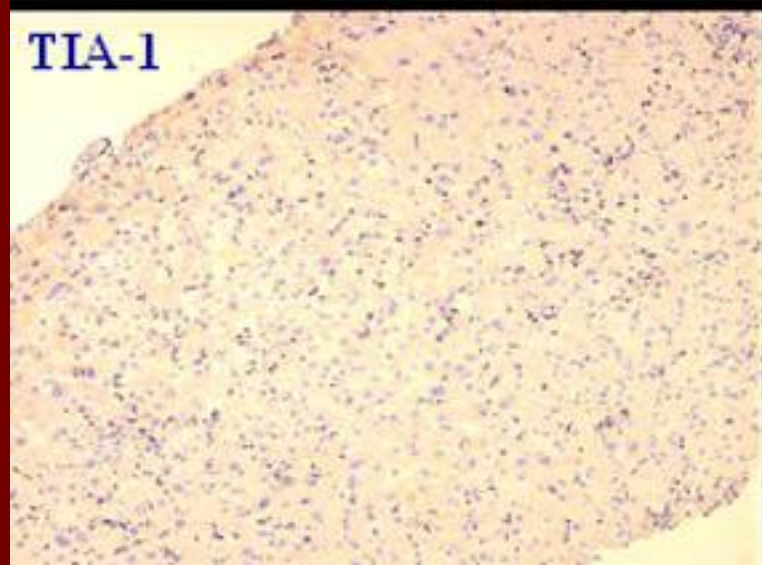
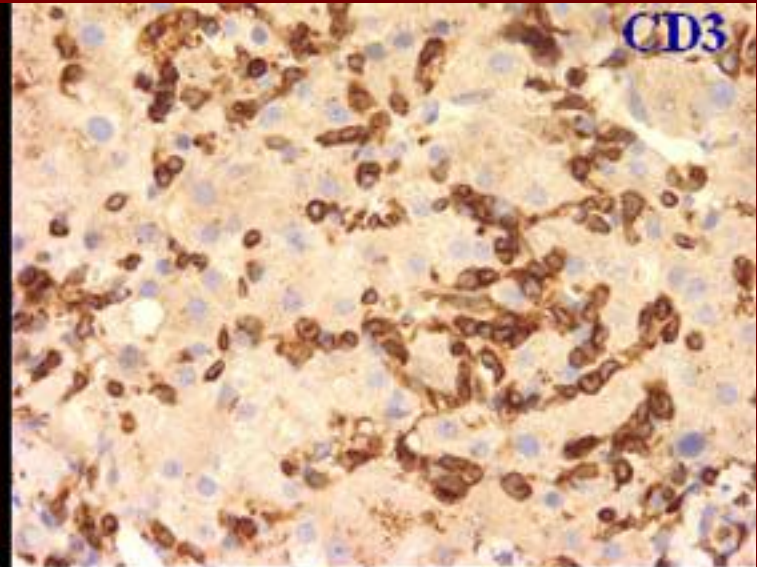
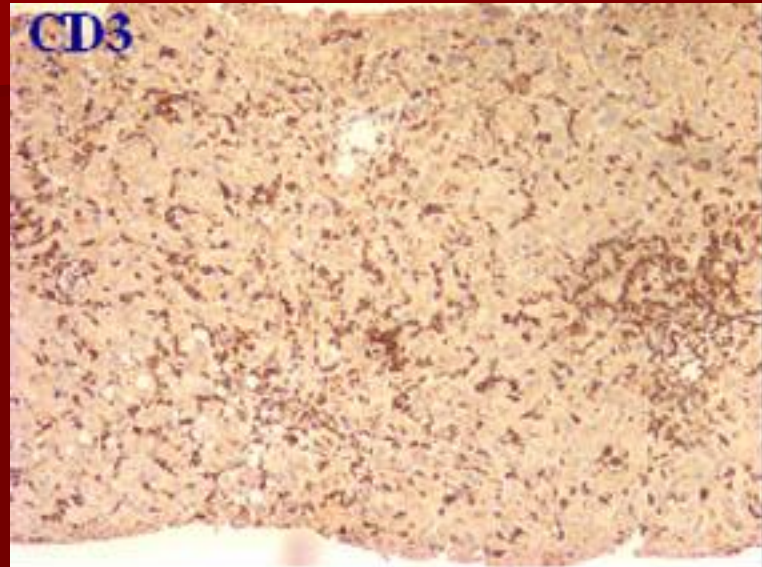
CD3



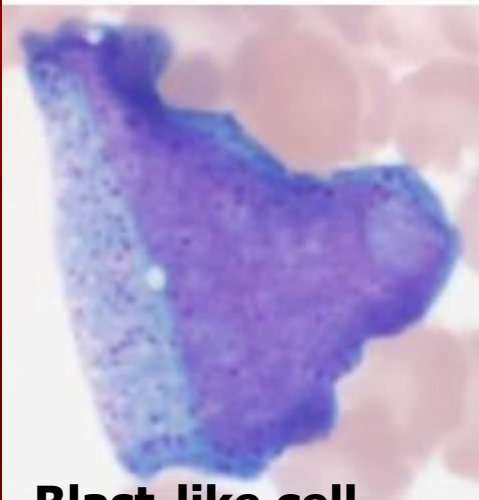
CD3



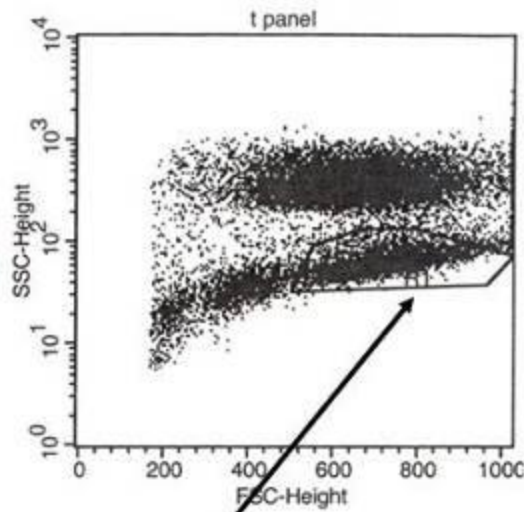
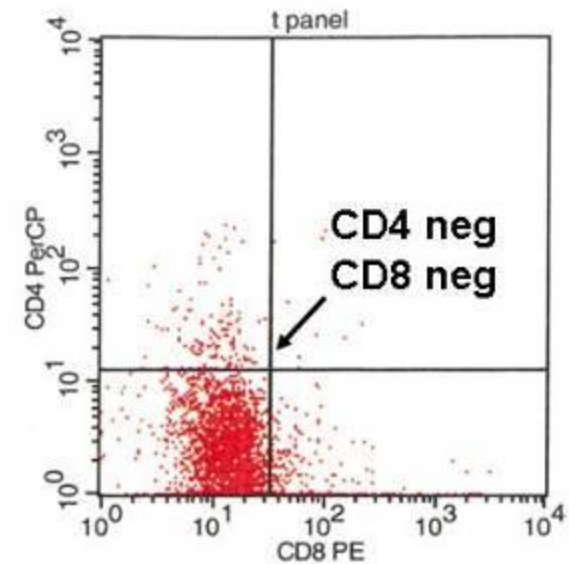
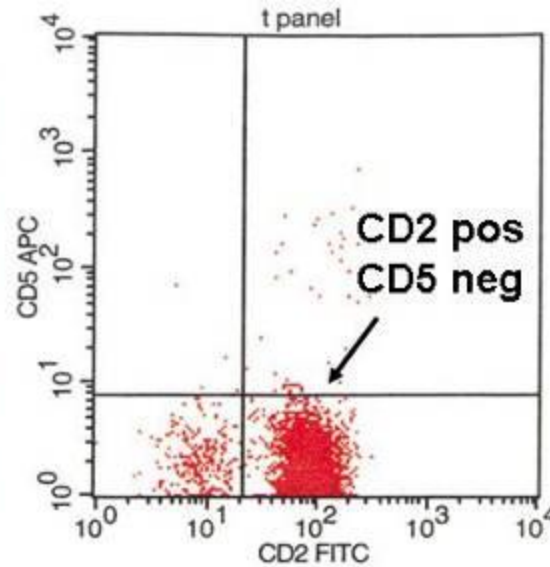
Hepatosplenic T-cell Lymphoma



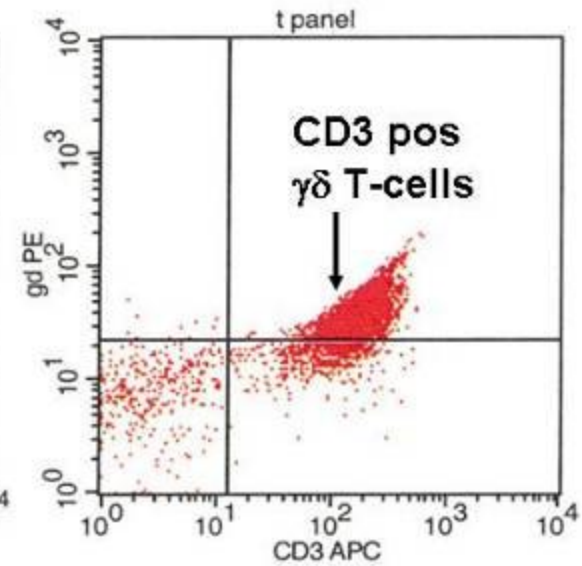
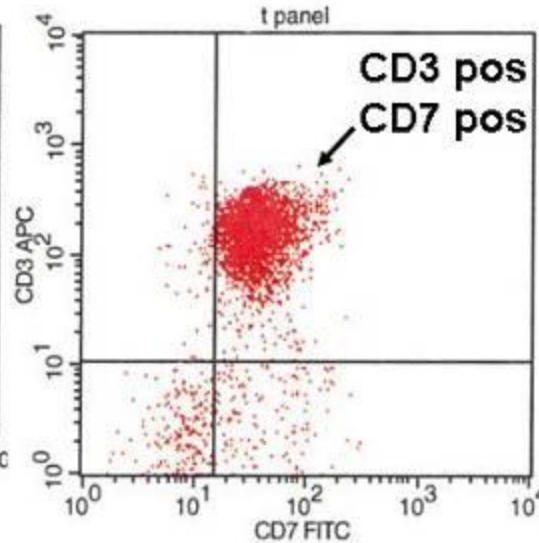
Hepatosplenic T-cell Lymphoma



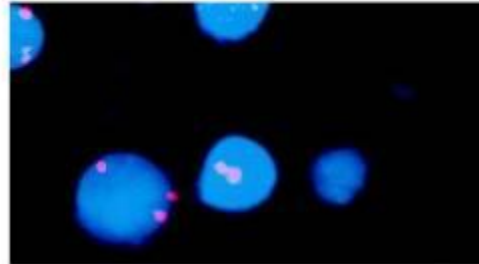
Blast-like cell



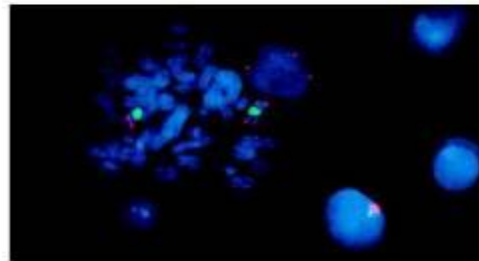
**Large Cells
By Light Scatter**



Hepatosplenic T-cell Lymphoma



Trisomy 8: Red chromosome
8 centromere



Isochromosome 7:
Green: chromosome 7
Centromere
Red: 7q3.1



**Clonal V-delta-1-J-delta-1 rearrangement
(Panel B, lane 1)**

Extranodal NK/T-cell Lymphoma, Nasal Type

Extranodal NK/T-cell Lymphoma, Nasal Type

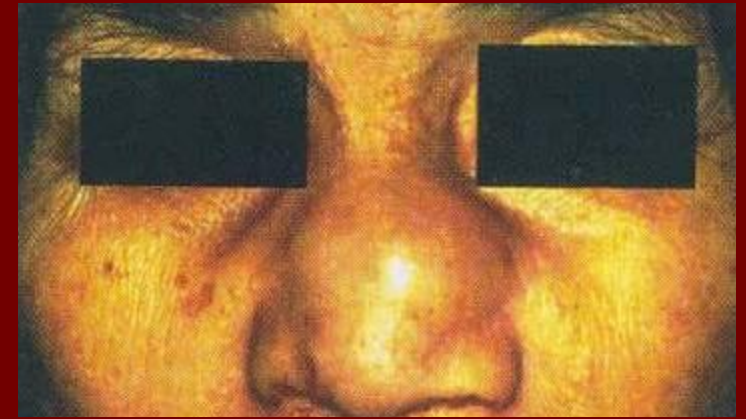
- Predominantly extranodal lymphoma with broad morphologic spectrum
- Nasal cavity most common site, but may occur anywhere
- More prevalent in Asia, Mexico and Central and South America
- M>F
- May occur in immunosuppressed and post-transplant patients

Extranodal NK/T-cell Lymphoma, Nasal Type

- Present with nasal obstruction or epistaxis due to mass lesion or extensive midfacial destructive lesions
- Variable presentation outside nasal cavity, e.g., skin ulceration, intestinal perforation
- May disseminate rapidly
- May have associated hemophagocytic syndrome
- May overlap with aggressive NK cell leukemia

Extranodal NK/T-cell Lymphoma, Nasal Type

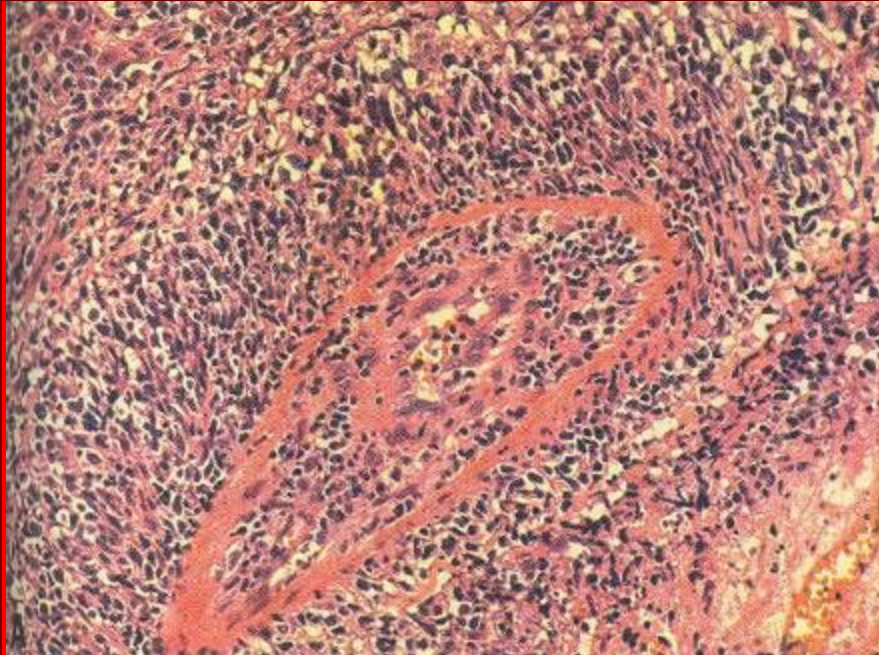
- Mucosal sites show extensive ulceration
- Diffuse infiltrate
- Angiocentric and angiodestructive pattern common with fibrinoid changes in vessels
- Coagulative necrosis and apoptotic bodies
- Cytologic spectrum broad from small to large anaplastic cells



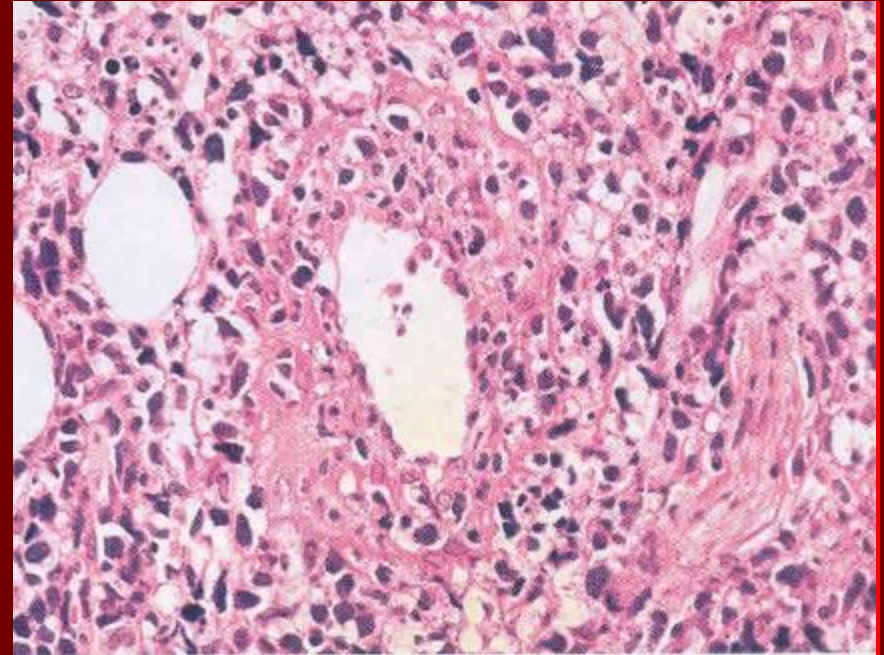
Expansion of the nasal bridge



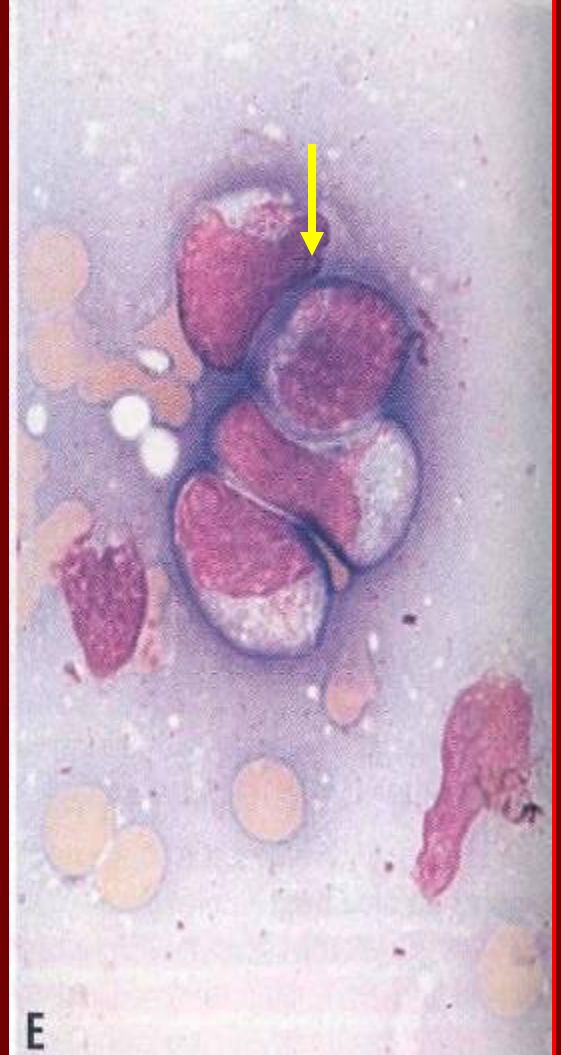
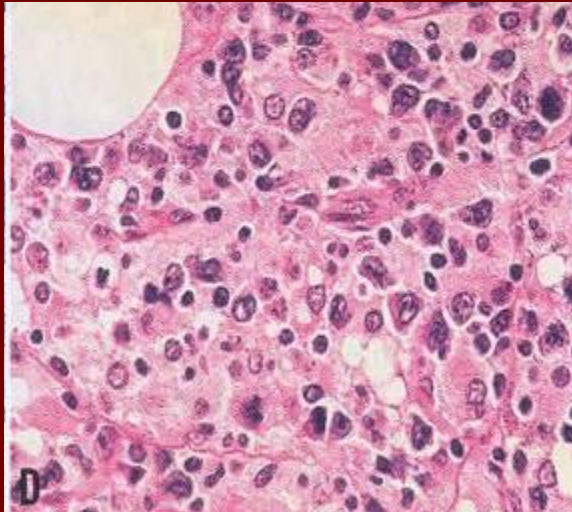
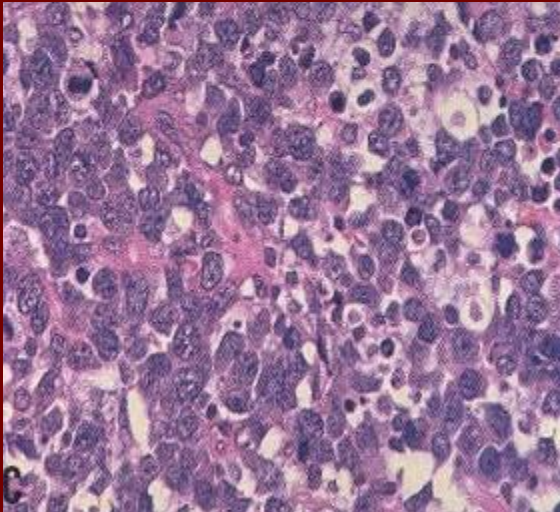
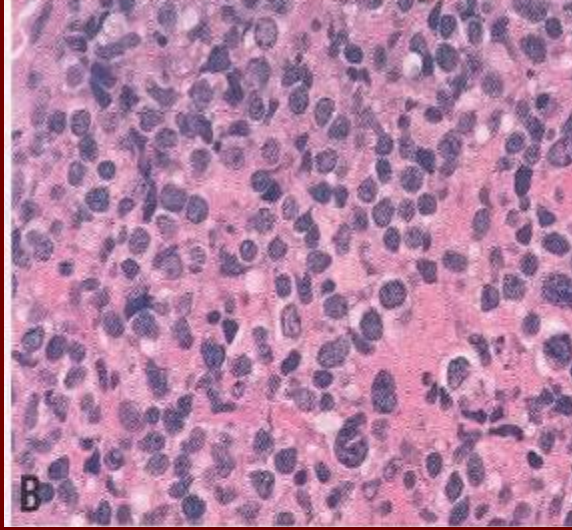
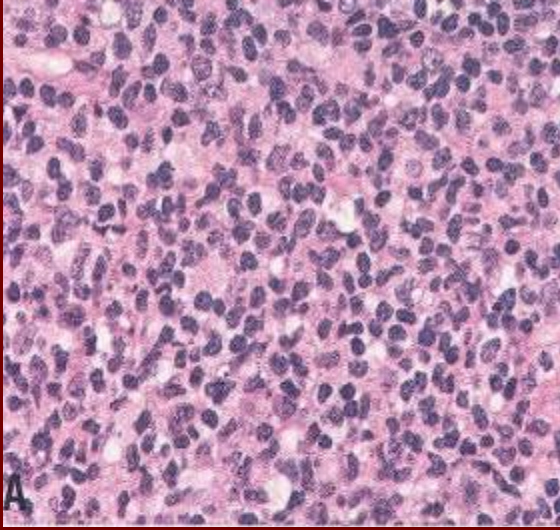
CT: tumor extends into the orbit



**Lymphocytic infiltrate with
destruction
of an artery**



**The lymphoma in skin with
angiocentric
angiodestructive property**



Extranodal NK/T:

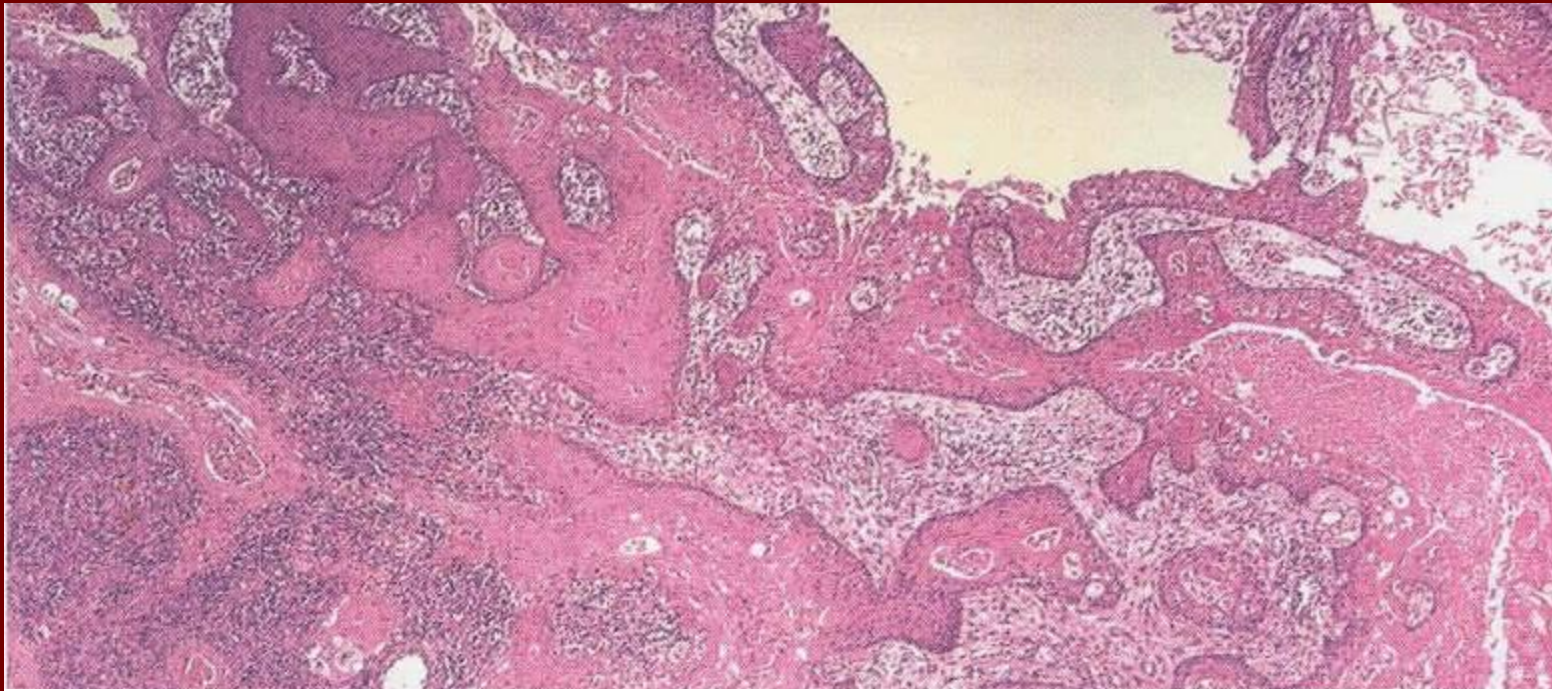
A-predominantly small cells

B-predominantly medium-sized cells

C-predominantly large cells with apoptosis

D-pleomorphic large cells with small cells

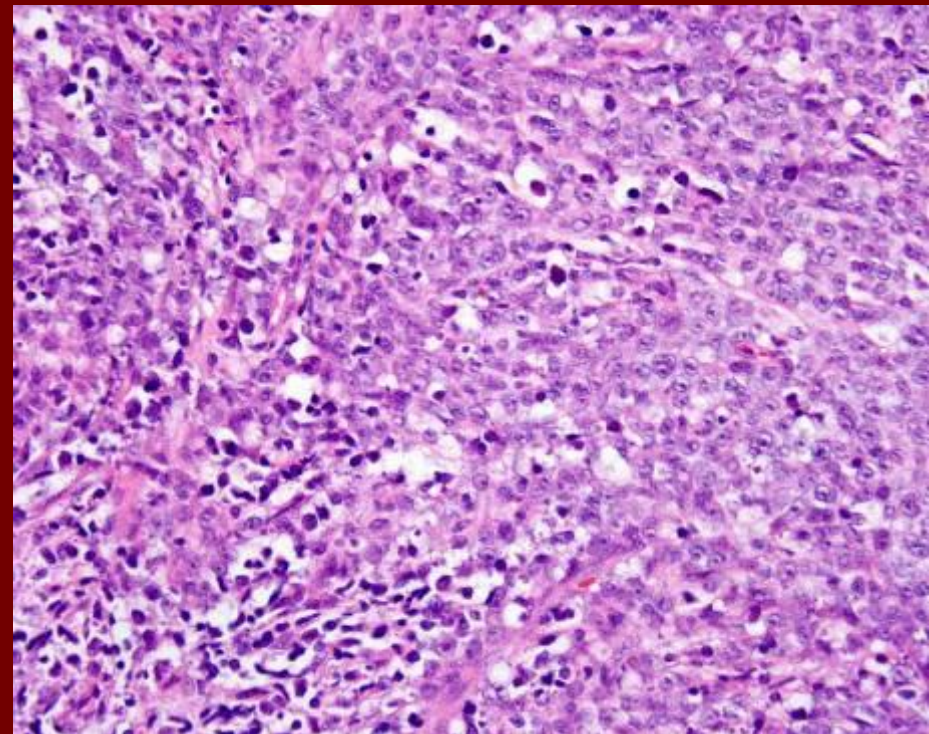
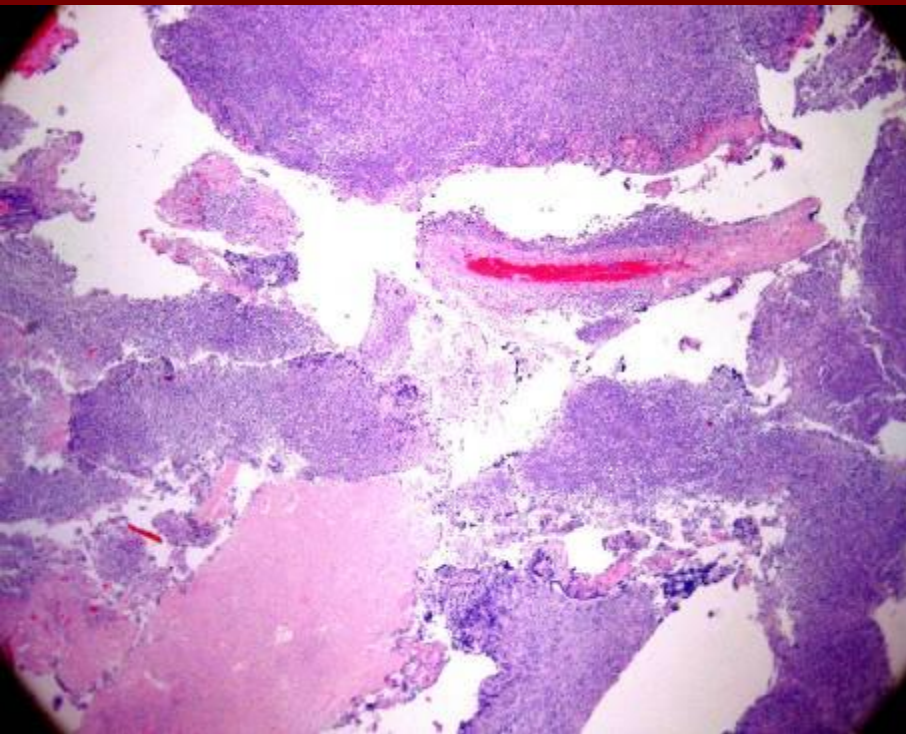
E-touch prep showing azurophilic granules



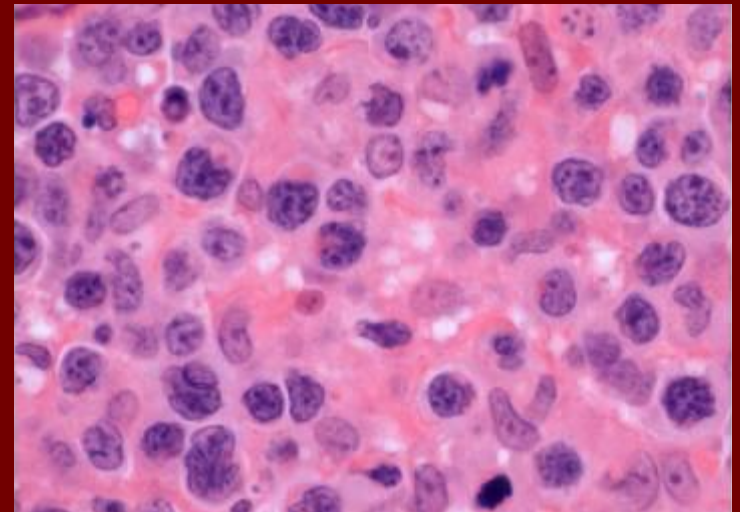
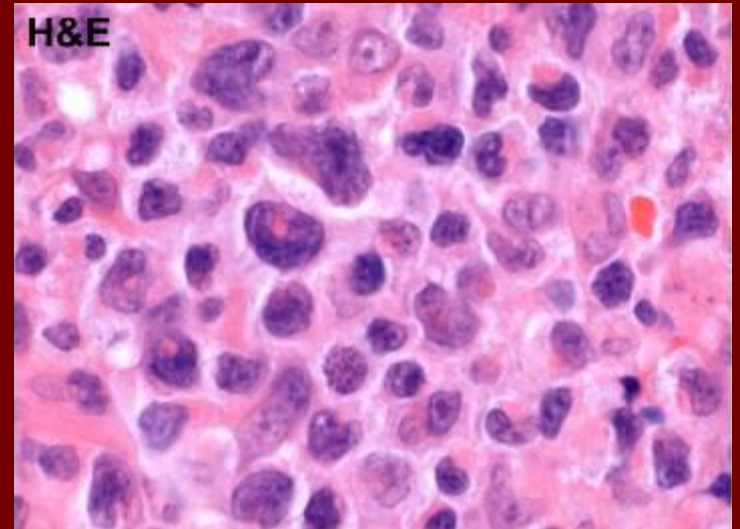
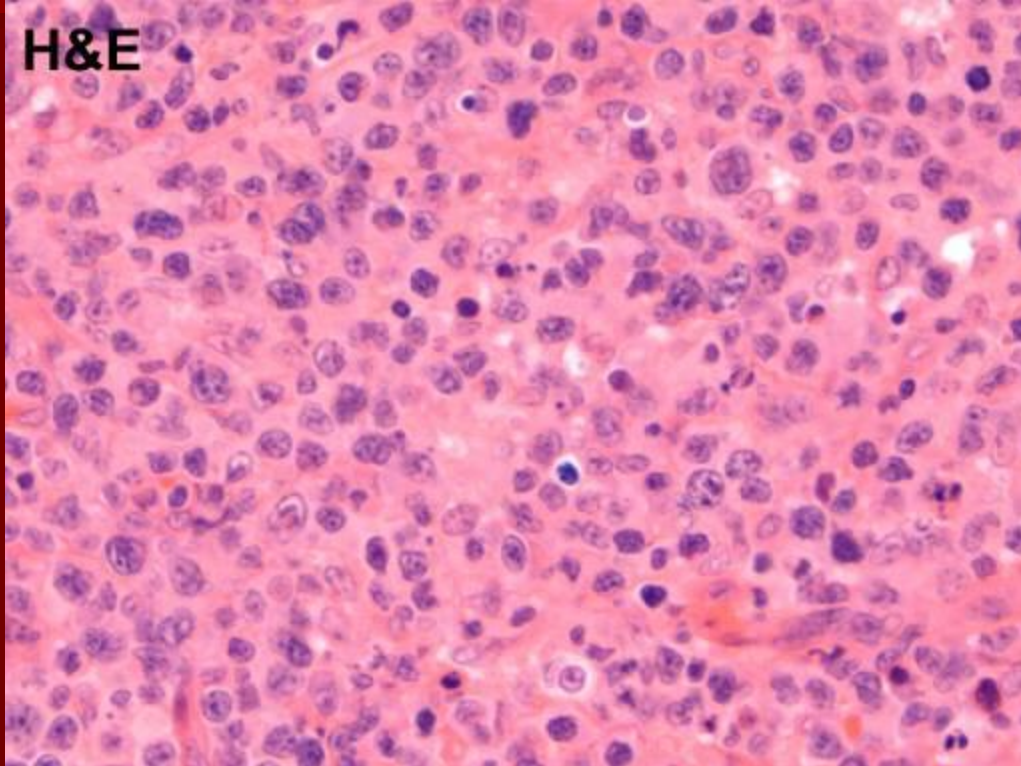
Nasal NK/T cell lymphoma with pseudoepitheliomatous hyperplasia
In mucosa epithelium mimicking squamous cell carcinoma

Extranodal NK/T-cell Lymphoma, Nasal Type

Nasal Cavity

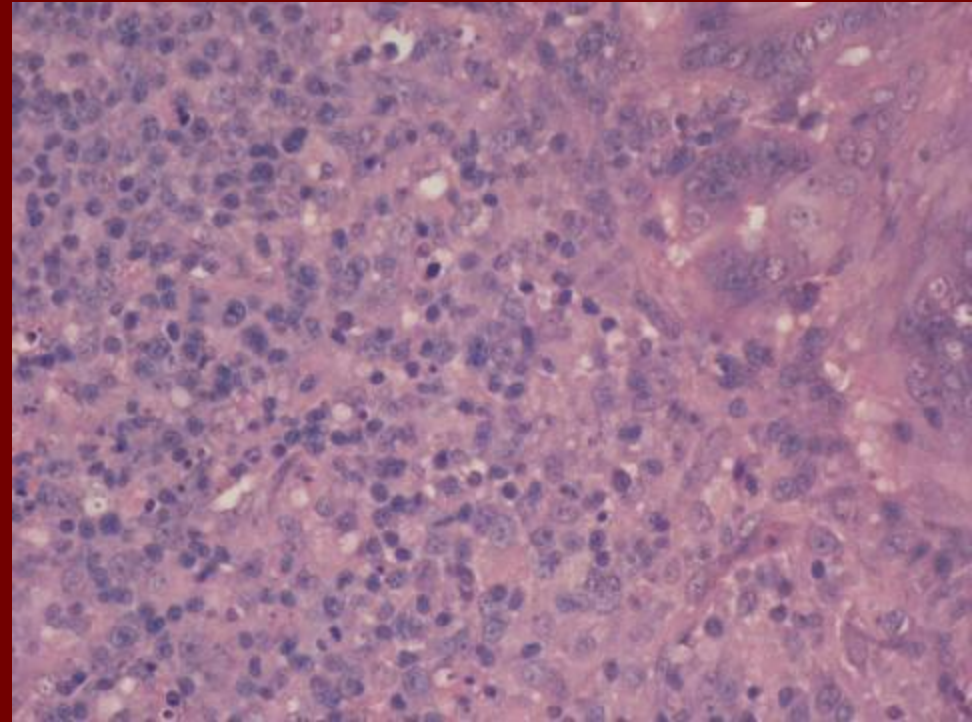
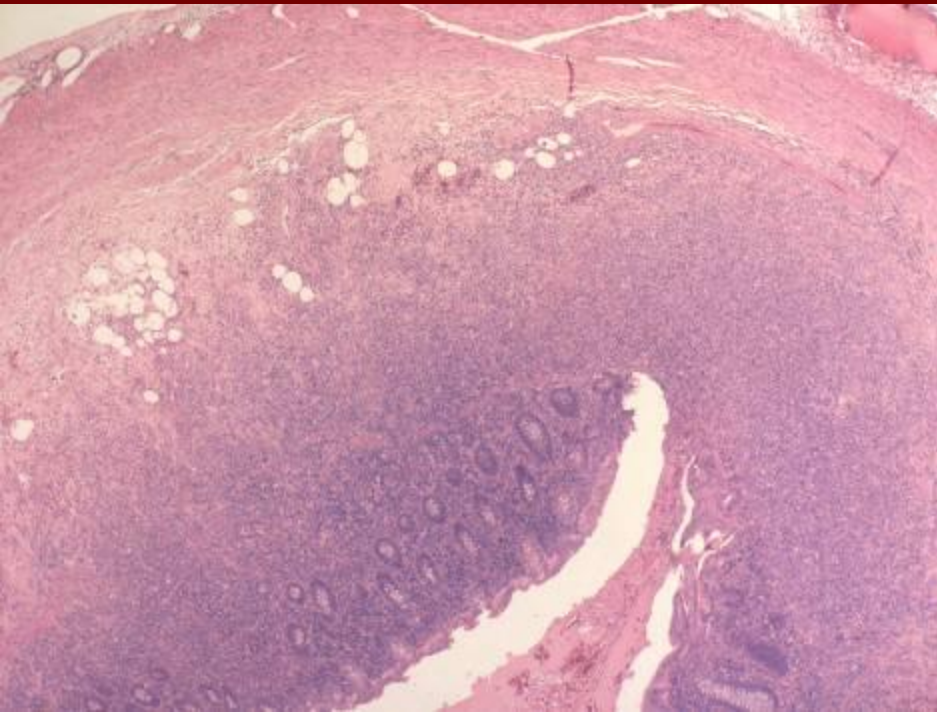


Extranodal NK/T-cell Lymphoma, Nasal Type



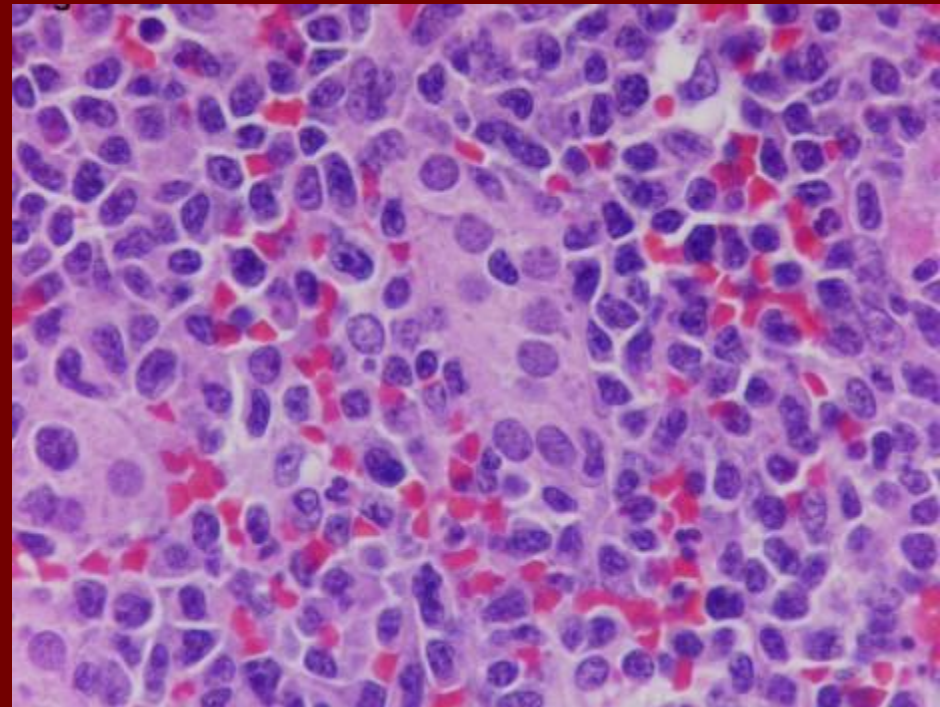
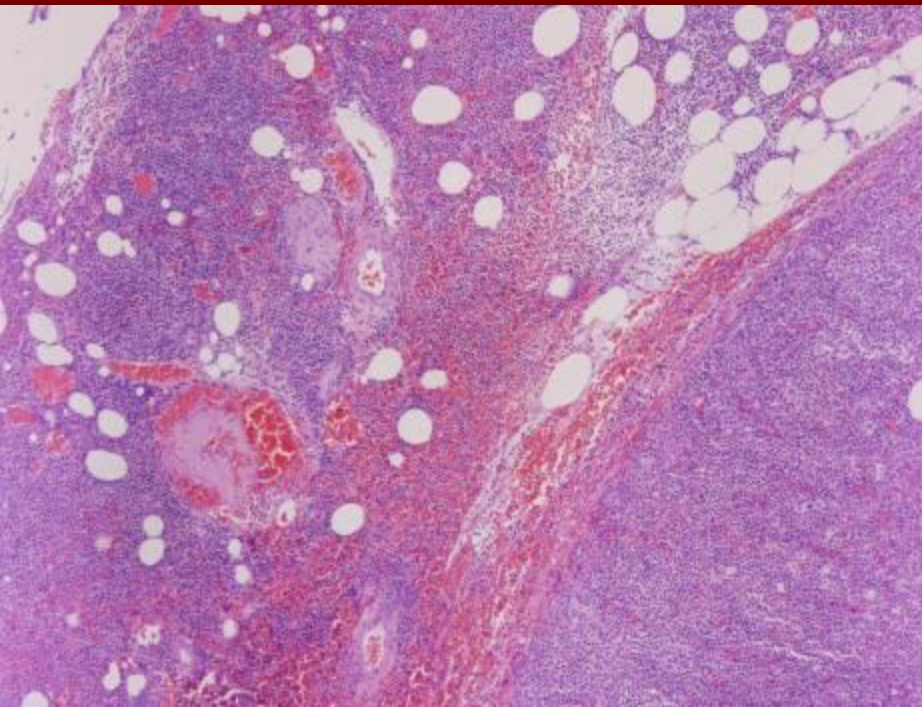
Extranodal NK/T-cell Lymphoma, Nasal Type

Bowel



Extranodal NK/T-cell Lymphoma, Nasal Type

Adrenal



Extranodal NK/T-cell Lymphoma, Nasal Type

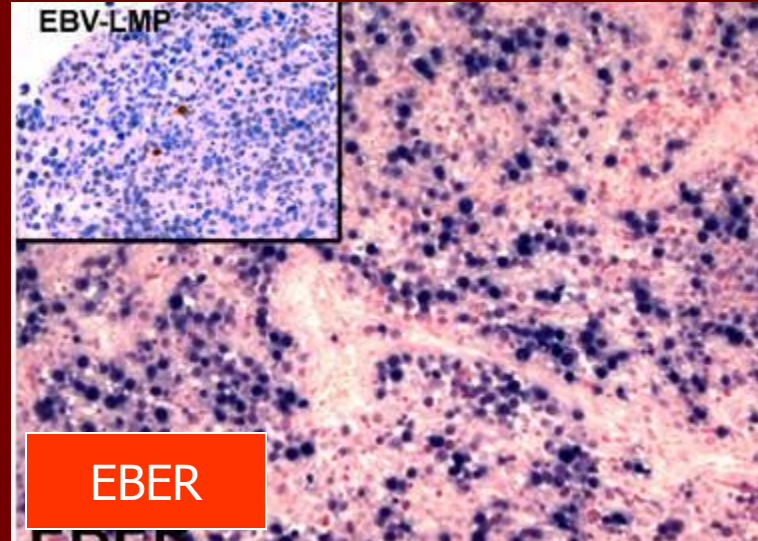
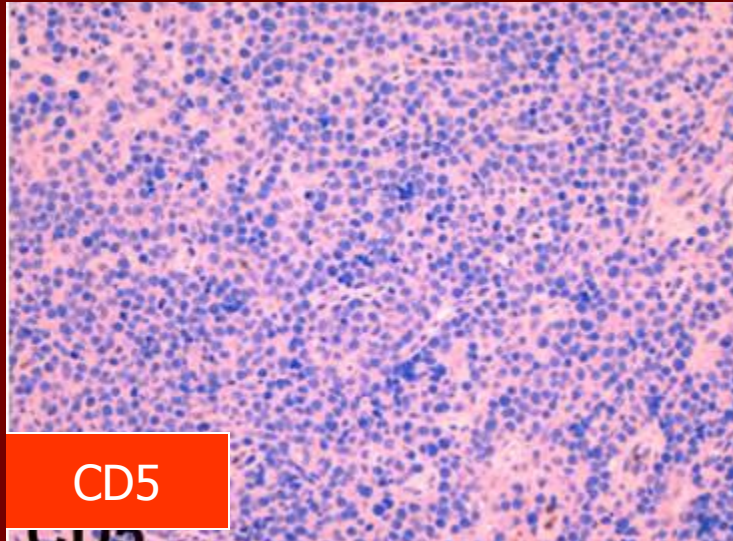
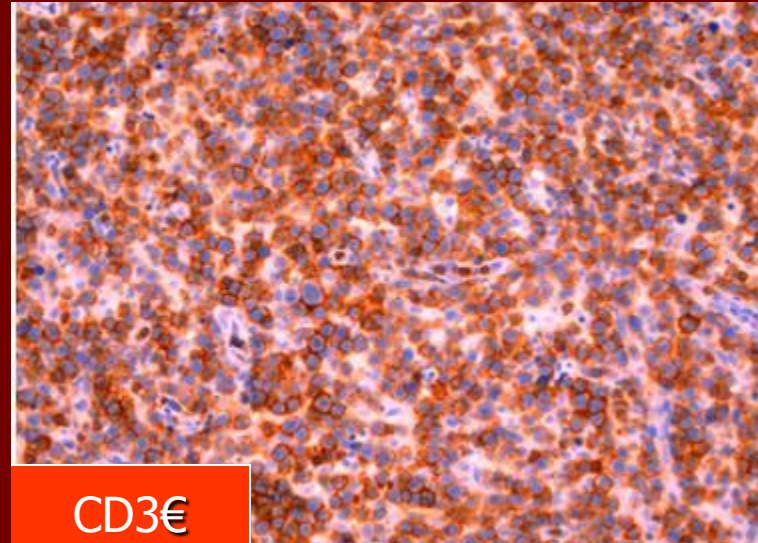
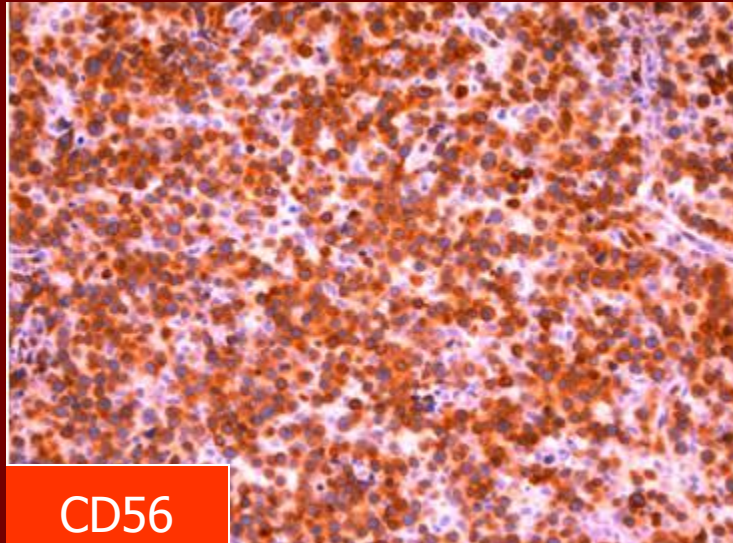
■ Immunophenotype

- CD2+, CD56+, sCD3-, cCD3ε+
- Positive for cytotoxic proteins
- Positive for EBV
- Other T-cell and NK-cell antigens negative

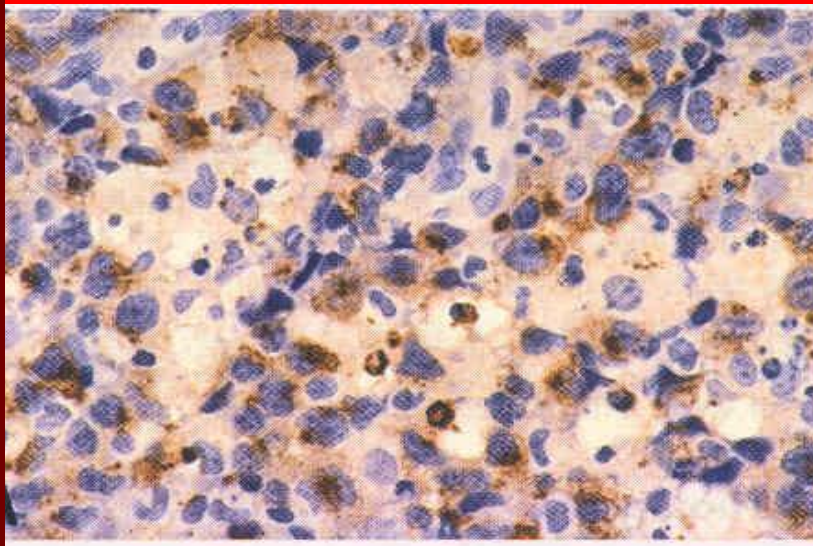
■ Genetics

- Usually TCR and Ig genes are germline

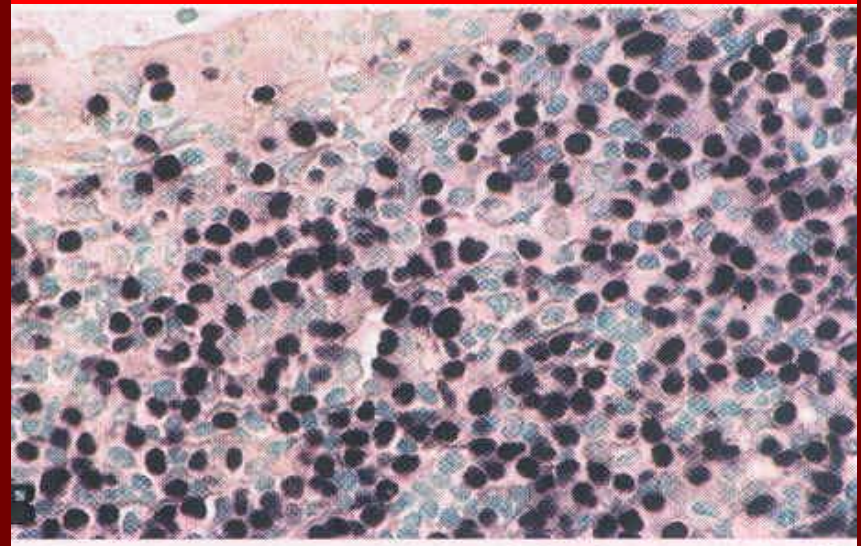
Extranodal NK/T-cell Lymphoma, Nasal Type



- Most cases also show cytotoxic granule associated proteins (granzyme B) and EBER



Granzyme B



EBER

Cell of Origin

- Activated NK cells or (rarely) cytotoxic T cells

Prognosis

- Variable
- Some pts respond well to therapy and others die of disseminated disease despite aggressive therapy