

**Subcutaneous
Panniculitis-Like
T-Cell Lymphoma**

Definition

- Cytotoxic T-cells of varying sizes
- Involving subcutaneous tissue with necrosis and karyorrhexis

Synonyms

- Lukes and Collins: not listed
- Kiel: not listed (pleomorphic T-cell lymphoma; small, medium sized, mixed or large cell; immunoblastic T-cell lymphoma)
- Working formulation: diffuse small cleaved cell; diffuse mixed small and large cell, diffuse large cell; immunoblastic

Synonyms

- REAL: Subcutaneous panniculitic T-cell lymphoma

Epidemiology

- Rare, <1% of non Hodgkin lymphomas
- M=F
- Broad age range, mostly young adults

Sites of Involvement

- Multiple subcutaneous nodules (0.5 to several cm)
- Extremities and trunk
- Other sites usually not involved (except in advanced disease)

Systemic Symptoms

- Variable
- Hemophagocytic syndrome with pancytopenia, fever, and hepatosplenomegaly in some (in aggressive fulminant cases)
- Lymph nodes rarely involved (usually in aggressive cases)

Etiology

- Most cases sporadic
- EBV negative
- Immunosuppression may be a factor in some ($\gamma\delta$ phenotype in these patients)

Precursor Lesions

- Probably no true precursor lesions
- Early biopsies may appear benign

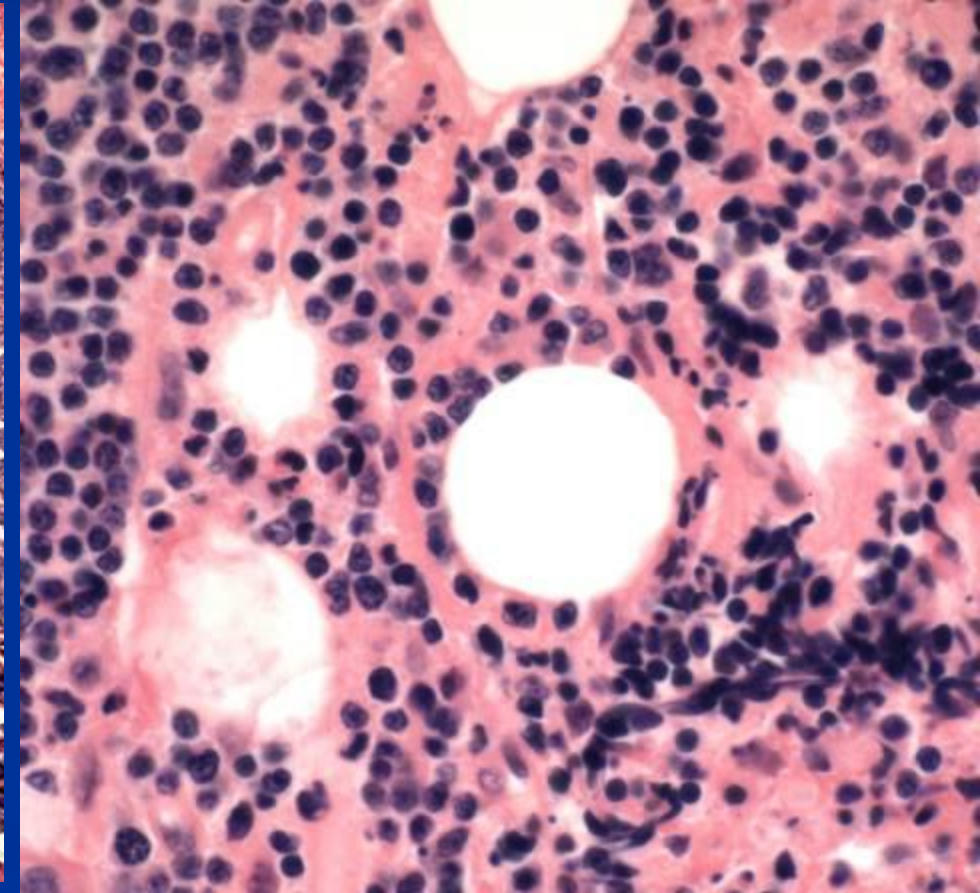
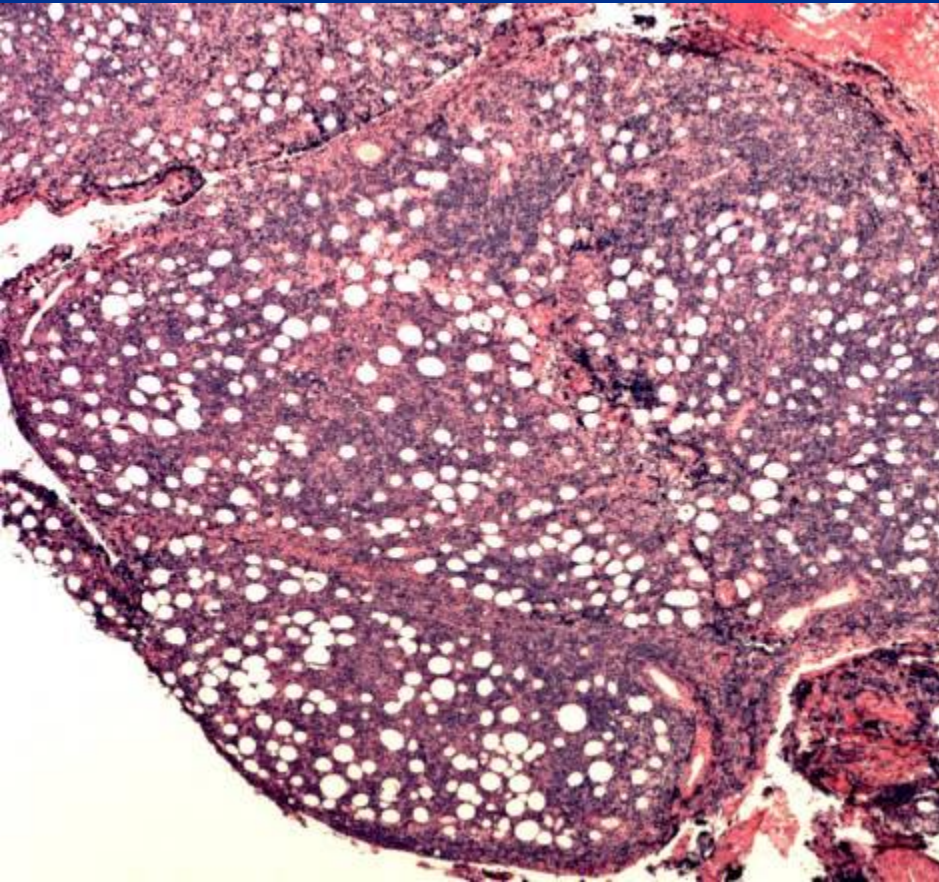
Histopathology

- Diffuse infiltrate in subcutis
- Septa involved (unlike in benign panniculitis)
- Dermis and epidermis usually spared (but often involved in $\gamma\delta$ cases)
- Lace-like pattern of rimming of fat by neoplastic cells

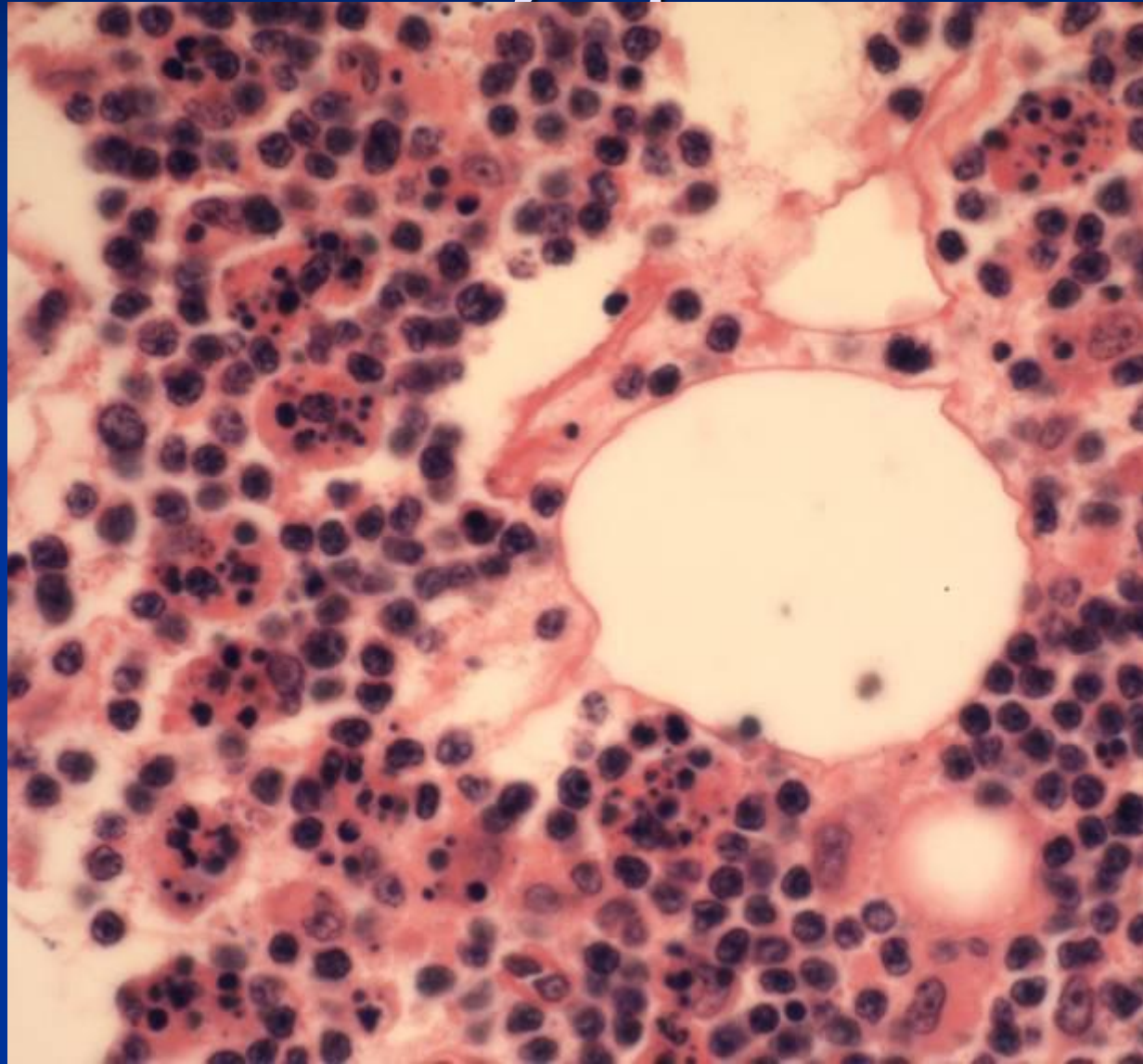
Histopathology

- Vacuolated histiocytes
- Necrosis
- Karyorrhexis (probably represents apoptosis)

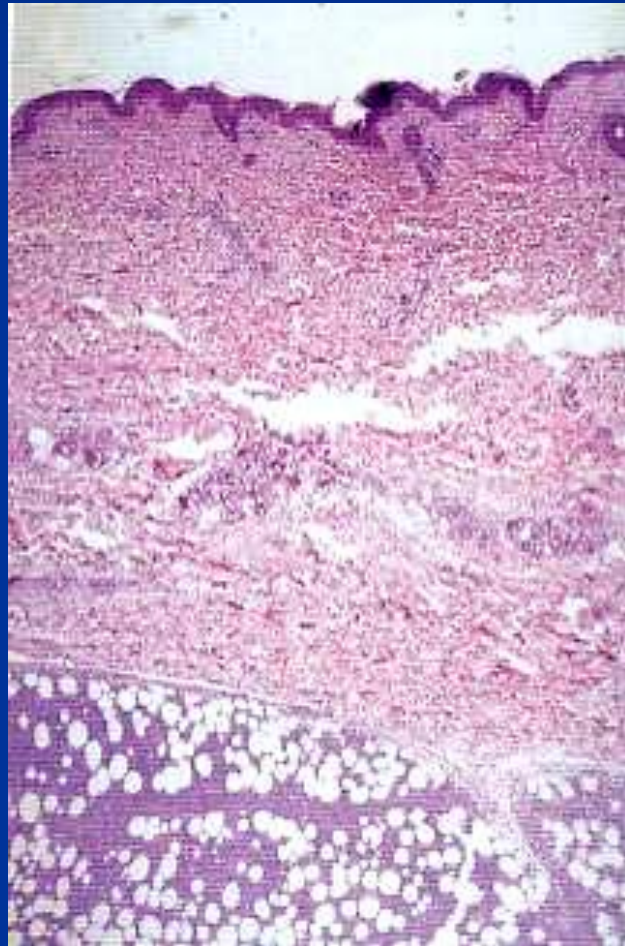
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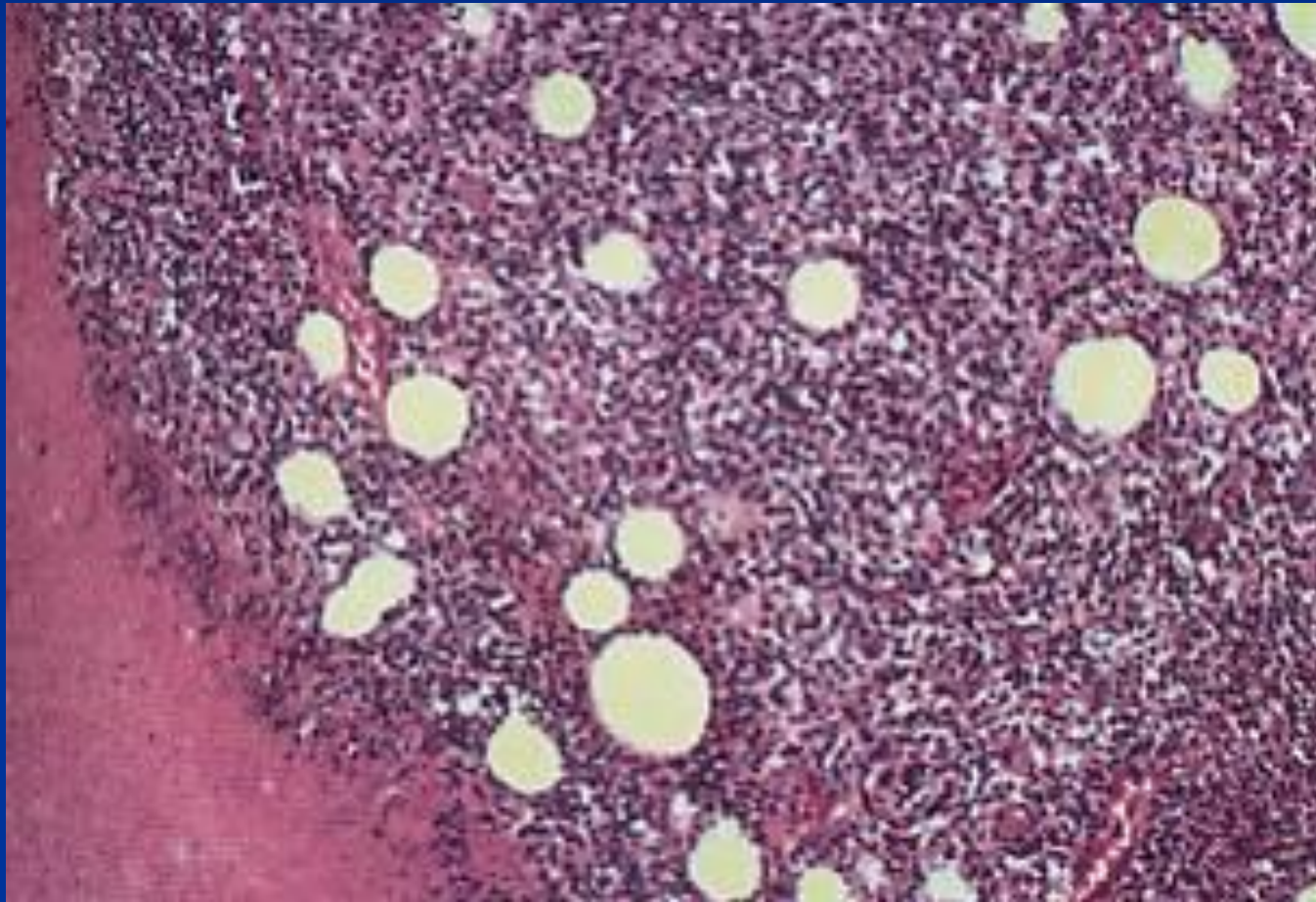
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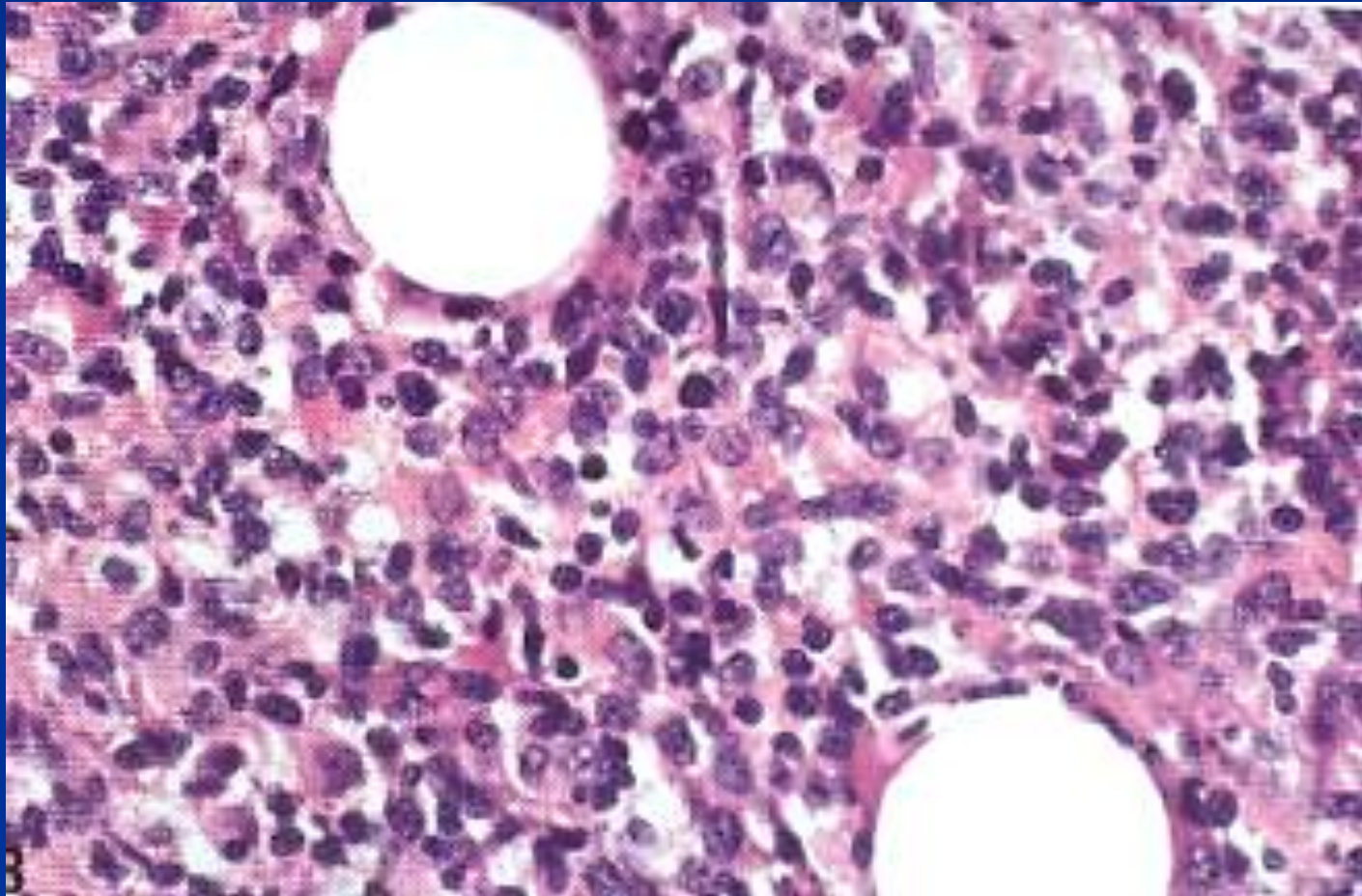
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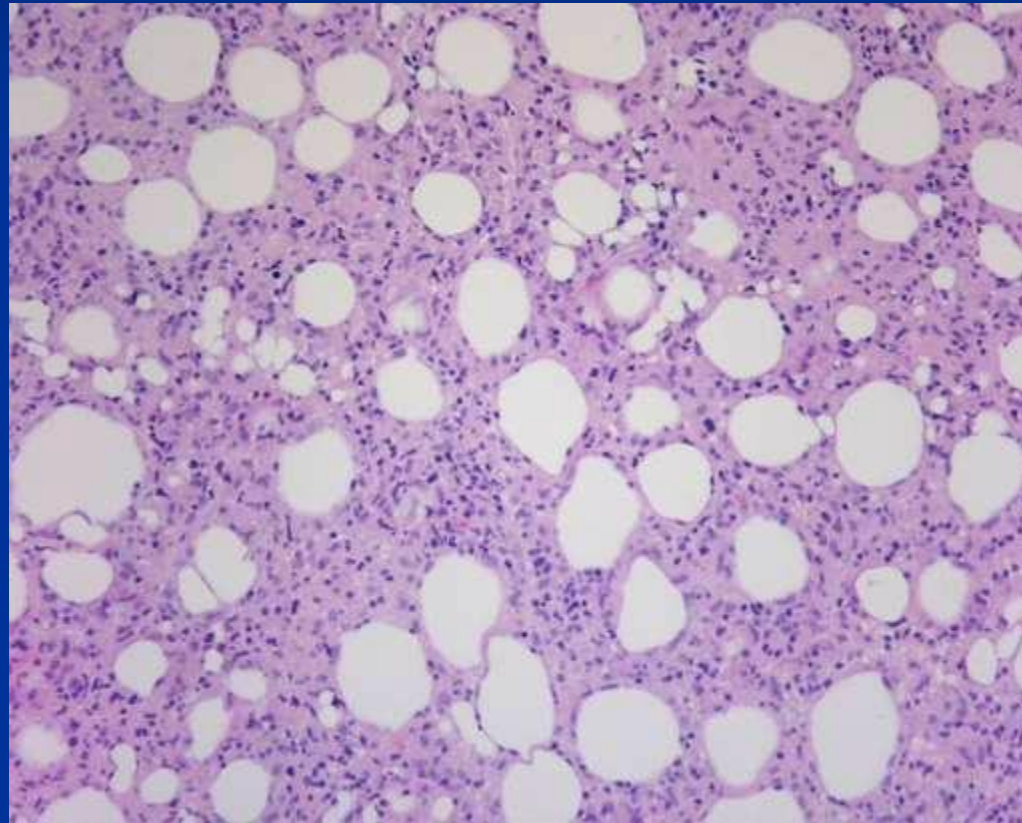
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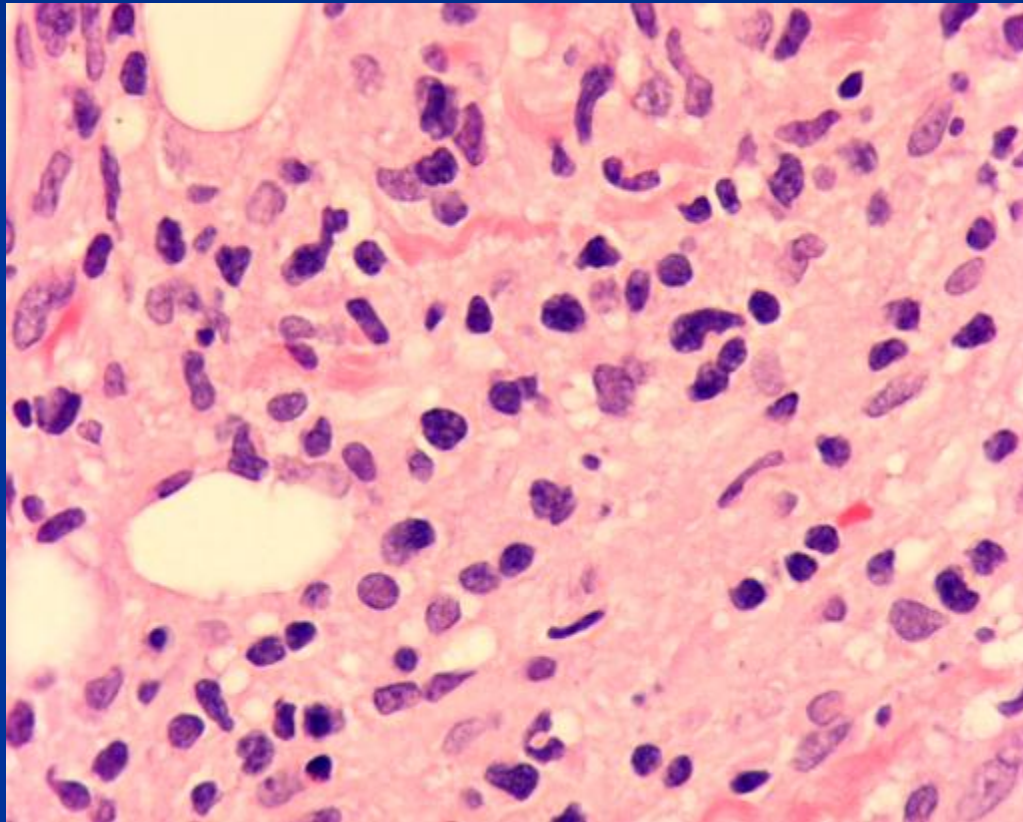
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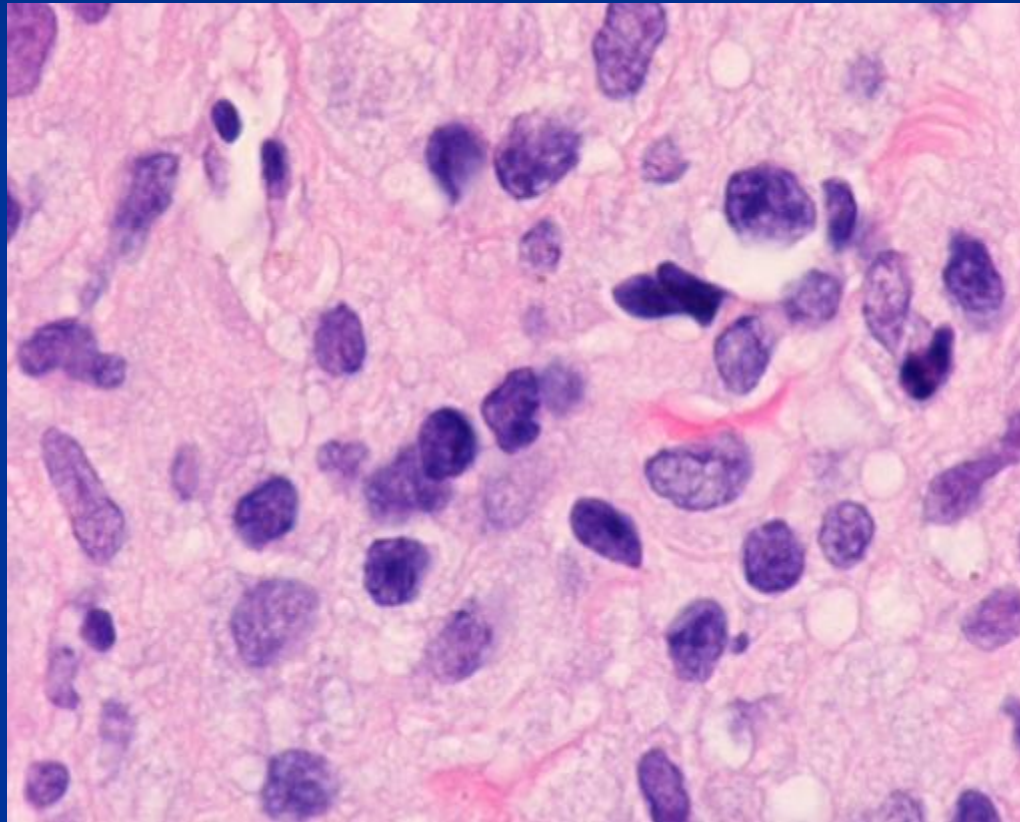
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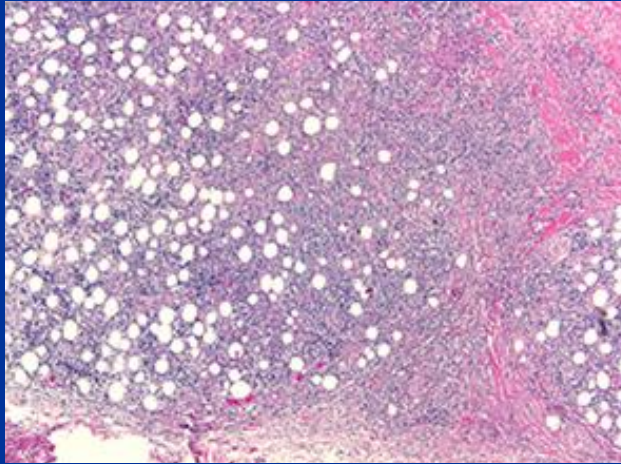
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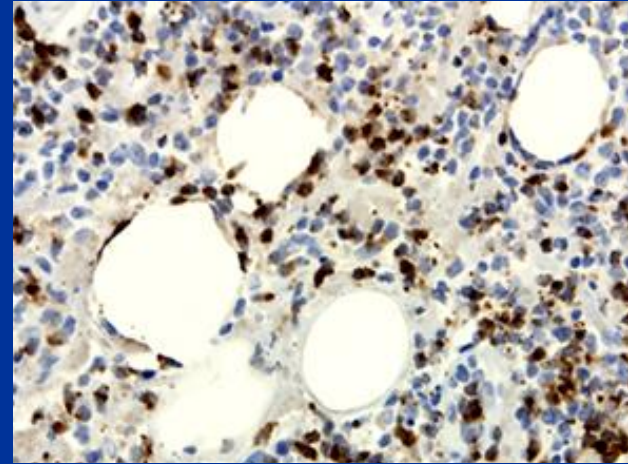
Immunophenotype

- Mature T-cell
- Usually CD8+
- Positive for cytotoxic molecules (granzyme B, perforin, and TIA-1)
- $\alpha\beta$ cases 75%
- $\gamma\delta$ cases 25% (often double negative for CD4 and CD8, and positive for CD56)

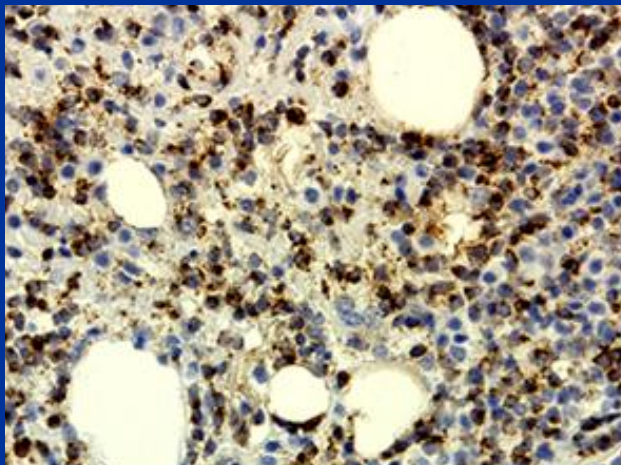
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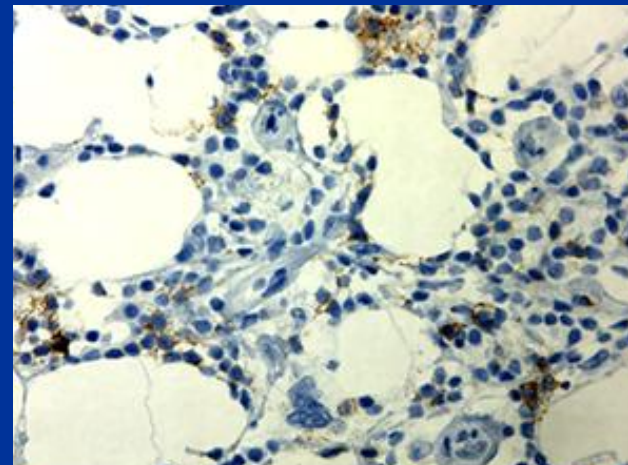
H&E: 40x



Granzyme

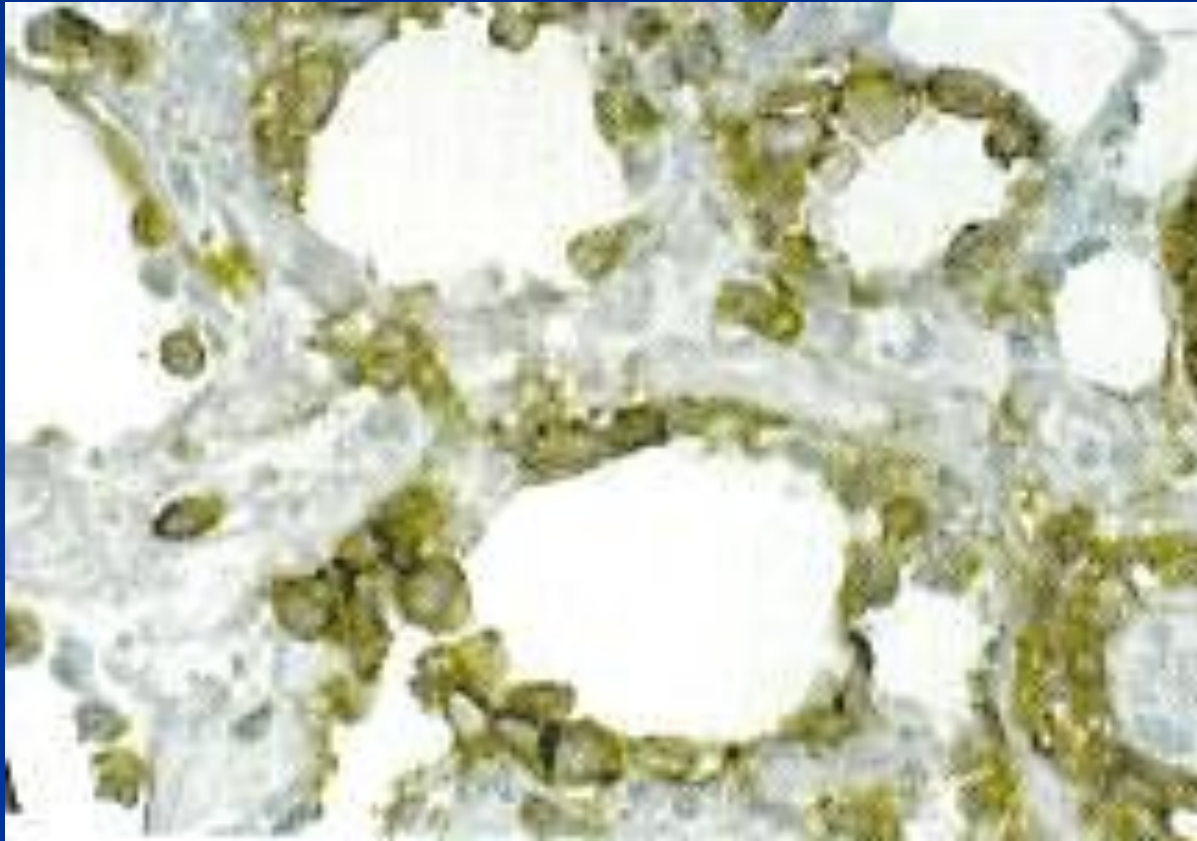


Perforin



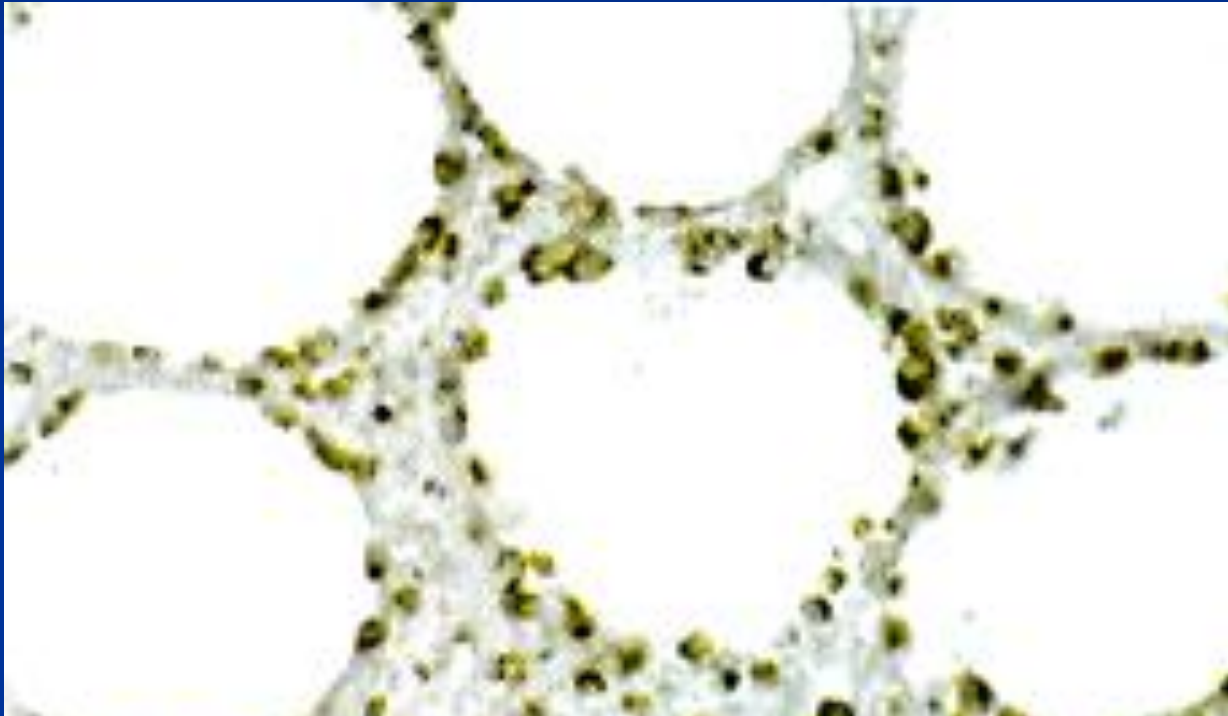
CD56

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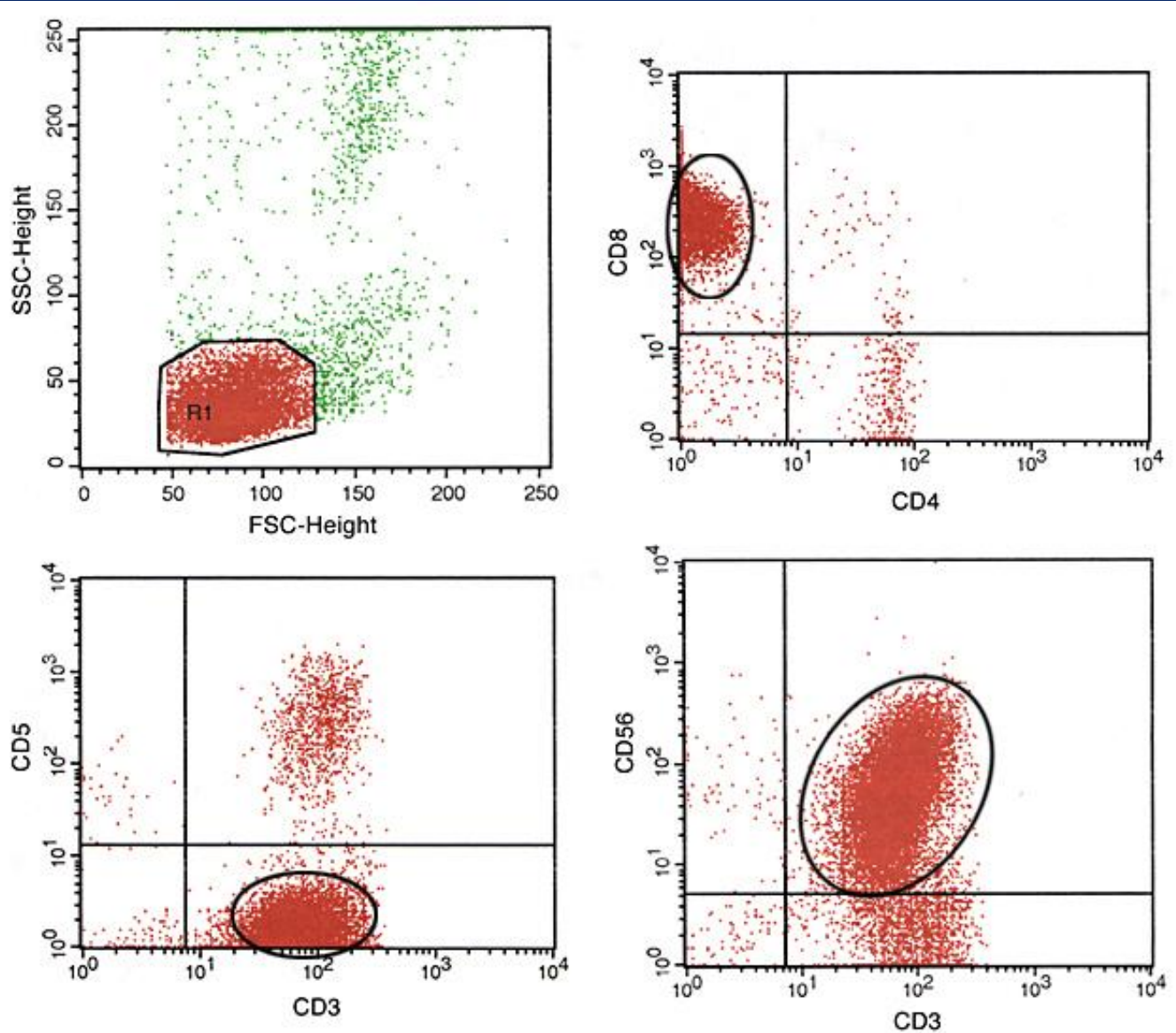
CD8

Subcutaneous Panniculitis-Like T-Cell Lymphoma



TIA-1

Subcutaneous Panniculitis-like T-cell Lymphoma: Flow Cytometry



Genetics

- Rearrangement for T-cell receptor genes
- Epstein Barr virus negative
- No other specific cytogenetic features

Differential Diagnosis

- Benign panniculitis
- Mycosis fungoides
- Extranodal T/NK-cell lymphoma, extranasal type
- Blastic NK-cell lymphoma
- Primary cutaneous anaplastic large cell lymphoma (ALCL)

Prognosis

- Aggressive disease with dissemination to lymph nodes and other organs in late course
- Hematophagocytic syndrome and lymph node disease precipitates a fulminant course
- Patients may respond to aggressive therapy