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 (γδ 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 γδ cases)
- Lace-like pattern of rimming of fat by neoplastic cells
Histopathology

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

- Mature T-cell
- Usually CD8+
- Positive for cytotoxic molecules (granzyme B, perforin, and TIA-1)
- αβ cases 75%
- γδ cases 25% (often double negative for CD4 and CD8, and positive for CD56)
Subcutaneous Panniculitis-like T-cell Lymphoma

H&E: 40x

Granzyeme

Perforin

CD56
Subcutaneous Panniculitis-Like T-Cell Lymphoma

CD8
Subcutaneous Panniculitis-Like T-Cell Lymphoma
Subcutaneous Panniculitis-like T-cell Lymphoma: Flow Cytometry
Genetics

- Rearrangement for T-cell receptor genes
- Ebstein 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