What we see

- Dense deep lobular atypical lymphocytic infiltrate involving subcutaneous tissue
- Rimming of fat cells by atypical lymphocytes
- Immunohistochemistry and flow cytometry was performed
Additional Studies

- Flow cytometry showed a dominant CD8 positive, TCR alpha/beta positive T cell population
  - Partial expression of CD56 and CD57
  - No aberrant loss of pan T cell markers (CD2, CD3, CD5, CD7)
Subcutaneous panniculitis like T cell lymphoma

(The likely trigger for her HLH)
Subcutaneous Panniculitis-like T cell Lymphoma

- Rare, <1% of NHL
- More common in women
- 20% of patients <20 yrs old, median 35 yrs
- 20% may have associated autoimmune disease (SLE most common)
  - Biggest differential dx is lupus profundus panniculitis
- CD8+ cytotoxic α/β T cells
  - γ/δ T cells are considered separately as primary cutaneous γ/δ T cell lymphoma and have a more aggressive course & worse prognosis
Multiple subcutaneous nodules
- Extremities, trunk, thighs most common
- Range from 0.5cm to several cm in diameter
- Kids may present with head and neck disease

Systemic symptoms in up to 50%
- Cytopenias, fever, anorexia
- Elevated LFT’s
- Nearly 50% with HLH

Lymphadenopathy, hepatosplenomegaly usu absent unless HLH also present
Infiltrate involves fat lobules, sparing septa

- “ringing” around adipocytes

Epidermis, dermis less involved

- May infiltrate eccrine coil
- Few atypical lymphocytes in epidermis
- Grenz zone present

Variable cell size (small, medium, large), but usually monotonous & consistent in the tumor

Irregular hyperchromatic nuclei, rim of pale cytoplasm
- Reactive histiocytes usu present – beware thinking it is just infectious!
- Histiocytes phagocytose debris
  - May see erythropagocytosis
- Plasma cells usu absent (unlike SLE)
- Karyorrhexis, necrosis, fat necrosis often prominent
- Vasculopathic changes vary
  - Thrombogenic vasculopathy, lymphocytic angioinvasion, mural & luminal fibrin deposition
Differential Diagnosis

- Many features overlap with lupus profundus
- May be very challenging differential
- SCPTCL generally lacks reactive germinal centers, plasma cells
- Dermal mucin can be seen in both
- Lymphocyte atypia not as severe in lupus
- No loss of pan T cell markers
SCPTCL

- Mature T cells – pan T markers
  - May lose CD5 and CD7, have decreased CD3 expression
- Alpha beta TCR+
  - βF1+ by IHC
  - Important to establish and distinguish from γ/δ T cell lymphoma which is more aggressive
- CD8+, cytotoxic markers+ (TIA1, granzyme B, perforin)
- Negative for CD56
- Proliferation index (Ki-67) intermediate to high
- CCL5+ (its ligand is CCR5, which is highly expressed on adipocytes)
Variable clinical course
- Some do well and spontaneously resolve, others more aggressive
- May wax and wane over years
- Local recurrences may occur, remain confined to subcutis

Relatively good prognosis
- Median 5 year survival 80%

Worse prognosis if have HLH
- Mortality often due to sepsis due to neutropenia
Follow up

- Patient treated with Decadron, then oral methotrexate and 6-MP with good response and resolution of lesions

- Currently in remission from both HLH and SCPTCL
Case #16

17 year old boy with history of tuberculosis, now with neck mass unresponsive to antibiotics. An excision of the mass is performed.
### Viable cells

**CD33 APC-A**

**CD19 PE-A**

-10 2 10 0 10 2 10 3 10 4 10 5

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<th>0x0 4.52% 4.42%</th>
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<tr>
<td><strong>Viable</strong></td>
<td><strong>CD33</strong></td>
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<tr>
<td><strong>cells</strong></td>
<td><strong>CD19</strong></td>
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**Lymphocytes = 84.05%**

### Forward Scatter vs Side Scatter

- **Lymphocytes**

### CD45

**Side Scatter**

- **CD45 V500-A**

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### CD33 vs CD19

**Forward Scatter**

- **CD33**
- **CD19**

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### CD3 vs CD19

**Side Scatter**

- **CD3**
- **CD19**

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<tr>
<td><strong>CD3</strong></td>
<td><strong>CD19</strong></td>
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### Lymphocytes

- **CD3 APC-H7-A**:
  - Side Scatter: 99.74% 0.00%
  - Myeloperoxidase: 0.08% 0.00%
  - TdT FITC-A: 0.09% 0.17%
  - Cytoplasmic markers: 17.79% 81.95%

- **MPO PE-A**:
  - Side Scatter: 99.74% 0.00%
  - Myeloperoxidase: 0.08% 0.00%
  - TdT FITC-A: 0.09% 0.17%
  - Cytoplasmic markers: 99.74% 0.26%