### Primary cutaneous T-cell lymphomas

**By Georgeanne Cornell, DO, Michael Visconti, DO, and Stephen Olsen, MD**

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<th>Clinical features</th>
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<th>Histopathology</th>
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<th>Prognosis</th>
<th>Management</th>
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<tr>
<td>Lymphomatoid papulosis (LyP)</td>
<td>Recurring papules/ nodules, clustering</td>
<td>Regress/ recur (frequently) MF/ALCL/ Hodgkin lymphoma in 20%</td>
<td>Five types: A. Large CD30+ lymphocytes with mixed inflammatory cells B. CD4+ epidermotropism C. Mimics pcALCL D. CD8+ epidermotropism E. Angioinvasive</td>
<td>CD30+ phenotypes vary Clonal T-cell gene rearrangement in 50%</td>
<td>PLEVA: Presence of CD30+ blast cells MF with large cell transformation: History of preceding patches/plaques ALCL: No history of recurring lesions</td>
<td>Limited or asymptomatic No therapy/topical steroids Symptomatic, progressive, widespread Phototherapy and/or low-dose methotrexate (MTX)</td>
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<td>Mycosis Fungoides (MF)</td>
<td>Patch/plaque stage: scaly red-brown patches and plaques in non-sun- exposed distribution Tumor stage: nodules with frequent ulceration in a background of patches/ plaques</td>
<td>Benign course in limited disease</td>
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<td>Variants: folliculotropic MF, pagetoid reticulosis, granulomatous slack skin</td>
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<tr>
<td>Primary cutaneous anaplastic large cell lymphoma (pcALCL)</td>
<td>Solitary&gt; multiple Grouping of firm nodules</td>
<td>Unlike LyP, lesions do not rapidly come and go</td>
<td>Sheets of large, atypical CD30+ lymphocytes</td>
<td>CD30+ CD4+/CD8-</td>
<td>ATLL: Histologically identical Sézary syndrome: Peripheral blood involvement Pagetoid reticulosis: CD8+ phenotype More extensive epidermotropism LyP: Self-healing Type B histologically indistinguishable Type D: CD8+, CD30+phenotype AECTCL: No cernifrom nuclei CD8+ phenotype</td>
<td>Excellent</td>
</tr>
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<td>Primary cutaneous CD8+ small/ medium T-cell lymphoproliferative disorder (PCSM-TCLPD)</td>
<td>Solitary plaque or nodule</td>
<td>May have aggressive course (need to rule out primary cutaneous peripheral T-cell lymphoma, not otherwise specified)</td>
<td>Dense nodular to diffuse small/ medium lymphocytic infiltrate Minimal to no epidermotropism</td>
<td>CD4+/CD8-/CD30- (MF-like)</td>
<td>Systemic ALCL: EMA+ (negative in pcALCL) LyP: Recurring lesions MF with large cell transformation: No history of preceding patches/ plaques</td>
<td>Solitary or localized disease Surgical excision, radiotherapy Limited or asymptomatic disease Radiotherapy, low-dose MTX, brentuximab</td>
</tr>
<tr>
<td>Primary cutaneous acral CD8+ T-cell lymphoma (ATCL)</td>
<td>Solitary papule/ nodule EAR = most common site</td>
<td>Benign course</td>
<td>Dense nodular to diffuse infiltrate • Grenz zone • No mitoses, necrosis, ulceration, or angiocentricity</td>
<td>CD3+CD8+/CD4-</td>
<td>Other cytotoxic CD8+ cutaneous T-cell lymphomas</td>
<td>Localized disease: Excision Topical/intralental steroids Radiotherapy</td>
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</table>
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| Subcutaneous panniculitis like T-cell lymphoma (SPTCL) | Subcutaneous nodules or deep plaques | Hemorrhagic syndrome (HPS) = worse prognosis | Lobular panniculitis of "fat rimming" neoplastic lymphocytes  
"Bean bag cells" (cytophagocytosis) | CD4-/CD8+/CD56-/TIA-1+/granzyme/+ perforin+/βF1+/αβ phenotype  
Lupus profundus: Plasma cells, interface change, nodular lymphoid aggregates | Good | w/o HPS: Oral corticosteroids w/ or w/o immunosuppressant (cyclosporine A or low-dose MTX)  
w/ HPS: High-dose corticosteroids w/ cyclosporine A  
High-dose chemotherapy with HSCT |
| Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma (PC-AECTCL) | Eruptive, ulcerated tumors | Visceral involvement | Malignant infiltrate with epidermotropism and angiodestruction | CD4-/CD8+/CD56+/TIA-1+/granzyeme+/perforin+  
Lupus profundus: MF, pagetoid reticulosis, LyP (D): Histologically indistinguishable (all CD8+)  
SPTCL: Does NOT have lichenoid interface | Lupus profundus: β8-  
Radiotherapy  
Multisagent chemotherapy w/ or w/o HSCT |
| Primary cutaneous γδ T-cell lymphoma (PCGD-TCL) | Multiple eroded nodules and plaques | HPS  
Viscerat involvement | CD4/CD8 (“double negative”)  
Expression of γδ receptor | Lupus profundus: γδ+  
Antivirals  
Multisagent chemotherapy w/ or w/o HSCT  
Localized: Radiotherapy  
Systemic: Chemotherapy |
| Adult T-cell leukemia/lymphoma (ATLL) | Small, confluent, violaceous papules, or firm/brown nodules | Hypercalcemia HTLV-1+ | Florot or clove leaf malignant T-cells  
Lack of β8 expression  
CD2+/CD3ε+/CD56+/TIA-1+/granzyme+/- perforin+/EBV-  
Lupus profundus: Malignant cells  
MTX, bexarotene, interferon Mogalizumab (if failed at least one systemic)  
Extracoporal photopheresis |
| Extranodal NK/T cell lymphoma (ENKTL) | Ulcerating plaques or tumors; May extend into the nose, sinuses, palate | HPS EBV+ | Dense infiltrate with extensive necrosis  
Fat rimming  
Angiocentricity and/or angiodestruction | Lupus profundus: Other causes of erythroderma: Will not have identical T-cell clone in the skin and blood |
| Sézary syndrome | Erythroderma, lymphadenopathy, Sézary cells | Circulating CD4+ neoplastic cells >1000 cells/µl  
Non-specific or resemble MF | CD3+/CD4+/CD8-/PD-1+  
MTX, bexarotene, interferon Mogalizumab (if failed at least one systemic)  
Extracoporal photopheresis |

### References: