44 year-old male

Clinical: Erythematous scaling papules symmetrically on the forearms, abdomen and lower back

DDX: Allergic contact dermatitis vs drug rash vs irritant dermatitis vs scabies vs pigmented purpuric dermatitis

Follicular Hyperkeratosis
- Discoid lupus
- Pityriasis rubra pilaris
- Lichen sclerosus
- Keratosis pilaris
Vacuolar Interface Dermatitis

- Lymph in every hole
- More holes than lymphs
- Mycosis fungoides
- LESA
- BL
- Sebaceous
- Occasional, fixed drug, lymphomatoid drug, lichen sclerosus
- Drug eruption
- Erythema multiforme
- Necrotic keratinocytes
- Drug eruption
- Fixed drug eruption
Skin, left volar forearm: Interface dermatitis with follicular plugging (see comment)

Comment: The microscopic differential diagnosis includes drug or viral eruption, connective tissue disease (including Wong type dermatomyositis), pigmented purpuric dermatoses, and pityriasis lichenoides.

Wong-type dermatomyositis
- Variant of DM with erythematous hyperkeratotic follicular papules similar to PRP
- Reported in children and adults
- Myositis occurs at similar rate of classic DM
- In Wong’s original report 52% developed malignancy
- Subsequent reports have shown no increased rate of malignancy
- Palmoplantar keratoderma is sometimes present

Additional History
- ROS positive for weakness and dysphagia
- 2 weeks prior PCP noted enlarge lymph nodes in the neck
- Metastatic tonsillar squamous cell carcinoma

Matsumoto A, Wang R, Carlson JA.

**Take Home**

Columnar dyskeratosis—A clue to Wong-type dermatomyositis?

Take Home Columnar dyskeratosis/pseudo-coronoid lamella Vacuolar Interface = Wong Type DM

31 year old G2P1 female at 8 weeks gestation
Subcutaneous Panniculitis-like T-cell Lymphoma
- B-F1 (αβ) positive
- GM1 (γδ) negative

**T-CELL CLONALITY BY PCR**

**Source:** SKIN, LEFT LATERAL LEG
**Results:** POSITIVE
**Comment:** Detection of T-cell receptor gamma chain gene rearrangement consistent with the presence of a clonal cell population. The clone is detectable with primers specific for the Vδ2-δ8 region.

**Primer Set:** Vγ2-M, Vγ2-M, Vγ3, Vγ2, Vγ1.1, Vγ1.2, and Vδ2-R
**Method:** PCR amplification with Differential Fluorescence Detection

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All was well, until……

After delivery developed rapid progression of new lesions despite treatment with increased prednisone, methotrexate and romidepsin

Fever
Leukopenia, anemia, hyperferritinemia, and increased LFTs
Ultrasound suggested cholecystitis
Admitted
Fatal HPS due to SPTCL
**Hemophagocytic Syndrome**
- Life-threatening syndrome of excessive immune activation
  - Histiocytes activated and secrete excess cytokines
  - Cytotoxic T lymphocytes and NK cells are unable to eliminate activated macrophages

**Familial Hemophagocytic Lymphohistiocytosis**
- Mutations in genes involved in perforin-dependent cytotoxicity
  - PRF1, UNC13D, STX11, STXB2
  - Most frequently presents in first year of life

**Reactive Hemophagocytic Syndrome**
- Viral illness
- Autoimmune disease (Macrophage activation syndrome)
- Lymphoma
- Immunodeficiency

**Hemophagocytosis**
- Presence of fragments or intact RBC, platelets, or WBCs within the cytoplasm of macrophages
What’s The Skin Got To Do With It?

- Cutaneous findings in 10 to 65%
- Manifestation of the trigger or underlying disease
  - Viral exanthem
  - Lupus, Dermatomyositis, Still’s Disease
  - Cutaneous lymphoma (SPLTCL)
- Manifestation of the associated liver dysfunction and coagulation problems

<table>
<thead>
<tr>
<th>Age</th>
<th>Underlying Disease</th>
<th>Cutaneous Findings</th>
<th>Dermatology Consult</th>
<th>Outcome</th>
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<tr>
<td>33</td>
<td>SPLTCL</td>
<td>SPLTCL</td>
<td>+</td>
<td>DOD</td>
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<tr>
<td>70</td>
<td>Adult Onset Still Disease</td>
<td>AOSD</td>
<td>+</td>
<td>DOD</td>
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<td>?</td>
<td>1st visit: none</td>
<td>2nd visit: HSV</td>
<td>+</td>
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<tr>
<td>25</td>
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<td></td>
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<td>Juvenile idiopathic arthritis</td>
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<td>26</td>
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<td>-</td>
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</tr>
<tr>
<td>56</td>
<td>HIV/DLBCL/Aspergillus pneumonia</td>
<td>None</td>
<td>-</td>
<td>DOD</td>
</tr>
</tbody>
</table>

70 yo male with intermittent fevers, joint pain, and persistent plaques

Eventually developed acute mental status changes and massive GI bleed resulting in his death
Adult Onset Stills Disease

- Spiking fever >=39 C
- Arthralgia/arthritis
- Evanescent rash (77-100%)-asymptomatic salmon pink macules/papules with fever on trunk, extremities and areas of pressure
- Persistent papules and plaques
- Increased WBC with 80% neutrophils
- 12-15% RHS

Take Home

- Histologic features of SPTCL and lupus panniculitis show marked overlap and may require prolonged follow-up
- Atypical CD8+/Ki-67+ lymphocytes rimming adipocytes suggest SPTCL

Take Home

- Non-specific morbilliform, purpuric or trigger-specific eruptions with fever, hepatosplenomegaly, neurologic symptoms, cytopenias, increased ferritin levels, liver function impairment or coagulation problems—think about Hemophagocytic Syndrome
- Persistent plaques of AOSD have apoptotic keratinocytes

32 year-old female

DDx: ? Scarring alopecia

DDx: ? Scarring alopecia

Tyler Technique

3/4/2019
32 year-old female

**Clinical:** Erythematous scalp with hair loss, scaling. Fungal culture was negative. Has Crohn’s. Patient on Remicaide.

**DDx:** ? Scarring alopecia.

- 5 months of scaling and hair loss
- Failed dandruff shampoo, econazole, and betamethasone

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**TNF-Inhibitor Induced Alopecia**

- 3/2016: started remicade
- 10/2017: c/o scaling and hair loss

Take Home

- TNF Inhibitor delayed reactions
- Psoriasiform dermatitis
  - Often with interface, spongiosis and mixed infiltrate
- Alopecia
  - AA histology with psoriasis, mixed infiltrate and atrophic sebaceous glands