Patient 1

Chief Complaint: Rash and Fever

Pediatric Annals; January 2012 - Volume 41 · Issue 1: e1-e5
What is the most appropriate treatment?

A. Slow prednisone taper
B. 14 day course of cephalexin
C. Acyclovir 400mg PO 5 times/day x 10 days
D. Cool compresses
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B. 14 day course of cephalexin
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Eczema Herpeticum

• Herpetic superinfection of a preexisting skin disease
  – Atopic dermatitis
  – Darier’s disease
  – Autoimmune blistering disorders
• Autoinnoculation or infected contact
• Life threatening
Eczema Herpeticum

- Securing diagnosis
  - Tzanck prep
  - Direct fluorescent antibody test (DFA)
  - Viral culture
Eczema Herpeticum

- Predisposing factors
  - Impaired skin barrier
  - Immunosuppression
Eczema Herpeticum Management

- Acyclovir
- Possible ophthalmology consultation
- Pain management
- Treat secondary infection
What is the most appropriate treatment?

A. Slow prednisone taper
B. 14 day course of cephalexin
C. Acyclovir 400mg PO 5 times/day x 10 days
D. Cool compresses
Patient 2

- 26yo woman
- 3-4 month history of bloating, abdominal pain and diarrhea
- Itchy, burning rash
Quiz

All of the following are important parts of this patients evaluation EXCEPT:
A. Skin biopsy
B. Wound culture
C. Thyroid studies
D. IgA endomysial antibody
Quiz

All of the following are important parts of this patient's evaluation EXCEPT:

A. Skin biopsy
B. Wound culture
C. Thyroid studies
D. IgA endomysial antibody
Dermatitis Herpetiformis

• Autoimmune blistering disorder associated with gluten sensitive enteropathy
• Northern European ancestry
  – HLA DQ2
• “virtually always” associated with celiac disease
• Up to 50% with thyroid disease
Dermatitis Herpetiformis

• Securing the diagnosis
  – Skin biopsy
    • H&E
    • Direct immunofluorescence
  IgA
  Neutrophils
Dermatitis Herpetiformis

- IgA anti-endomysial antibodies
- Tissue transglutaminase antibody - tTG (IgA)
- Deamidated gliadin peptide antibody - dGP (IgA and IgG)
- Gliadin assay (IgA and IgG)
Dermatitis Herpetiformis

- Management
  - Gluten free diet
  - Dapsone or sulfapyridine
Quiz

All of the following are important parts of this patient's evaluation EXCEPT:

A. Skin biopsy
B. Wound culture
C. Thyroid studies
D. IgA endomysial antibody
Patient 3

Patient 3

- Skin biopsy - subepidermal blister containing neutrophils and nuclear dust
- DIF - IgG, IgM and C3 at the basement membrane zone
- ANA 3+ positive
- Complement components
  - C3 48 (nl 80-200mg/dl)
  - C4 4 (nl 20-50mg/dl)
- Anti-dsDNA 89% (nl 0-25%)
- Anti-Sm 1:102,400
- Lymphopenia
Quiz

Besides the skin, which other organ system is most likely to be involved?

A. CNS
B. Hematologic
C. Musculoskeletal
D. Renal
Quiz

Besides the skin, which other organ system is most likely to be involved?
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Bullous Systemic Lupus Erythematosus

- Uncommon presentation of systemic lupus
- Bullous flares often parallel disease flares in other organ systems particularly renal
- Blisters not limited to sun exposed areas
## Bullous Systemic Lupus Erythematosus

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Definition</th>
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<tbody>
<tr>
<td>Malar rash</td>
<td>Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds</td>
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<td>Discoid rash</td>
<td>Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions</td>
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<td>Photosensitivity</td>
<td>Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation</td>
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<td>Oral ulcers</td>
<td>Oral or nasopharyngeal ulceration, usually painless, observed by a physician</td>
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<td><strong>Arthritis</strong></td>
<td>Nonerosive arthritis involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion</td>
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<td>Serositis</td>
<td>Pleuritis - convincing history of pleuritic pain or rub heard by a physician or evidence of pleural effusion OR</td>
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<td>Pericarditis - documented by EKG, rub or evidence of pericardial effusion</td>
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<td>Renal disorder</td>
<td>Persistent proteinuria greater than 0.5 grams per day or greater than 3+ if quantitation not performed OR</td>
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<td>Cellular casts - may be red cell, hemoglobin, granular, tubular, or mixed</td>
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<td><strong>Neurologic disorder</strong></td>
<td>Seizures OR psychosis - in the absence of offending drugs or known metabolic derangements (uremia, ketoacidosisis, or electrolyte imbalance)</td>
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<td>Hemolytic anemia - with reticulocytosis OR</td>
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<td><strong>Hematologic disorder</strong></td>
<td>Leukopenia - less than 4,000/mm^3 total on two or more occasions OR</td>
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<td></td>
<td>Lymphopenia - less than 1,500/mm^3 on two or more occasions OR</td>
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<tr>
<td></td>
<td>Thrombocytopenia - less than 100,000/mm^3 in the absence of offending drugs</td>
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<tr>
<td>Immunologic disorders</td>
<td>Positive antiphospholipid antibody OR</td>
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<td>Anti-DNA - antibody to native DNA in abnormal titer OR</td>
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<td></td>
<td>Anti-Sm - presence of antibody to Sm nuclear antigen OR</td>
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<td>False positive serologic test for syphilis known to be positive for at least six months and confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test</td>
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<tr>
<td>Antinuclear antibody</td>
<td>An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with &quot;drug-induced lupus&quot; syndrome</td>
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</table>
Bullous Systemic Lupus Erythematosus

- Management
  - Dapsone
  - Systemic corticosteroids
  - Pulsed cyclophosphamide

Lupus nephritis
Quiz

Besides the skin, which other organ system is most likely to be involved?

A. CNS
B. Hematologic
C. Musculoskeletal
D. Renal
Quiz

Which of these infectious diseases is most commonly associated with this disorder?

A. Hepatitis B
B. Hepatitis C
C. Syphilis
D. Varicella
Quiz

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Porphyria Cutanea Tarda

- Deficiency of heme synthetic enzyme uroporphyrinogen decarboxylase (UROD)
  - Acquired
    - Induced by estrogen or alcohol
    - Associated with Hepatitis C and HIV
  - Familial
    - Autosomal dominant with incomplete penetrance
- ↑↑ Copro- and uroporphyrins
Porphyria Cutanea Tarda

- Blisters on sun-exposed skin
- Skin fragility
- Milia
- Hypertrichosis

Tx—serial phlebotomy, antimalarials
Quiz

Which of these infectious diseases is most commonly associated with this disorder?

A. Hepatitis B
B. Hepatitis C
C. Syphilis
D. Varicella
Thank You!

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