Uncommon Rashes in Kids

2nd Annual Dermatology Refresher Symposium
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Objectives

• Recognize *uncommon* rashes in pediatric population.

• Be alert when *common* rashes are somewhat *uncommon* based on manner of presentation or response to treatments.

• Become familiar with basic treatment options.

• Understand when referral to sub-specialist is needed.
Uncommon Rashes

Langerhans Cell Histiocytosis
Daries Disease
Cutaneous Larva Migrans
Langerhans Cell Histiocytosis
Congenital Self Healing Reticulohistiocytosis
Six weeks later . . .

Different baby
Congenital Self Healing Reticulohistiocytosis

- Hashimoto-Pritzker Disease

- Langerhan’s cell histiocytosis
  - CD1a and S100 positive

- Traditionally considered a benign disease
  - Spontaneous involution over 3 months
    - otherwise, healthy without systemic involvement

- 3% risk of mortality & 10% chance of relapse
Evaluation/Work-up

- Rule out systemic involvement

- Example: Letterer-Siwe disease
  - Potentially fatal variant of LCH
  - Also occurs < 1y/o but has significant visceral involvement

<table>
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<th>Table 2. Histiocyte Society Guidelines for Diagnosis</th>
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<td>Physical examination</td>
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<td>Hematologic and coagulation studies</td>
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<td>Liver function tests</td>
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<td>Skeletal radiographic survey</td>
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<td>Chest radiography</td>
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Table 2: Guidelines from the Histiocyte Society for evaluation of an infant with biopsy confirmed LCH.

6 weeks of age—near clearance
Outcome

- Most common long term sequella: post-inflammatory pigmentary changes

- Involuted cutaneous lesions may also leave
  - Residual scars, Milia, Anetoderma

- Relapse rate of approximately 10 %
  - 6% developed systemic involvement after cutaneous resolution
  - All of the relapses occurred < 1 year of age
  - 31% of the infants with a reported relapse died

And then there is more . . .

- Solitary lesion in healthy infant
- Tissue culture negative (fungal, bacterial, mycobacterial)
- HSV negative, labs normal
- Skin Biopsy: LCH
  - SQ infiltrate of cells with lobulated reniform nuclei
  - CD1a, S100, CD68 & CD43 positive
  - AFB, GMS, HSV stains negative
- CXR, skeletal survey normal
Darier’s Disease
Diagnosis

- Hereditary acantholytic dyskeratosis
  - Autosomal dominant

- Biopsy of skin
  - Suprabasal acantholysis with dyskeratosis

- Mutation in ATP2A2 gene,
  - SERCA enzyme/pump required to transport calcium
  - Desmosomes cannot assemble
Clinical Findings

- Harsh, coarse, sandpaper like skin colored to yellow/brown papules located:
  - Seborrhoeic areas of the face, Scalp and neck
  - Central chest and back
  - Skin folds
- Coalesce to form wart plaques with odor
- Pits on palms/soles
- Nail changes
  - V-nicking & red/white longitudinal bands
- Mucous membranes with cobblestone pattern
- Flares: heat, topical steroids, & superinfection
Treatment Options

• Topical
  • Tretinoin
  • Keratolytics

• Systemic
  • Isotretinoin
    • Teratogenic
    • Requires lab monitoring
  • Acitretin
    • Requires lab monitoring
  • Cyclosporine

• Other (prevent superinfection)
  • Antibiotics for bacterial & fungal
Cutaneous Larva Migrans
Ancylostoma braziliense/caninum
https://www.cdc.gov/dpdx/hookworm/index.html
Ground Itch

- Upper level of dermis of skin
- Rarely can cause
  - Iron deficiency anemia
  - GI issues
  - Respiratory issues
- Less commonly, larvae may migrate to the bowel
  - cause an eosinophilic enteritis
- Advance a few mm to a few cm daily
- Treatment: most lesions resolve without treatment within 4-8 weeks
  - Mebendazole/ivermectin, topical steroids & antihistamines
Common Rashes that are *Uncommon* (per se)

Tinea
Atopic Dermatitis
Psoriasis
Acrodermatitis Enteropathica
Tinea Incognito
Tinea Incognito
Widespread Tinea Corporis

- Diffuse annular plaques throughout entire body
- Associated pruritus
- Genital and facial involvement as well
- S/p extensive work up to include:
  - Skin biopsy → spongiotic dermatitis c/w Erythema Annulare Centrifugum (EAC)
  - RPR, ANA, CBC, CMP → elevated ANA
Other Tinea Types
Kerion

- Immediate initiation of ORAL antifungals to prevent permanent damage
- Hair EMERGENCY
- NOT a bacterial infection → does NOT respond to Abx
KOH Hair

- Hyphae and spores within hair shaft
- Destruction and breakage leads to permanent scarring
Classic Tinea

- Hyphae Elements noted on KOH exam
- Topical Treatments for localized involvement
Atopic Dermatitis
Pathophysiology

• Chronic skin condition characterized by dry, itchy skin
  • 10-20% in school age kids; 90% have dz before 5

• Pathogenesis:
  • Impaired Barrier Function
  • Excess cytokine release (Type 2 inflammation)
  • Deficiency in ceramide
    • Increased transepidermal water loss
    • Lower skin surface hydration levels
    • Increased susceptibility to environmental factors
  • Loss of function in filaggrin gene
Treatment Options
General Skin Care

- Moisturization, moisturization, moisturization . . .
- Cream, washes, lotions, ointments, sprays and more
- Bleach Baths
  - 1/4 cup bleach in 1/2 bath tub
  - 1TSP bleach per gallon of water
- Wet Wraps/Wet PJs
- Avoidance of known irritants/triggers
- Fragrances, weather changes, pet dander, environmental
Topical rx steroids

• Low-potency
  • Safe on the face, groin, under arms for all patients AND infants < 1 year
  • When used less than 2-weeks at a time
    • Hydrocortisone 2.5%, desonide

• Mid-Potency
  • Safe on the body for children > 1 year
  • When used less than 2-weeks at a time
    • Triamcinolone, mometasone

• High-Potency (select uses)
  • Clobetasol, betamethasone
Topical Rx Non steroids

**Calcineurin Inhibitors**
- Tacrolimus-- 0.03% & 0.1% ointment (2 yrs & up)
  - Black Box Warning→Lymphoma
- Pimecrolimus– 1% cream (2 years & up)
  - Black Box Warning→Lymphoma

**Phosphodiesterase (PDE4) Inhibitors**
- Crisaborale-- 1% ointment (3 mo & up)
  - Mild to moderate AD
  - No limit on location, amount or duration
  - Irritation/redness and application site pain

**Januse Kinase (JAK) Inhibitors**
- Ruxolitinib1.5% cream– (12 yrs & up)
  - Short term non continuous treatment of mild to moderate AD
  - Black Box Warning
    - Serious infection, malignancies, major adverse cardiac events (MACE),
    - Thrombosis, thrombocytopenia, anemia, neutropenia, lipid elevation
  - Limit to 20% BSA for no more than 8 wks
  - Not in combination with other immunosuppressive medications
Systemic Rx Options

**NOT FDA approved for AD– specialist referral**

- Prednisone, methotrexate, cyclosporine, azathioprine, mycophenolate

**FDA approved for AD– specialist referral**

- **Dupilumab– Interleukin 4/13 receptor blocker**
  - Decreases inflammatory cytokines thought to contribute to AD signs & sx
  - Side effects: injection reaction, conjunctivitis
  - Lab monitoring: NONE
  - Efficacy– 75% of pediatric patients were 75% improved in AD at week 16
  - Method of Administration: SQ injection at home
    - Monthly to every other week– weight-based dosing
    - 6y/o and above
  - Non-immunosuppressive

- **Tralokinumab– Interleukin 13 receptor blocker– like above**
Janus Kinase Inhibitors (JAKi)

- **Upadacitinib**— oral JAKi (Higher JAK1)
  - Once daily dose 15mg or 30mg
  - 12 y/o and up

- **Abrocitinib**— oral JAKi (Higher JAK1)
  - Once daily dose 100mg or 200mg
  - 18y/o and up

- **Indication:**
  - Moderate to severe AD not responding to other pills, injections, or biologic medications
  - Not with other systemic medications for AD

- **Black Box Warning**
  - MACE, Thrombosis

- **Laboratory Monitoring:**
  - Anemia, neutropenia, lymphopenia, hyperlipidemia, transaminitis

- **Cytochrome Drug Interactions**
Other *Uncommon* Eczematous Conditions
Nummular Eczema

- Coin shaped lesions, commonly on extremities
- More difficult treatment response than classic AD
- Topical/oral steroids
Dyshidrotic Eczema

- Pompholyx
- Itchy, watery blisters that affect hands/feet
- Worsens with stress, sensitivities, heat, sweat & wet condition
Allergic Contact Dermatitis

- Patch Testing ➔ 80 item North American Comprehensive Panel
- Topical steroids and avoidance of allergen
- Overlying ACD can exacerbate atopic dermatitis
Eczema Herpeticum

- HSV viral superinfection of AD requiring specialist referral and possible hospitalization
- Avoid immunosuppressive medications
- Treat with IV/oral acyclovir +/- oral Abx for bacterial superinfection
Molluscum Dermatitis & Gianotti Crosti Like Dermatitis

- Reaction patterns to molluscum infection
- Topical regimen for sxs but improve when viral infection is resolved
Dermatitis Herpetiformis

- Celiac Disease → Avoidance of Gluten
- Biopsy PLUS DIF → Subepidermal with neuts and DIF with granular IgA and C3 along DEJ and within papillae
Psoriasis
Psoriasis

- Chronic immune-mediated disease
  - 2% of the U.S. population
- Age of onset
  - ages 20-30 & ages 50-60
  - 30% of patients with first-degree relative
- Subtypes
  - Plaque
  - Guttate
  - Inverse
  - Erythrodermic
  - Palmoplantar
  - Pustular
Subtypes: Plaque & Guttate
Subtypes: Inverse & Erythrodermic
Subtypes: Palmoplantar
Subtypes: Pustular Psoriasis
Other *Uncommon* Psoriasis Types
An Autoinflammatory Disease with Deficiency of the Interleukin-1–Receptor Antagonist

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DIRA vs DITRA

• DITRA: Deficiency in Interleukin-36 Receptor Antagonist

• DIRA: Deficiency in Interleukin-1 Receptor Antagonist

• Auto-inflammatory syndrome
  • Pustular Rash (pustular psoriasis)
  • Joint swelling with arthritis/pain
  • Nail changes
  • Bony changes
Autoinflammatory Conditions

DITRA

• Sudden repeated episodes of high-grade fever, generalized rash, and disseminated pustules

• Hyperleukocytosis and elevated serum levels of C-reactive protein

• Txt: Remicaide, Stelara, Cosyntex

DIRA

• Similar to an acute severe systemic infection/infection of the bone

• Increased ESR/CRP

• Characteristic Xray changes
  • balloon-like swelling of the ends of the ribs (all children)
  • periosteal elevation along multiple long bones
  • heterotopic ossification around the hip
  • multifocal osteolytic lesions (may result in vertebral crush fracture)

• Txt: Anakinra
DIRA
Treatment Options

Psoriasis
# Topical Treatments in Psoriasis

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<th>Medication</th>
<th>Uses in Psoriasis</th>
<th>Side Effects</th>
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<td>Topical steroids</td>
<td>All types of psoriasis</td>
<td>Skin atrophy, hypopigmentation, striae</td>
</tr>
<tr>
<td>Calcipotriene (Vitamin D derivative)</td>
<td>Use in combination or rotation with topical steroids for added benefit</td>
<td>Skin irritation, photosensitivity (but no contraindication with UVB phototherapy)</td>
</tr>
<tr>
<td>Tazarotene (Topical retinoid)</td>
<td>Plaque-type psoriasis. Best when used with topical corticosteroids.</td>
<td>Skin irritation, photosensitivity</td>
</tr>
<tr>
<td>Coal tar</td>
<td>Plaque-type psoriasis</td>
<td>Skin irritation, odor, staining of clothes</td>
</tr>
<tr>
<td>Calcineurin inhibitors</td>
<td>Off-label use for facial and intertriginous psoriasis</td>
<td>Skin burning and itching</td>
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Systemic Treatments

• Phototherapy
  • narrow-band ultraviolet B light (NBUVB)
  • broad-band ultraviolet B light (BBUVB)
  • psoralen plus ultraviolet A light (PUVA)

• Traditional Oral Medications
  • Methotrexate
  • Acitretin
  • Cyclosporine
  • Apremilast (March 2014)

• Biologic Agents
Systemic Treatments- Biologic Agents

- **TNF-α inhibitors**
  - Etanercept- 4 years and up
  - Infliximab
  - *Adalimumab

- **IL 12/23 blocker**
  - Ustekinumab- 6 years and up

- **IL 23**
  - Guselkumab
  - Risankizumab
  - Tildrakizumab

- **IL-17 blocker**
  - Secukinumab- 6 years and up
  - Ixekizumab- 6 years and up

* 2years & up for JIA; 12 years & up for HS
Updates In Psoriasis: Cardiovascular Disease

- Link to cardiovascular & cardiometabolic disease
  - Inflammation, epidermal proliferation & angiogenesis

- Severe disease results in death 5 years younger (based on patient risk factors for mortality)
  - Risk is similar to that of diabetes
  - Recs for screening for CV risk factors

- Increases in aortic inflammation is equivalent to a decade of aging
  - The greater the skin involvement, the higher the aortic inflammation

- Subcutaneous fat is increased under plaques
Acrodermatitis Enteropathica
Zinc Deficiency

• Triad: dermatitis, diarrhea & alopecia

• Congenital Form:
  • Mutation in SLC39A4 gene (zinc transporter protein)

• Acquired Form:
  • Mother unable to secrete zinc in breast milk (absent zinc binding factor)
  • After GI surgery

• Presents upon discontinuation of breast milk

• “Horseshoe” facial dermatitis

• Diaper area- plaques with peripheral scale
Questions

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CME Question

Which of the following uncommon rashes can signify an underlying condition and requires specialist referral/evaluation?

A. Acrodermatitis Enteropathica
B. Langerhan’s Cell Histiocytosis
C. Eczema Herpeticum
D. Erythrodermic or Pustular Psoriasis
E. All of the above