Pearls for Challenging Cases in Skin of Color in Pediatric Dermatology

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Disclosure

• I have no conflicts of interest or financial relationships to disclose.
Case 1

Inpatient Consult for “generalized rash”
Case 1

• 16 yo Somali female with presented to the ER for 10 day history of worsening rash affecting entire body.

• Prior treatments include Permethrin for suspected scabies without improvement and triple antibiotic therapy for newly diagnosed gonorrhea, and empiric therapy for chlamydia and syphilis (Penicillin, Ceftriazone and Azythromycin).

• Patient acutely decompensated and became febrile, tachycardic and hypotensive and was transferred to PICU. She received Vancomycin, blood cultures and skin cultures from pustules noted on admission were negative.

• Dermatology was consulted on hospital day 2.
Differential Diagnosis

• Pustular psoriasis
• Generalized gonorrhea
• Acute generalized exanthematous pustulosis
• Drug rash with eosinophilia and systemic symptoms (DRESS)
Shave biopsy, left upper arm: Subcorneal pustular dermatosis with psoriasiform dermatitis and spongiform pustules, consistent with pustular psoriasis.

Elongation and hypertrophy of rete ridges, epidermal hyperplasia and neutrophils in the parakeratotic layer (Munro microabscess)

Hyperkeratosis and parakeratosis

Extensive neutrophilic infiltrate with spongiform pustules below horny layer (Kogoj’s microabscesses)
Psoriasis is uncommon compared to Caucasians. It is seen in about 0.1% of American and West African Blacks, 1.4% of East African Blacks.

Childhood psoriasis is rare and has no gender predilection.

Clinically, underlying erythema more difficult to detect and overall skin involvement may be more extensive at time of diagnosis.
Pustular Psoriasis

Possible triggers may include:

- Sudden withdrawal of injected or oral corticosteroids
- Drugs: lithium, aspirin, indomethacin, iodide, terbinafine, minocycline and some beta-blockers

In infants and children there is not usually an implicated drug.
Pustular Psoriasis

• Generalized pustular psoriasis can be life-threatening, hospitalization may be considered. Systemic symptoms include: Fever, tachycardia, cachexia and muscle weakness.

• Complications:
  • Cardiorespiratory failure during the acute phase
  • Secondary bacterial infection
  • Anemia, lymphopenia with increased inflammatory markers (ESR, CRP)
  • Protein and electrolyte imbalance: low albumin, low calcium and low zinc
  • Renal and liver impairment
  • Malabsorption
Pustular Psoriasis - Treatment

• **Acitretin** is the drug of choice, with rapid response.

• Alternatives: Methotrexate, Cyclosporin, Prednisone (with caution) and biologics including Infliximab, Etanercept, Ustekinumab and Adalimumab.

• Our patient was treated with Cyclosporine 5 mg/kg/d divided BID and topical steroids (desonide & triamcinolone for face and body respectively).
After 3 days of Cyclosporine
After 3 days of Cyclosporine
 Pearls: Psoriasis & Skin of Color

• Erythema is not prominent, lesions appear violaceous or hyperpigmented.
• Post-inflammatory hypo- or hyperpigmentation is very common.
• Consider potential clinical mimickers: lichen planus (especially hypertrophic type), cutaneous lupus erythematosus (discoid and subacute).
• Potential extensive involvement at initial presentation.
• Managing scalp psoriasis: Consider impact of hair texture, styling practices, and washing frequency when evaluating severity and selecting topical therapy.
Case 2

Outpatient visit for “eczema flare”
Case 2

- 9 year old African American male presented to Dermatology clinic for follow-up evaluation of eczema. He was last seen 2 years ago and prescribed Desonide 0.05% ointment and Triamcinolone 0.1% ointment BID for face and body respectively, and oral antihistamines (Cetirizine 5 mg and Hydroxyzine 20 mg) for pruritus.

- Parents used topical therapy for less than 2 months, after noticing his skin was “turning white with the treatment”.

- He is currently using an OTC moisturizer lotion about once to twice daily. His scalp is very itchy, sleep is not disturbed.

- Eczema is now flaring. He is otherwise healthy, had had no preceeding illness or fever.

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Differential Diagnosis

- Psoriasiform dermatitis
- Hypopigmented mycosis fungoides
- Eczematous dermatitis
- Pityriasis rubra pilaris
- Pityriasis lichenoides choronica
- Syphilis
- Nutritional deficiency
4x shows overall and focal of follicular plugging. 10x shows alternating hyper- and hypo-granulosis, and some variation in the stratum corneum. Findings are subtle but suggestive of pityriasis rubra pilaris (PRP).
• Folliculocentric keratotic “nutmeg-grater” papules lacking erythematous base
• Absence of salmon-colored plaques, instead numerous hypopigmented plaques with “islands of sparing”
• Waxy skin colored to hyperpigmented (not orange) keratoderma of palms and soles.
Pityriasis Rubra Pilaris

<table>
<thead>
<tr>
<th>Type</th>
<th>Frequency</th>
<th>Clinical features</th>
</tr>
</thead>
<tbody>
<tr>
<td>I Classic adult</td>
<td>Most adults</td>
<td>Follicular keratotic papules, first on face and extending caudally; progresses to generalized keratoderma with islands of sparing; palms and soles usually involved; generally clears within 3 years</td>
</tr>
<tr>
<td>II Atypical adult</td>
<td>Rare</td>
<td>More ichthyosiform scaling; coarse palmoplantar keratoderma; long duration</td>
</tr>
<tr>
<td>III Classic juvenile</td>
<td>14-35% of children</td>
<td>Same as type I</td>
</tr>
<tr>
<td>IV Circumscribed juvenile</td>
<td>Most common type in children</td>
<td>Thick plaques on knees, elbows; palms, and soles involved</td>
</tr>
<tr>
<td>V Atypical juvenile</td>
<td>Rare, familial, Onset in first years of life</td>
<td>'Sclerodermatous' changes on palms and soles; follicular hyperkeratosis</td>
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- PRP in children is a non-inherited dermatosis with no gender predilection
- Most common presentation: Type III Classic juvenile form
- Retinoids should be considered as first-line treatment for PRP.
- Other therapies include topical corticosteroids, topical calcineurin inhibitors, anti-TNF alpha inhibitors
Pearls: Pityriasis Rubra Pilaris & Skin of Color

- High index of suspicion with presentation of psoriasiform dermatitis and keratoderma
- Erythema is not prominent, lesions appear hypo or hyperpigmented.
- Post-inflammatory hypo- or hyperpigmentation is very common in comparison to classic salmon colored plaques with “islands of sparing”
- Consider potential clinical mimickers: hypopigmented mycosis fungoides, psoriasis
Case 3

Inpatient consult for “swollen face”
Case 3

- 16 year old African American female presented with 1 week of fever (102-104 °C) and general malaise
- Recently treated with amoxicillin for acute otitis media
- Following admission, she developed facial rash associated to rapidly progressive periorbital edema
- Labwork was remarkable for pancytopenia
Differential Diagnosis

• Reactive process (drug or viral exanthem)
• Manifestation of an autoimmune disorder
Work-up

- Skin biopsy from nodule on right leg revealed superficial and deep mixed perivascular and interstitial dermatitis. Histotologic features were not diagnostic. Pattern of inflammation favor a variant of palisaded and neutrophilic granulomatous dermatitis, which can be associated with underlying systemic and connective tissue disorders. A drug eruption, reactive erythema, exuberant urticarial or reactive process, and other similar dermatoses could also present with these features.”
Diagnosis

• During hospitalization, she developed arthritis, hemolytic anemia, leukopenia, thrombocytopenia, hypocomplementemia, ANA, Anti-Sm, Antiphospholipid Antibodies.

• **Diagnosis: Systemic lupus erythematosus.**

• Management: IV Steroids and Plaquinil.

• Rash and periorbital edema quickly improved. Fevers stopped
Systemic Lupus Erythematosus

• Severe organ involvement and significant disease activity are primary characteristics in children with juvenile SLE.
• African Americans are at highest risk for severe organ involvement.
• The clinical manifestations of lupus nephritis (LN) in children are diverse.
• Early treatment may result in significant disease remission.
Periorbital erythema and swelling as a presenting sign of lupus erythematosus in tertiary referral centers and literature review.

Wu MY1,2, Wang CS3,4, Ng CY1,2, Kuo TT2,4, Chang YC1,2, Yang CS1,2, Lin YH1,2, Ho HC1,2, Chung WH1,2, Shen CS1,2.

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Bilateral periorbital swelling as the initial presentation of cutaneous lupus erythematosus.

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Lupus and Skin of Color
In contrast to newborns
Neonatal Lupus Erythematosus

• Caused by transplacental passage of maternal antibodies, most commonly anti-Ro/SS-A
• 50% of women with a child with NLE are asymptomatic at time of child’s birth
• Cardiac disease results in 20% mortality -> Congenital heart block
• Women who have a prior child with NLE have a 25% risk of NLE in subsequent children
Pearls: Lupus and Skin of Color

• Chronic cutaneous lupus Erythematous is a scarring subtype, more prevalent in blacks

• Periorbital edema has been described as a presenting sign of lupus

• Skin findings tend to be exaggerated with an increased risk of post-inflamatory hyperpigmentation and hypertrophic scarring

• Consider potential clinical mimickers in neonatal lupus: seborrheic dermatitis, pityriasis alba and atopic dermatitis