Case Studies in Pediatric Dermatology

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Case Studies in Pediatric Dermatology

I have no relevant conflicts of interest to declare!!
Case Studies in Pediatric Dermatology

Will discuss cases in the following categories:

- Neonatal Dermatoses & Lesions
- Childhood Dermatoses & Lesions
- Adolescence
- Miscellaneous Case Discussions
Neonatal Dermatoses & Lesions
Neonatal Dermatoses & Lesions

- Infant born with hypopigmented patch on abdomen, inner thighs
- Borders somewhat “feathery”
- No preceding rash
- o/w healthy
- Pregnancy uneventful
Neonatal Dermatoses & Lesions
Pigmentary Mosaicism

- Heterogeneous group of disorders with patterned streaks of hypo/hyperpigmentation
- Streaks/whorls tend to follow lines of Blaschko – lines of ectodermal embryologic development
- Reflects gene mosaicism of affected areas
- In extensive involvement can see gross chromosomal abnormalities (60%)
- Do not tend to be hereditary
Pigmentary Mosaicism

- Not always present at birth, can “evolve” over time
- Can have associated abnormalities of the bones, eyes, and/or CNS (30%)
- No effective treatment
- Other names:
  - Hypomelanosis of Ito = incontinentia pigmenti achromians
  - Linear and whorled nevoid hypermelanosis
  - Nevus depigmentosus
Pigmentary Mosaicism

This is not the same thing as vitiligo!!
Neonatal Dermatoses & Lesions

- Neonate born to a 21 yo healthy woman, no complications during pregnancy
- Found to have diffuse involvement with brown plaques and nodules
- Hypoplastic right arm
Neonatal Dermatoses & Lesions
Congenital Melanocytic Nevus

• Present at birth; some small-medium sized CMN occur during 1\textsuperscript{st} yr of life
• Usually classified by greatest diameter in adulthood:
  – Small <1.5 cm
  – Medium 1.5 – 19.9 cm
  – Large 20 cm or greater
  – Giant CMN occupy significant portion of skin surface
Congenital Melanocytic Nevus

• 1% of newborns have small CMN, large occur in 1/20,000, giant occur in 1/500,000
• Large/giant CMN may be associated with cosmetic disfigurement, ↑’d risk of melanoma, neurocutaneous melanosis (NCM)
• NCM more likely in patients with axial involvement and satellite lesions
• NCM sx’s usually occur during 1st 3 years, prognosis poor
  – >90% die from dz, 70% before age 10
• Magnitude of risk for MM remains controversial, probably at least 5-10% for large/giant nevi
Neonatal Dermatoses & Lesions
Congenital Melanocytic Nevus

- Therapy: individualize for the patient
- Excision a consideration for lesions
  - Study on surgical excisions: 74% of patients preferred scar over nevus, 26% did not!* 
- Close clinical surveillance
- Education: www.aad.org
- Support groups: Nevus Outreach, Inc. www.nevus.org

Neonatal Dermatoses & Lesions

• Infant born with a light, hypopigmented patch on upper chest
• Area turned bright red within 2-3 weeks
• Lesion continued to grow for another 6 months
• Patient o/w healthy
Hemangioma of Infancy

- 2.5% of all neonates: most common benign tumor of childhood, up to 10% of Caucasians by age 1
- Female: male = 3:1, more common in prematurity
- Demonstrate endothelial cell hyperplasia
- 30% faint pink or white macules at birth
- Most appear between 1-4 weeks
- Head and face most common site (60%)
- 80% solitary
Hemangioma of Infancy

- Pathogenesis is unknown
- Positive endothelial cell staining with GLUT1, the erythrocyte-type glucose transporter protein, noted in all growth phases (absent in other VAs)
- This protein normally expressed in the microvascular endothelia of blood-tissue barriers
- ? From embolized placental cells
Hemangioma of Infancy

- Three types: superficial, deep, and mixed
- Proliferation: 1-10 months
  - Most rapid between 1-4 months
  - Watch for complications
- Involution: After 6-10 months
  - Softens, develops gray surface
  - 50% gone by age 5
  - 70% gone by age 7
  - 90% gone by age 9
Superficial Hemangioma of Infancy
Deep Hemangioma of Infancy
Mixed Hemangioma of Infancy
Hemangioma of Infancy

- Lesions do not always leave normal skin!!
- 20% of patients may have: residual wrinkling, atrophy, telangiectasias, hypopigmentation, and redundant fibrofatty tissue or skin
- Fewer than 2% of lesions leave significant cosmetic disfigurement requiring surgical intervention
Hemangioma of Infancy

• Complications:
  – Ulceration and/or infection 10-15% of cases
  – Alters vital functions
  – Kasabach-Merritt Syndrome
  – Congestive heart failure
  – Posterior fossa defects
  – Residual skin changes
Hemangioma of Infancy

• ~20% of patients will have some residual defect

• Traditional forms of therapy:
  – Observation
  – Pulse dye laser
  – Corticosteroids: oral, intra-lesional, topical
  – Imiquimod
  – Interferon alpha
  – Vincristine
  – Surgery
Hemangioma of Infancy

- Propranolol used in 11 children aged 2-6 months with severe HOI:
  - Propranolol started at 2 mg/kg/dy
  - 24 hrs after initiation dramatic softening, with intense red to purple color change
  - No regrowth after steroids stopped
  - HOI continued to improve until nearly flat, with residual telangiectasia

Hemangioma of Infancy

• But this is off-label usage in kids!!
• Potential side effects:
  – Typically mild and temporary
  – Hypoglycemia
  – Bradycardia and hypotension
  – Dizziness, lethargy, fatigue
  – Diarrhea, nausea, vomiting
  – Bronchospasm
  – Hypersensitivity reactions rare: rash (including EM, SJS, TEN), urticaria, fever, agranulocytosis, anaphylaxis
Hemangioma of Infancy

• Suggested use:
  – Cardiology referral for clearance
  – 2 mg/kg/day divided tid, monitor BP and HR for several hours when initiate
  – Slowly taper off when course completed
  – About 6 months of therapy seems best
  – Each month of therapy seems comparable to ~1 year of spontaneous resolution
Hemangioma of Infancy
Hemangioma of Infancy
Hemangioma of Infancy

- Be wary of segmental or beard hemangiomas
Hemangioma of Infancy
6 month old girl was born with these pink marks on her face and occipital scalp/neck. The patches have faded over time, remain flat. Patches become more pronounced when patient is crying. o/w healthy.
Salmon Patch = Nevus Simplex

- Most common vascular lesion of infancy
- Occurs in 30 – 40% of all newborns
- Often on glabella, forehead, upper eyelids, occasionally nose and nasolabial area
- When seen on nape of neck = “stork bite” and “angel kiss” when on forehead
- Most common form of capillary malformation but many think a form of fetal circulation
Salmon Patch = Nevus Simplex

- Usually isolated, no associated findings
- 95% of facial lesions fade during 1st 2 yrs of life
- Stork bites may fade but many persist indefinitely
- Parents should be educated about “reappearance” of facial lesions during episodes of increased intra-vascular pressure (crying, exertion, straining)
- Pulsed dye laser can be used if see persistence
Neonatal Dermatoses & Lesions

- 4 yo girl was born with this red mark on her face
- The patch has not changed over time, remains flat
- o/w healthy
Port Wine Stain = Nevus Flammeus

- 0.3 % of all newborns
- Capillary malformation
- Equal sex distribution
- Present at birth, darken with advancing age
- Represent ectasia of superficial vascular plexus
- Usually unilateral and segmental (~85 %)
- Face and neck most often affected
Port Wine Stain = Nevus Flammeus

- < 10% of PWSs in trigeminal distribution associated with ocular or CNS involvement
- Can see underlying soft tissue hypertrophy
- Pyogenic granulomas and basal cell carcinomas can sometimes occur within PWS
- PWSs can be seen in syndromes:
  - Sturge Weber
  - Klippel-Trenaunay/Weber
  - Cobb
Port Wine Stain = Nevus Flammeus

• Rationale for treatment:
  – Psychosocial morbidity in young children (but no increase in serious mental illness)
  – Early treatment *may* prevent thickening and nodularity
  – Early treatment *may* prevent soft tissue hypertrophy
• Treatment is with the pulsed dye laser (585 nm)
Childhood Dermatoses & Lesions
Childhood Dermatoses & Lesions

- 9 yo boy presents with a slightly crusted, slightly sore area beneath his nose, present for 1 week
- He has a h/o dry, sensitive skin, but no eczema
Impetigo

- A common, contagious superficial skin infection caused by strep, staph, or both
- Can occur anywhere on body, but mostly on exposed areas
- Two classic forms:
  - Bullous (usually S. aureus)
  - Nonbullous = crusted (>70% of cases, strep was main organism (GABHS), but now staph more common)
- Fever & lymphadenopathy can occur, more with GABHS (and rarely see glomerulonephritis or scarlet fever)
- Rx can be topical or systemic
Childhood Dermatoses & Lesions

- 8 yo girl with 1-2 week h/o small “sores” spreading on her cheek, somewhat tender
- She does have a h/o eczema
- No recent illnesses
Cutaneous Herpes Simplex

- Can occur on any body surface area
- Can be easily misdiagnosed as impetigo or zoster
- Can get secondary bacterial infection or lymphangitis
- Usually caused by HSV-1 in infants and children
- Eczema herpeticum is a disseminated HSV infection occurring in pts with atopic dermatitis
- In disseminated cases, need IV acyclovir
Cutaneous Herpes Simplex
Childhood Dermatoses & Lesions

- 9 yo with fairly rapid onset of small bumps over the course of a couple weeks
- Some areas associated with itching and redness
- Child has a h/o eczema
Molluscum Contagiosum

- Caused by pox virus
- Often affect head, neck & trunk and may be extensive in children with atopic dermatitis
- Genital MC increasing in sexually active young adults
- May spontaneously regress
- Treatments: cantharidin, cryotherapy, aldara, tretinoin cream, salicylic acid, 5-FU, imiquimod cream (curetage in adults)
Childhood Dermatoses & Lesions

- 12 yo girl has been developing numerous flat, brownish bumps diffusely over the face
- They continue to spread
- Occasional itch, mostly no Sx’s
Flat Warts = Verrucae Plana

- HPV types 3, 10, 28, 41
- Occur primarily on the face, neck, arms, and legs
- Can get several hundred lesions!
- Often see koebnerization
- Treatment difficult due to number of lesions:
  - Cryotherapy, topical retinoids, aldara (immunomodulatory), topical 5-FU, oral cimetidine
Common Warts = Verrucae Vulgaris

- HPV types 1, 2, 4, 7
- Most common site is hands
- Transmission skin–skin, fomites
- Incubation 1 - >6 months
- Very common in children and adolescents
- Treatments: topicals such as salicylic acid, cryotherapy for resistant warts, immunotherapy

And education/patience!!!
- 2/3 resolve spontaneously by 2 yrs
Common Warts
Plantar Warts = Verrucae Plantaris

- HPV types 1 & 4
- Sole of foot: single or in group causing ‘mosaic’ appearance
- Hyperkeratotic
- May cause walking pain
- Treatments: plasters, cryotherapy, laser, curettage, immunotherapy
- Very difficult to treat!
Childhood Dermatoses & Lesions

- 8 yo child developed recent onset nails breaking, “lifting off” nail bed
- Child o/w healthy
- She did have diarrhea with sores in her mouth ~6 weeks prior
Onychomadesis

- Onychomadesis = proximal complete separation of the nail
- Results from full, but temporary, arrest of growth of the nail matrix
- Described in a variety of stressful events, e.g., Hand-Foot-Mouth Disease
- Beau’s lines = transverse grooves that originate under the proximal nail fold, from partial arrest of the nail matrix
Childhood Dermatoses & Lesions

- 7 yr old healthy boy with a 5 month appearance of several new depigmented patches
- No preceding rash
- No scaling, itchiness
Childhood Dermatoses & Lesions

- Several depigmented well-demarcated patches have occurred
Vitiligo

- Affects ~1% of the population
- May begin at any age, 25% develop before age 8, 50% begin prior to age 20
- Mean age onset in kids: 6.2 years
- Rarely congenital
- Most pts otherwise healthy, occasional alopecia areata and thyroid disease seen
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Vitiligo

- Pathomechanism: most likely autoimmune
- Autoantibodies that destroy melanocytes have been found (directed against melanocyte-specific antigens like tyrosinase and TRYP-1)
- See increased frequency of autoimmune disorders in family members
- Genetic component: 7-12% in first degree relatives
- Generalized vitiligo linked to chromosome 1p31
Vitiligo

• Location, size, shape of lesions vary considerably
• Typically ivory-white areas, with well-defined, convex borders
• Usually bilateral, can be asymmetric: “segmental” vitiligo (localized, unilateral) seen more in kids
• Universal or total vitiligo is rare
• 75% first lesions occur in exposed areas
• Predilection for body folds, orifices, bony prominences
Vitiligo

- 12% of pts show white hairs (leukotrichia)
- Halo nevi often found, and Koebner phenomena
- Pattern subtypes:
  - Generalized
  - Focal
  - Segmental
  - Acrofacial
  - Mucosal
  - Universal
Vitiligo: Koebner Phenomenon
Vitiligo: Treatment

• Children tend to respond more than adults with complete and permanent repigmentation
• Response is better with early onset disease
• Repigmentation usually occurs in hair follicles
• Potent topical steroids, twice daily
• Calcineurin inhibitors (especially for face)
• PUVA and narrowband UVB phototherapy
Vitiligo: Treatment

• Surgical methods:
  – Autologous grafting of non-lesional epidermis or cultured melanocytes
  – 308 nm excimer laser

• Benoquin = 20% monobenzyl ether of hydroquinone for complete depigmentation

• Cosmetic camouflage or aniline dye stains

• Photoprotection

• National Vitiligo Foundation: www.vitiligofoundation.org
DDX: Pityriasis Alba
DDX: Pigmentary Mosaicism
DDX: Pigmentary Mosaicism
DDX: Post-inflammatory
Adolescence
Comedonal Acne

• Consists of open and closed comedones
• The least inflammatory acne but paradoxically can be the most difficult to treat
• Treat with topical retinoids
Comedonal Acne
Papulopustular Acne

- Inflammatory acne
- Sometimes can be treated with topical retinoids and topical antibiotics
- Often requires treatment with systemic antibiotics
Nodulocystic Acne

- Can cause severe, permanent, disfiguring scarring
- Needs treatment with 13-cis-isotretinoin (Accutane)
Nodulocystic Acne

- Isotretinoin (13-cis retinoic acid):
  - Only for severe inflammatory acne
  - Not a first-line agent; try systemic abx’s first
  - 4 - 5 month course
  - 0.5 - 1 mg/kg/day
  - Monthly labs: CBC, LFTs, TG/Chol, HCG
  - Strict adherence to pregnancy prevention program:
    iPLEDGE Program www.ipledgeprogram.com/
Nodulocystic Acne

- Isotretinoin side effects:
  - Skin and mucous membrane dryness
  - Increased sun sensitivity
  - Hair loss, epistaxis, conjunctivitis
  - Triglyceride elevation
  - Musculoskeletal discomfort & skeletal changes
  - Depression
  - Severe teratogenicity
Acne: Does Diet Play a Role?

• Diet may play a role: low-glycemic load diet improves acne*

• Positive association with acne for intake of total milk and skim milk: may be because of the presence of hormones and bioactive molecules in milk?**

Case Discussions