Neonatal Dermatology Review

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Session Summary

This presentation will provide a review of the basic anatomy and function of the skin, identify transient benign lesions with a discussion of origin, significance and common resolution. More commonly identified developmental and infectious lesions in the newborn period will also be reviewed.

Session Objectives

Upon completion of this presentation, the participant will be able to:

- list the functions of the skin;
- recognize developmental characteristics of neonatal skin;
- identify common benign skin lesions in the newborn;
- recognize pathology associated with skin lesions in the newborn;
- identify the key features of skin lesions, color, including borders, location, onset and characteristics.

References


Some slides compliments of Dr. Tom Harris (TRH)

Session Outline

See presentation handout on the following pages.
Neonatal Dermatology

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Presented October 14, 2014
For FANNP

Functions of the Skin

1) Physical protection (barrier function):
   Provides mechanical, chemical (e.g., vernix), and bacterial (normal flora) protection for the inner body

2) Heat regulation:
   - Production and evaporation of sweat
   - Dilatation & constriction of blood vessels
   - Insulation of body by subcutaneous fat

3) Sense perception (heat, touch, pain, pressure)

4) Immunological properties (cutaneous immunosurveillance)

5) Useful for estimating gestation age by exam, and assessing nutritional status at birth (reflecting acute or chronic placental insufficiency, and resulting IUUGR/SGA, postmature and “dysmature” babies)
Lanugo

Estimating gestational age

Fingernails curving around fingertips indicate > 43 wks gest.

Estimating gestational age

Footsoles of markedly postmature, meconium-stained newborn, with deep, discolored plantar creases

Estimating gestational age

Translucent skin of the extremely premature, immature newborn infant, on the margin of viability

Assessing nutritional status

Size-10 skin on a body recently shrunk to size 8

Assessing nutritional status

Loss of subcutaneous fat tissue in a term but “dysmature” infant
FUNCTIONS OF THE SKIN (continued)

6) The skin is also useful as a medium through which to assess cardiovascular and/or respiratory status (e.g., capillary refill time, cyanosis, skin mottling etc.), or confirm the presence of birth injury, or suspicion of local or systemic infection, or identify minor anomalies or stigmata suggestive of occult major malformations or hereditary or chromosomal syndromes.

Testing for capillary refill time (CRT)

Mottled skin color and mild central cyanosis in a possibly septic baby!

The skin “mottling” could also be due to poor perfusion from congestive heart failure, or from metabolic acidosis in association with an inborn error of metabolism, etc.

Physiologic Phenomena:
Cutis Marmorata
- Reticulated bluish mottling
- Trunk and extremities
- Normal response to chilling
- Resolves in weeks to months

Physiologic Phenomena:
Cutis Marmorata
- If persists with warming, consider:
  - Cutis marmorata telangiectasia congenita
  - Cornelia de Lange
  - Down’s syndrome
- Cutis marmora alba
  - Hypertonia of deep vessels

Milia: Tiny inclusion cysts within the epidermis that contain concentric layers of trapped keratinized stratum corneum
Miliaria crystallina: Tiny, superficial 1-2 mm vesicles due to obstructions of the eccrine (sweat) glands at the subcorneal or intracorneal level. Miliaria rubra or "prickly heat" is composed of 1-3 mm papulopustules, and represents dermal inflammation around occluded eccrine ducts. Both of these benign conditions are most commonly seen in febrile or overheated infants.

Neonatal Acne: (to be distinguished from infantile acne) Erythematous papules & pustules (but without comedones) that appear at birth or within the first 2 to 3 wis of life. Usually in male infants on face. May clear within first 2 to 3 months or may transition into infantile acne (with comedones) lasting a year. Need no treatment.

Acne Neonatorum


Transient Neonatal Pustular Melanosis

- Benign, self-limited (24-48 hrs)
- MC in dark skinned infants
- Superficial sterile pustules that rupture leaving collarette of fine scale around hyperpigmented macule
- Histopathology: Subcorneal pustule of neutrophils
- Incidence: ~0.5-2% lasting up to 3 months.

Harlequin sign

Temporary imbalance of the autonomic regulatory mechanism of (dependent) cutaneous vessels

Complications Diagnostic Procedures

- Digital loss
- Cutaneous puncture marks
- Scalp electrode infection
- Burns
- Anetoderma of prematurity
- Calcinosis cutis
- Scars, lacerations
- Ocular trauma, blindness

Harlequin sign

Temporary imbalance of the autonomic regulatory mechanism of (dependent) cutaneous vessels

Transient neonatal pustular melanosis first appears as small superficial pustules without inflammation

Complications Diagnostic Procedures

- Digital loss
- Cutaneous puncture marks
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- Calcinosis cutis
- Scars, lacerations
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Developmental Anomalies, Disruptions, and Tumors of Newborn Skin

Occult Spinal Dysraphism
- A combination of 2 or more congenital midline skin lesions is the strongest marker of OSD.
- Spinal MRI is the diagnostic modality of choice
- Ultrasound may be considered for infants younger than 4 mo of age

Mongolian spots
Collections of melanocytes located in the dermis, thought to be due to failed migration of melanocytes from neural crest up to dermal-epidermal junction

Nevus of Ito
Easily confused with “Bathing trunk” Giant hairy nevus

Capillary Malformation (CM) or Salmon Patch
“Angle kiss” (when on forehead); “Stork bite” (when on nape)
- Midline vascular malformation of developmental origin; in essence, capillary ectasias
- Seen in nearly 50% of all newborn infants, so can’t be considered a significant minor anomaly
- Usually disappear by 1 to 2 yrs
- No pattern of inheritance, and not associated with any specific syndromes, in contrast to Port Wine Stains (aka “nevus flammeus”) which are off to one side, grow proportionate to the child’s growth and persist throughout life if left untreated.

Port Wine Stains (PWS) are also ectatic, malformed capillaries within the dermis.
However, they grow proportionate to the child’s growth and persist throughout the person’s life if left untreated.
Their thickness increases over the years and their color becomes crimson red or a deep purple hue.
Any infant with Port Wine Stain involvement of the skin in the V1 area of the Trigeminal Nerve (i.e., the ophthalmic division of CN V) is at risk for Sturge-Weber Syndrome & its accompanying problems.

Adnexal polyp composed of adnexal structures, i.e., hair follicles, vestigial sebaceous glands and eccrine glands

These polyps fall off spontaneously shortly after birth.

Nevus Sebaceous
Hamartoma or linear nodules of superfluous tissue, absent of hair
Persist and enlarge with 10-15% ultimately showing malignant degeneration

Strawberry Hemangioma
- Vascular tumor that may be superficial (as in this case) or deep or both, i.e., "combined"
- If multiple, may indicate visceral involvement
- Initially increase in size (especially in premature infants), reaching full size at about 4 months corrected gestational age. Then begin to slowly involute, becoming barely visible by school age
- Occur with increasing frequency with decreasing gestational age: 23% in babies < 1000 gms; 16% between 1,000 – 1,500 grams (possibly due to increased levels of vascular endothelial growth factor)

Deep or Combined Hemangiomata

If crucial structures are involved, the lesion may require steroids (to accelerate natural involution) or laser surgery
The same girl at 12 years of age after intensive steroid and laser ablation treatment

Nascent Hemangioma

A nascent hemangioma is as the name implies, a precursor to a deep form of hemangioma. They are permanent but require no treatment.

Congenital Erosive Vesicular Dermatosis

(A) Congenital erosive and vesicular dermatosis: newborn with extensive symmetrical ulcerative and erosive changes. (B) Superficial scar at age 4 months. (C) Acropatellar scarring at birth.

Cutis Aplasia Congenita

Often represents a benign, isolated autosomal dominant trait.

Newborn Baby

Baby’s Mother

However, in the Adams-Oliver Syndrome which also has an autosomal dominant inheritance pattern, the large scalp defect may involve the underlying cranium, and be associated with distal limb reduction defects that vary from partial absence of fingers or toes to loss of an entire distal limb.

Congenital melanocytic nevus (CMN)

aka: Congenital nevocellular nevus (CNCN) due to proliferation of nested melanocytes in skin

The risk of malignancy or change into malignant melanoma (MM) depends on lesion size, location, histology, and the patient age:

Small CMN (congenital melanocytic nevus) defined by some as < 1.5 cm and by others as < 2.5 cm.

Lifelong risk of MM is ~ 2 – 3%!
Large “Giant hairy nevus” with nodular texture
Lifetime risk of malignant melanoma (MM) is approx. 6 - 8%

Café au lait macules or CALMs are associated with Neurofibromatosis Type I Disease if 6 or more are present and measure at least > 0.5cm in length

Patients will have Lisch nodules or small pigmented spots on their iris seen after 6 years of age by slit lamp examination

To diagnose the autosomal dominant Waardenburg’s Syndrome, there should be iris heterochromia, lateral displacement of the medial canthi (called dystopia cathomorum) and hearing loss, beside the white forelock.

“Collodion baby” with lamellar ichthyosis. Only 5-6% of these babies will ultimately replace the collodion membrane with normal skin. Although the stratum corneum is thick, it is a poor barrier in both directions, allowing for excessive water, heat, and electrolyte loses, and absorption of toxins or invasion by pathogenic organisms.

Epidermolysis bullosa

Blisters develop at the mildest provocation
“Prune Belly” syndrome resulting from prior (i.e., early in utero) abdominal distension associated with congenital GU malformations. Notice also the left-sided Erb’s Palsy and equinovarus clubbing of the left foot.

Amniotic band sequence as the cause of the scalp skin disruption: Typical placenta with the torn amnion collapsed down around the insertion of the cord. Note also the hemorrhagic amniotic band attachments that tore away from the scalp at the time of delivery.

What happened?

Amniotic constriction bands

- Congenital constriction deformities
- Congenital amputation
- Rupture of amnion

Amniotic constriction bands: multiple anomalies of the feet

Skin Signs of Localized Bacterial Infection

Scalded Skin Syndrome due to certain species of Staph aureus
Bullous Impetigo
- Etiology: Staph, occasionally Strep
- Also known as pemphigous neonatorum
- Increased in areas of warmth and moisture
- Bulla become wrinkled, flaccid and rupture producing ulcers that become crusted

Skin Signs of Congenital Viral Infections

The Blueberry Muffin Baby
- What are the blueberries?
  - Extramedullary hematopoiesis

Neonatal Varicella
- Neonatal varicella: generalized crusted papules and vesicles
- Fetal varicella syndrome: Segmental deep scars, dermatomal distribution
Congenital Varicella

- Varicella during 1st 20 wks of gestation
- Highest risk between 13 – 20 wks
- Various findings: low birth wt, ophthalmologic defects, CNS defects, limb hypoplasia / contractures, GI/GU defects
- Cutaneous: vesicles, scarring, ACC-like
- Mother >5d before or infant 1-4 DOL = mild
- Mother w/in 5d or 2d post or infant 5-10 DOL = severe, disseminated dz (Pneumonia, Hepatitis, meningoencephalitis)
- Mortality 30%

Cytomegalovirus (CMV)

- “Blueberry muffin”
- Jaundice appears within 1st 24 hours of life
- Hepatosplenomegaly, abdominal distension, anemia, thrombocytopenia, macular chorioretinitis, periventricular calcification with enlarged lateral ventricles, hearing defect, microcephaly

Herpes

- Skin lesions are present at birth
- Other clinical manifestations vary and may not be present until the 4th to 8th DOL
- Lethargy, poor feeding, temp instability, jaundice, HSM all can lead to widespread dissemination, encephalitis and death
- Treatment: IV Acyclovir

Examples of HSV Lesions

Congenital HSV vs Neonatal

- Neonatal herpes simplex with multiple vesicles
- Intrauterine HSV infection. Deep atrophic/ulcerative lesions suggesting epidermolysis bullosa or aplasia cutis
Neonatal HSV

- Risk factors – vaginal birth
  - Primary genital HSV (40-50%)
  - Recurrent HSV (2-5%)
  - Fetal scalp monitoring
- 3 clinical presentations
  - Skin, eyes and mouth disease (SEM)
  - CNS disease
  - Disseminated disease
- Treatment - Acyclovir 60 mg/kg/day

Skin Signs of Neonatal Fungal Infections

Diaper Dermatitis

- Chafing dermatitis - friction
- Irritant contact dermatitis (ICD) – spares folds
- Candida – beefy red, pustular satellites

Candida Diaper Dermatitis

- Symmetric
- Involves intertriginous areas
- Erythematous, swollen, slightly scaly skin
- Satellite lesions
- Can lead to hypopigmentation if not treated appropriately

Irritant Contact Dermatitis

Diaper Dermatitis: Seborrheic dermatitis
**Congenital Syphilis**

- Treponema pallidum
- Transmission 70-100% for untreated
- 40% stillborn
- Early: birth to 2 yrs
- Late: after 2 yrs
- Treatment: crystalline Penicillin G

**Early Congenital Syphilis**

The raw hemorrhagic lesions on the soles of the feet so typical of Congenital Syphilis

(From Eichenfield et al., 2008)

The discoid lesions of Congenital Lupus look somewhat similar but never involve the palms of the hands or the soles of the feet.

(From Eichenfield et al., 2001)

**Describing Lesions**

- Primary Lesions
- Secondary Lesions
- Color, borders, configuration, and distribution of lesions
- Hyperpigmentation and hypopigmentation

**Primary Lesions**

Primary lesions are defined as lesions that arise de novo (not from another lesion) and are therefore most characteristic of the disease process.
Macule
A circumscribed, flat lesion with color change. Usually up to 1 cm. Not palpable. Examples: café au lait macules and capillary malformations

Café au lait macules


Patch
A circumscribed, flat lesion with color change. Greater than 1 cm in size. Examples: Nevus depigmentosus, mongolian spots, nevus simplex

Hemangioma precursor


Papule
A circumscribed, elevated, solid lesion, up to 1 cm in size. Elevation may be accentuated with oblique lighting. Examples: Verrucae, milia, and juvenile xanthogranuloma.

Umbilical granuloma


Plaque
A circumscribed, elevated, plateau-like, solid lesion, greater than 1 cm in size. Examples: Marked cytoma, nevus sebaceous

Nevus sebaceous


Nodule
A circumscribed, elevated, solid lesion with depth, up to 2 cm in size. Examples: Dermoid cysts, neuroblastoma

Juvenile xanthogranuloma


Tumor
A circumscribed, elevated, solid lesion with depth, greater than 2 cm in size. Examples: Hemangioma, Spina, rhabdomyosarcoma

Hemangioma

Vesicle
A circumscribed, elevated, fluid-filled lesion up to 1 cm in size.
Examples: Herpes simplex, varicella, miliaria crystallina

Bulla
A circumscribed, elevated, fluid-filled lesion greater than 1 cm in size.
Examples: Sucking blisters, epidermolysis bullosa, bullous impetigo

Wheal
A circumscribed, elevated, edematous, often evanescent lesion, caused by accumulation of fluid within the dermis.
Examples: Urticaria, bite reactions, drug eruptions

Pustule
A circumscribed, elevated lesion filled with purulent fluid, less than 1 cm in size.
Examples: Transient neonatal pustular melanosis, erythema toxicum neonatorum, infantile acropustulosis

Abscess
A circumscribed, elevated lesion filled with purulent fluid, greater than 1 cm in size.
Example: Pyoderma

Secondary Lesions
Secondary lesions are characteristically brought about by modification of primary lesions, either by the individual or through the natural evolution of the lesion in the environment.
**Crust**
Results from dried exudate overlying an impaired epidermis. Can be composed of serum, blood, or pus. Examples: Epidermolysis bullosa, impetigo

**Scale**
Results from increased shedding or accumulation of stratum corneum as a result of abnormal keratinization and exfoliation. Examples: Ichthyosis, postmaturity desquamation, seborrheic dermatitis.

**Erosion**
Intraepithelial loss of epidermis. Heals without scarring. Example: Herpes simplex, certain types of epidermolysis bullosa.

**Ulcer**
Full-thickness loss of the epidermis, with damage into the dermis. Will heal with scarring. Examples: Ulcerated hemangiomas, aplasia cutis congenita

**Fissure**
Linear, often painful break within the skin surface, as a result of excessive xerosis (dry skin). Examples: Inherited keratoderma, hand and foot eczema.

**Lichenification**
Thickening of the epidermis with exaggeration of normal skin markings caused by chronic scratching or rubbing. Examples: Sucking callus, atopic dermatitis
Atrophy

Localized diminution of skin. May be steroid induced. Example: Aplasia cutis congenita, intrauterine scarring and focal dermal hypoplasia.

Focal dermal hypoplasia

Scar

Permanent fibrotic skin changes that develop as a consequence of tissue injury. In utero scarring from infections or amniocentesis or postnatally from variety of external factors. Examples: Congenital varicella, aplasia cutis congenita.