Congenital Nevi: When to Worry?

Elizabeth I. McBurney, MD, FACP
Clinical Professor of Dermatology
LSU School of Medicine
Tulane Health Science Center
Private Practice, Lafayette, LA
Objectives

- To identify congenital nevi at risk for melanoma
- When to order MRIs
- Learn appropriate management of congenital nevi
- What to tell parents
Congenital Nevi

Present at Birth

- Small  < 1.5cm
- Medium  1.5cm – 19.9cm
- Giant  > 20cm
- 9cm on scalp, 6cm on trunk in newborns
- Garment  > 40cm
CONGENITAL NEVUS
LT. HIP
04-29-2008
Natural History of Congenital Melanocytic Nevi

- Darker or lighter color
- Increase in overall thickness
- Changes in topography
Management of Children with Giant Congenital Nevi

Surgical Excision vs. Long Term clinical exams

- Consider surgical excision – wait 6 months of age to avoid period of highest anesthesia risk
- If no excision, life long examination of nevus each 6-12 months plus photography
- Biopsy enlarging, firm papulonodules and new areas of induration or ulceration
Neurocutaneous Melanosis (NCM)

Risk Factors:

- Multiple satellite lesions
- Giant/Garment Nevus
  (>40cm projected adult size)
- Posterior axial location
When to do MRI of Head and Spine for Detection of Neurocutaneous Melanosis (NCM)

- Asymptomatic patients with at least one risk factor
- Asymptomatic patients with intermediate risk (< 10-20 satellite lesions, 20-40 cm as adult)
- Symptomatic patients – signs and symptoms of increased intracranial pressure
Risk of Melanoma in Congenital Nevi

Comprehensive review of 14 studies

- Overall risk in all cases: 0.7%
- Risk in large nevi: 2.5%
- Risk in garment nevi: 3.1%
Cutaneous Melanomas in Congenital Nevi

- Small congenital nevi – melanoma develops at dermal-epidermal junction
- Large congenital nevi – two thirds develop in dermis, subcutaneous fat or deeper
Estimated size of nevus at birth necessary to reach 20 cm in full grown adult

<table>
<thead>
<tr>
<th>Location of CMN</th>
<th>Diameter at birth (cm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Head</td>
<td>12</td>
</tr>
<tr>
<td>Hands, feet, torso, forearms, arms, and buttocks</td>
<td>7</td>
</tr>
<tr>
<td>Thighs</td>
<td>5.8</td>
</tr>
<tr>
<td>Legs</td>
<td>6</td>
</tr>
</tbody>
</table>

Leptomeningeal Melanomas

- Arise in patients with neurocutaneous melanosis
- Most commonly involve frontal and temporal lobes
Melanomas Associated with Large/Giant CMN

• Cutaneous melanomas often arise sub-epidermally, making recognition difficult
• Extracutaneous primary sites are relatively common
  - CNS
  - retroperitoneum
• Occasionally, the primary site is not found
Large/Giant CMN – Risk of Melanoma

• Higher risk
  posterior axial location
  large number of satellite nevi
  larger size of CMN (e.g. >50 cm)

• Lower risk
  restricted to an extremity or the head
  satellite nevi themselves
Congenital Nevi

- Surgical excision with histological control of margins is preferable option for almost all that are indicated to be removed
- Laser treatment will decrease nevomelanocytic load but recurrence is the rule
How Does Excision of Large CMN Affect the Risk of Cutaneous Melanoma?

• No clear-cut answer in the literature
  Trend toward lower incidence of melanoma in patients whose nevi were excised. However, the largest nevi (which have a higher melanoma risk) are more likely to be inoperable.

• Although early and "complete" excision is often recommended, it is usually impossible to remove every nevus cell in the lesion
  Location
  Extensive size
  Involvement of deeper structures
Procedures for Cosmetic and Psychological Benefit (*not* to decrease melanoma risk)

- Neonatal curettage
  Takes advantage of a cleavage plane between the upper dermis (where “active” nevus cells are concentrated) and mid dermis in first 2-3 weeks of life.

- Dermabrasion

- Laser resurfacing (e.g. CO2, erbium:YAG)

- Other lasers (e.g. Nd-YAG, ruby, alexandrite)
Congenital Nevus

Q-switched ruby laser 694nm

6 months post treatment

Congenital nevus of left cheek of a 1-month-old infant before treatment (top) and after four treatments with the Q-switched ruby laser (bottom)
Neurocutaneous Melanocytosis

• Symptomatic NCM
  Classically 3-4% of those with high-risk nevi
  Median onset -2 y. median survival of 6 months
  Hydrocephalus, seizures, vomiting
  With localised mass, median onset -9y. And more likely focal sensorimotor deficits

• Asymptomatic NCM
  +MRI in 5-25% of those with high-risk nevi but no neurologic symptoms
  In 5-y. follow-up study, 1 of 10 asymptomatic patients with +MRI went on to develop neurologic symptoms
Small and Medium-Sized CMN: Longitudinal Evaluation

- Baseline photography
- (self) skin examination by patient/parents
  Bring focal changes in color, border or topography to dermatologist’s attention
Longitudinal Evaluation for Life

- Photography
- Bipsy of suspicious areas
- General physical examination yearly
- Education regarding signs of cutaneous melanoma
- Education regarding signs of mass lesions in CNS