A Rare Case of a Giant Congenital Nevus in a Young Adult Filipino Female

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Introduction
Congenital melanocytic nevi (CMN) usually are present at birth or develop shortly thereafter. Lesions are classified as small (< 1.5 cm), medium (1.5-10 cm), and giant CMN (>20 cm). Presence of more than 50 satellite nevi is a significant factor in prognosis. Incidence of giant nevi is estimated at 0.005% and is common in both sexes. Clinically, they present as light brown to black patches or plaques often covered with hair with lesions varying in sizes, ranging from small resembling acquired melanocytic nevi to very large lesions often affecting certain anatomic regions. Larger lesions lead to more difficult treatment, creating a social stigma and deterioration of quality of life.

Case Report
- 21-year-old, Filipino, female from Palawan, Philippines
- With a history of enlarging inborn generalized nevi accompanied by pruritus and heaviness centered on the trunk
- Unremarkable past medical and family histories
- Consulted a plastic reconstructive surgeon referred to Dermatology service
- On PE: (figures 1-3)
  - brown-black, non scaly plaques overlying soft, redundant, and pendulous skin, on the lower half of the trunk, extending to genitals and both thighs with greatest circumference of 58cm
  - depigmented patches on the left lower back with some hairy areas on left thigh
  - generalized hyperpigmented satellite macules, papules and plaques with diameters measuring 0.2 to 3.5 cm, including palms, soles, both ears, oral mucosa and sclera of left eye
  - Other observations: solitary hyperpigmented, soft, non-movable, non-tender mass measuring 8.2x6.0cm on the right knee, and bowing of lower extremities
  - rest of the physical findings were normal
  - several excisional biopsies taken from the face and trunk area
  - Histopathology: Deep Congenital Melanocytic Nevus (figure 4)
  - Referred to Psychiatry, Orthopedics, Ophthalmology, Gynecology, ENT, Internal Medicine and Neurology services for work up
  - Cranial MRI and ancillary procedures all normal, ruling out Neurocutaneous Melanosis
  - Underwent primary Panniculectomy, Thighplasty, Left Labioplasty and excision of multiple nevi of the face with no complications (figure 5-6)
  - Second debulking stage was not done due to financial constraints

Case Discussion
- Giant congenital nevi (GCN) are seen in 1:500,000 newborns
- Caused by spontaneous mutations during fetal development
- May also be genetically inherited
- Risk of development of malignant melanoma is 6%
  - by 2 years old, the risk of developing into melanoma is 50% and increases to 80% by 7 years of age
  - Increased risk of malignancy: larger nevi (>50 cm), axial location (trunk, head and neck), multiple satellite lesions and the existence of nodules; all of which were seen in our patient
  - Baseline MRI should be done within 4-6 months but our patient underwent MRI as an adult
  - Results were negative, ruling out Neurocutaneous Melanosis
  - Yearly examinations for the first 3 years of life are recommended, with reassessment every 2 to 5 years
  - Surgical treatment of giant CMN can be addressed as early as the age of 6 months
  - Due to the depth of some lesions, especially if the leptomeninges are involved, excisions may not totally eliminate the risk for developing melanoma
  - CO2 laser, Er:YAG and Q-switched ruby laser have all been recently used for resurfacing and for selectively removing the lesions
  - Family support is essential for managing psychological distress

Conclusion
- A rare case of a Giant Congenital Nevus in a 21-year old female from Palawan, Philippines is presented. Dermatopathology showed deep congenital melanocytic nevus. Patient underwent primary reconstructive surgery and multiple nevus excision with no complications. A holistic approach is recommended involving a multidisciplinary approach to improve the patient’s quality of life.