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Picture of the Month

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HYPERPIGMENTED SKIN lesions were present at birth on the infants and children pictured below (**Figure 1**, **Figure 2**, **Figure 3**, and **Figure 4**). Findings from the physical examinations were other-

wise unremarkable. There was no family history of similar findings.

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Figure 1.



Figure 3.



Figure 2.



Figure 4.

Denouement and Discussion

Congenital Pigmented (Nevocellular) Nevi

Figure 1. A medium-sized congenital pigmented nevus on the right cheek.

Figure 2. A giant hairy nevus involving the scalp, neck, shoulders, and upper back.

Figure 3. A "bathing trunk" nevus with irregular hyperpigmentation.

Figure 4. A giant hairy nevus of the left posterior shoulder area.

CLINICAL AND PATHOLOGIC FEATURES

Congenital pigmented (nevocellular) nevi are present in approximately 1% of newborns.¹ The incidence in black infants is higher, 1.8%.² For descriptive purposes, congenital nevi have been arbitrarily classified into 3 categories based on size: small <1.5 cm; medium, 1.5 to 20 cm; and, giant nevi, >20 cm. The congenital nevi that are small to medium typically appear as tan or brown irregularly shaped macules. Over time, the nevi may become elevated and intensely pigmented, sometimes covered with coarse dark hair.³ Small to medium nevi frequently darken at puberty, with pregnancy, or with the use of oral contraceptives.⁴

Giant congenital nevi, or giant hairy nevi, occur in 1 in 20 000 live births.^{4,5} They have an irregular margin often with a verrucous surface. Their color is typically dark brown to black and >95% of them have dark, coarse surface hair. Satellite lesions are often present beyond the periphery of the main lesion and may be scattered over the entire skin surface. Around age 10 years, the giant nevus becomes more elevated, verrucous, and hyperkeratotic and the surface hair thicker.⁶

Giant hairy nevi on the scalp and neck may be associated with leptomeningeal melanocytosis and neurologic disorders that include epilepsy or focal neurologic abnormalities. Lesions over the vertebral column may be associated with spina bifida or meningocele.³ Infants with lesions on the scalp or posterior midline should undergo neuroimaging studies to detect associated conditions that may affect treatment and prognosis.^{7,8}

RISK FOR MALIGNANT CHANGE

Congenital pigmented nevi have a predisposition to undergo malignant change. The size of the lesion correlates with the potential for malignant transformation. The risk of malignant melanoma in patients with small to medium ones ranges from 2.6% to 4.9%, while for giant nevi the risk is felt to be approximately 6%.⁹ Small congenital nevi pose a much greater practical risk since they occur more frequently than giant nevi. Malignant change of small nevi usually occurs after puberty, whereas 60% of all malignant melanomas arising from giant nevi will develop in the first decade of life.⁹ In contrast to the greater frequency of congenital nevi in blacks, the frequency of melanoma in blacks is lower than in whites by a factor of 10 to 20.²

MANAGEMENT AND PROGNOSIS

The management of congenital pigmented nevi remains controversial; no absolute guidelines can be recommended. Management is individualized and centers around: cosmetic appearance, risk of neoplastic proliferation, and the psychological impact on the patient and family.

Because it is impractical to prophylactically excise all non-giant congenital nevi, yearly examination for the first 3 years of life is recommended, with reassessment every 2- to 5 years afterward depending on the confidence of the parents to monitor the lesion(s).⁶ Biopsy specimens are obtained from lesions that undergo suspicious alteration.⁷

The impact of giant hairy nevi is greater because of the considerable cosmetic disfiguration as well as the higher malignant potential. It is more difficult to recognize early malignant changes within giant pigmented nevi because of their typical irregular surface and pigmentation. Some have recommended total surgical excision as early as technically practical, while others are more cautious because of the large areas involved and the resulting disfigurement and complications from scarring. Due to the depth of some lesions, especially if the leptomeninges are involved, excision may not eliminate the risk for developing melanoma.⁷ In a prospective study of 92 patients with large lesions, all 3 who subsequently developed melanomas did so in extracutaneous sites.¹⁰

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