Congenital and Infantile Benign Skin Lesions Affecting the Hand and Upper Extremity, Part 2: Nonvascular Neoplasms

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This article is part 2 of a 2-part series presented to aid the hand surgeon in becoming familiar with dermatological lesions that may be present on the upper extremity during infancy. The discussion focuses on nonvascular neoplasms grouped into the following categories: epithelial, melanocytic, histiocytic, dermal, fibroblastic, and adipocytic neoplasms. Diagnostic tips are offered, including clinical photographs, to help differentiate between these lesions. In addition, the recommended treatment for each is discussed. *(J Hand Surg 2013;38A:2284–2292. Copyright © 2013 by the American Society for Surgery of the Hand. All rights reserved.)*

Key words Nevus, melanocytic, fibroma, hamartoma, tumor.

Skin lesions are common on the hands and upper extremities of children and infants. These lesions can be broadly categorized as benign or malignant as well as vascular or nonvascular. This article focuses on benign nonvascular neoplasms. Although many of these lesions occur in isolation, some may be associated with a syndrome.

EPITHELIAL NEOPLASMS

Epidermal nevus

Epidermal nevi present at birth as tan or flesh-colored patches or plaques in a linear distribution, which often follow Blaschko lines (skin lines representing embryonic migration of cells). They can affect any part of the body, including the distal extremities, and are generally asymptomatic (Fig. 1). On hands, "divided" or "kissing" epidermal nevi can involve opposing portions of neighboring digits.[1] Thickening and darkening of the lesion typically occurs around puberty, resulting in a verrucous plaque that does not resolve spontaneously.[2]

Epidermal nevi may occur in isolation or in association with a variety of syndromes, including epidermal nevus syndrome (epidermal nevi and systemic features affecting numerous organs) (Figs. 2, 3), Proteus syndrome, SOLAMEN (segmental overgrowth, lipomatosis, arteriovenous malformation, and epidermal nevi) syndrome, CLOVE (congenital lipomatous overgrowth, vascular malformations, and epidermal nevi) syndrome, and others.[2]

On histopathology, hyperkeratosis and papillomatosis can be seen. These features are nonspecific and can additionally be seen in acquired lesions such as seborrheic keratoses. Features of epidermolytic hyperkeratosis (EHK) can be seen and, if present, may indicate a risk of transmitting widespread cutaneous EHK to the patient's offspring.[2] A fibroblast growth factor receptor 3 (FGFR3) mutation may be identified in up to 30% of keratinocyte-derived epidermal nevi as well as acquired seborrheic keratoses. There is a very low risk of malignant transformation.[2]

Nonoperative treatment may not be curative because topical retinoids, curettage, and cryotherapy may help initially, but lesions will generally recur. Excision, when feasible, is curative. Carbon dioxide (CO_2) laser treatment may be helpful but carries a risk of scarring.[2]

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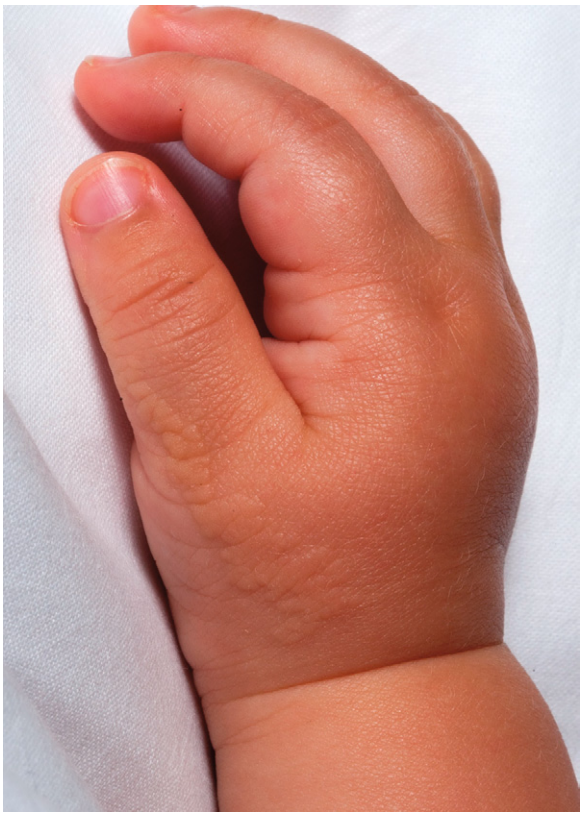

FIGURE 1: Epidermal nevus presents as a tan plaque in a linear distribution on the right thumb. (By permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

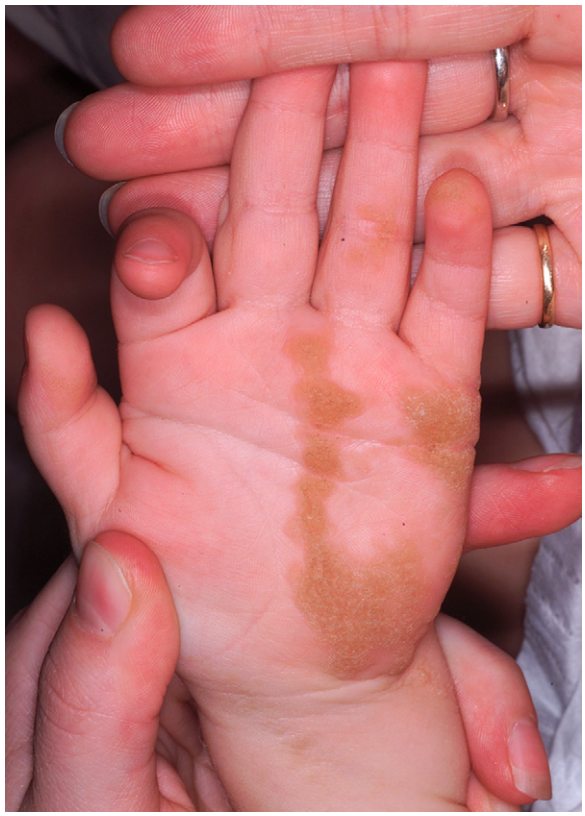

FIGURE 2: Epidermal nevus on the left palm presents as a linear, verrucous, hyperkeratotic plaque, in the setting of epidermal nevus syndrome. (By permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

Porokeratotic eccrine ostial dermal duct nevus

Porokeratotic eccrine ostial dermal duct nevus (PEODDN) is a rare congenital hamartoma that presents as punctuate pits with comedo-like plugs that form hyperkeratotic linear plaques on the distal extremities, particularly the palms and soles. Generally, these are asymptomatic but may be associated with pruritus or localized hyperhidrosis overlying the lesion. PEODDN affects male and female infants equally. Etiology is unknown; however, PEODDN is not associated with other congenital anomalies.[3]

On histopathology, PEODDN is characterized by epidermal invaginations containing parakeratotic columns of stratum corneum overlying eccrine ducts with dilated acrosyringium. Treatment is often unsatisfying; various ineffective therapies reported include topical agents such as steroids, tar, ultraviolet B (UVB), psoralen with ultraviolet A (PUVA), and anthralin. Retinoids or keratolytics may be beneficial.[3] Use of ultrapulsed CO_2 or erbium/CO_2 laser has been reported, but both require numerous sessions and lesions may recur.[4,5] Photodynamic therapy has been reported to have some efficacy after 2 sessions in 1 patient.[6] Localized lesions may be excised. Rarely, squamous cell carcinoma may develop within PEODDN; thus, regular monitoring is advised.[4]

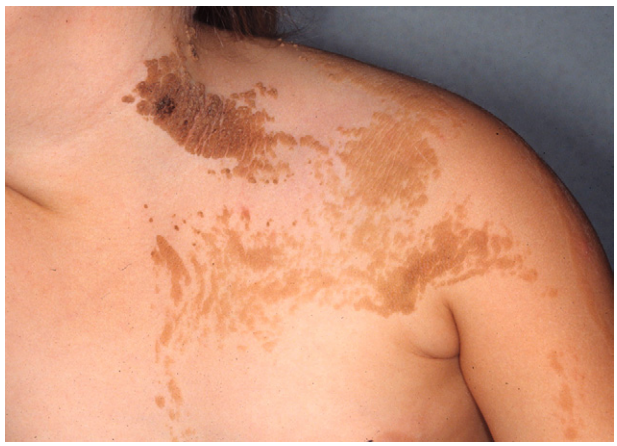

FIGURE 3: Epidermal nevus syndrome demonstrates a whorled (Blaschkoid) morphology on the left shoulder region. (By permission of Mayo Foundation for Medical Education and Research. All rights reserved.)