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Alvin H. Jacobs, MD

MELANOCYTIC NEVI

Acquired Moles

The pigmented melanocytic nevus, or mole, represents an hereditary malformation of melanocyte-directed embryonic neuroectodermal cells arrested at the dermoepidermal junction or below. These cells, designated "nevus cells," aggregate in groups or nests, the predominant site of which determines their histologic classification (junctional, compound, or intradermal).

The so-called acquired melanocytic nevi are not present at birth. They usually develop between the second and sixth year and may appear in crops at puberty. The latter phenomenon may occur in association with darkening of previously existing melanocytic nevi. They may occur on any body surface including nail beds and mucous membranes (Fig 1). There is no greater significance to palmar, plantar, genital, or mucous membrane lesions in terms of malignant potential. Similarly, the histologic type of nevus bears little significance regarding malignant degeneration except that junctional and compound nevi are more likely to become melanomas than are intradermal moles. The individual risk of eventual malignant transformation in acquired melanocytic nevi is extremely low when certain facts are considered: (1) The per capita average number of pigmented nevi is between 20 and 30. (2) The incidence of melanoma is truly rare in childhood (but rises appreciably after puberty). In all age groups approximately 9,300 cases occur per year in the USA. (3) Less than 50% of melanomas arise in preexisting melanocytic nevi. Giant congenital melanocytic nevi show a significantly increased potential for malignant degeneration (see below).

Excisional biopsy and histologic examination of acquired melanocytic nevi is rarely indicated in the pediatric patient. Suspected malignancy would be the sole motivating factor for such a procedure. Clues to malignant transformation of acquired melanocytic nevi include the following: (1) rapid growth associated with wide diffusion of pigment with irregular margins; (2) irregular darkening of pigmental lesions associated with bluish discoloration; (3) partial loss of pigment associated with apparent spontaneous scarring; (4) erythematous halo in the absence of trauma; (5) spontaneous bleeding or increased fragility; and (6) development of nodular components. It should be emphasized that the presence of hairs within a pigmented lesion gives no assurance of its being benign. When indicated, biopsy should be done by excisional surgery although wide excision can await the results of the biopsy.

Halo nevus is the term designating an otherwise ordinary intradermal or compound nevus around which a collar of depigmentation develops (Fig 2). The process is seen most commonly occurring on the trunk in the second decade of life. The centrally located nevus usually disappears as the depigmentation advances. The normal pigmentation eventually returns to the area in most cases when the process is complete. Halo nevi may be multiple or occur as successive events. Treatment of halo nevus is rarely indicated (Fig 3).

Congenital Pigmented Nevi

A great deal of confusion exists regarding the definition and significance of congenital pigmented nevi. When confronted with any large pigmented lesion in a pediatric patient, the single most important determination is whether or not the lesion was present at birth. Frequently,
parents describe a nevus as being "present since birth," but on further questioning it becomes evident that the lesion developed later. A truly congenital nevus should have its history dating to the actual time of birth. This information is of greatest importance when dealing with a relatively small or medium sized nevus (Fig 4) which may not have all the clinical characteristics of the giant nevus which commonly covers an entire anatomical region (garment nevus) (Fig 5). In general, congenital moles tend to be larger than the acquired type. The very large (giant) lesions are so remarkable that no confusion exists regarding their origin. However, no consensus exists regarding what physical dimensions constitute a "giant" congenital melanocytic nevus. Therefore, it is probably more appropriate to ignore size alone and allow therapy to be guided by whether the lesion was congenital or acquired. This may seem to be a minor point, but the incidence of malignant melanoma arising in congenital melanocytic nevi may approach 10%. Decision concerning excision of the congenital lesions cannot be delayed, since 60% of the melanomas that have occurred in congenital nevi have occurred in the first decade of life. In contrast with the 10% malignancy potential of congenital nevi, acquired pigmented moles have only about one in a million chance of developing malignancy.

Fortunately, the garment-like congenital melanocytic nevus is rare. They are dark brown to black in color with an uneven papillomatous surface and irregular margins frequently exhibiting smaller satellite nevi at the periphery (Fig 6). Although many such lesions have a hairy component that consists of large, coarse terminal hairs, the presence or absence of hair has no bearing on the malignancy potential. Treatment is directed toward total excision and grafting, when necessary, to cover the resultant defect. This eliminates the malignant potential and serves to partially ameliorate the adverse cosmetic impact.

Café-au-Lait Spots

The birthmark with the greatest array of possible significant associations is the café-au-lait spot. They usually are present at birth or appear in early childhood as light tan macules of varying size and shape (Fig 7). About 10% of normal individuals have café-au-lait spots, but when six or more spots are present, especially if they measure 1.5 cm or greater, there is a strong possibility that the patient has, or will develop, neurofibromatosis (Fig 8). Café-au-lait spots are usually the first, and one of the best cutaneous signs of neurofibromatosis, occurring in about 90% of cases (Fig 9).

Similar pigmented lesions occur in Albright's syndrome but they have a more irregular outline ("coast of Maine") and more limited distribution on the trunk, buttocks, and thighs (Fig 10). There may be unilateral localization favoring the side of the greatest bony involvement.

Café-au-lait spots are also associated with other syndromes and visceral diseases. These include: (1) tuberous sclerosis, (2) pulmonary stenosis, and (3) temporal lobe dysrhythmias.

EPIDERMAL NEVI

The final group of embryonic errors producing cutaneous defects are epidermal nevi. This is often a confusing group of lesions owing to the descriptive terminology selected to designate these aggregates of epithelial tissue derived from epidermis or epidermal appendages (i.e., apocrine, eccrine, or sebaceous glands, or hair follicle elements). Individual
Fig 4. Relatively small congenital pigmented nevus.

Fig 5. This giant congenital nevus or "garment" nevus is partly hairy. Malignant degeneration could occur in either the hairy or non-hairy portion. Removal of this giant nevus presents a problem to the plastic surgeon, but removal should be carried out.

Fig 6. Giant nevus with multiple satellite nevi.

Fig 7. Large, perianal café-au-lait spot.

Fig 8. More than six café-au-lait spots indicate that this infant will undoubtedly develop other manifestations of neurofibromatosis.

Fig 9. This young man with multiple café-au-lait spots has developed several small neurofibromas.

Fig 10. This child with a large unilateral "coast of Maine" café-au-lait spot and premature puberty has Albright's syndrome.
lesions show mixed histologic components, but one basic element usually predominates, thereby determining the name. The most common of these hamartomas are the verrucous nevus and the sebaceous nevus.

Verrucous Nevi

The verrucous nevus is either congenital or appears in early infancy on any skin surface. They tend to be unilateral and there is no genetic or sex predilection. The initial appearance is that of a flesh-colored to yellow-brown, rough, warty, oval or linear plaque. Frequently, a multiplicity of tiny individual warty papules form the familiar linear streaks seen on the extremities or the horizontal or gyrate configurations seen on the trunk (Figs 11 and 12). If the lesions are extensive and involve intertriginous sites, the surface may become macerated and foul smelling.

Treatment is for cosmetic reasons. Operative therapy usually should not be undertaken until after puberty when the lesion has attained its maximum size. Excision is preferable for small lesions or moderate size linear ones. Curettage with electrodessication or dermabrasion can be helpful, but recurrences are frequent. Bowen’s disease and squamous cell carcinoma have been reported in the preexisting epithelial nevi in only the rarest of circumstances.

Sebaceous Nevi

The sebaceous nevus is frequently present at birth or develops within the first year. The lesion is most commonly encountered on the face or scalp as an isolated elevated waxy orange to orange-brown plaque with a granular surface (Fig 13). They may reach up to 10 cm in greatest dimension and are devoid of hair (Fig 14). They evolve through childhood with slow growth, and at puberty they become thickened and develop a multiplicity of relatively uniform papillomatous projections from the surface.

Total surgical excision is the treatment of choice since basal cell carcinoma may eventuate in many cases. Treatment may be delayed, however, since such degeneration usually occurs in middle life.

Epidemial Nevus Syndrome

Solomon and associates have described a relatively rare syndrome in which an extensive epidermal and/or sebaceous nevus is associated with multiple skeletal and neurologic abnormalities.

SUMMARY

— Acquired moles—very rare malignancy during childhood
— Congenital moles—10% chance of malignancy
— Café-au-lait spots—associated with neurofibromatosis, Albright’s syndrome, tuberous sclerosis, pulmonary hypertension, and temporal lobe dysrhythmias

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