

# Congenital Café-au-lait Spot and Sequentially Occurred Nevus Spilus

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There has been one case reported that café-au-lait spot, nevus spilus and melanocytic nevi following Blaschko's lines concurrently occurred at birth.

An 11-year-old girl had an uneven, oval, uniformly light brown colored patch on the right side of the trunk since birth. Later a light brown, large patch based marking with numerous scattered darker brown macules progressively developed along the upper trunk and the medial side of right arm following Blaschko's lines. She had not had any other skin abnormality, underlying disease, malformation associated with neurofibromatosis or familial history. Histological examination taken at the light brown colored macule on the trunk revealed basal hyperpigmentation with rete ridge elongation and no dermal melanophages, compatible with café-au-lait spot<sup>1</sup>.

Hereby we report an unusual case of congenital café-au-lait spot and nevus spilus which sequentially occurred in later life by linear unilateral distribution, following Blaschko's lines. (*Ann Dermatol* 16(4) 191~193, 2004)

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**Key Words:** Blaschko's lines, Café-au-lait spot, Nevus spilus

Nevus spilus (speckled lentiginous nevus) presents as a circumscribed, usually lightly pigmented patch containing scattered, usually more smaller, darker pigmented spots<sup>2</sup>. Café-au-lait spot is discrete, uniformly tan or light brown, well circumscribed, less than one to several centimeters large patch<sup>2</sup>. This is an interesting case of congenital café-au-lait spot and sequentially developed nevus spilus following Blaschko's lines which reflect the migratory paths of melanoblast clones.

## CASE REPORT

An 11-year-old girl had an uneven, oval, uni-

formly light brown colored patch on the right side of the trunk since birth and later a light brown large patch based mark with numerous scattered darker brown macules progressively developed along the upper trunk and the medial side of right arm by linear appearance (Fig. 1). She had not any other skin abnormality, underlying disease, malformation associated with neurofibromatosis or familial history.

Histological examination taken at the light brown colored macule on the trunk revealed basal hyperpigmentation with rete ridge elongation and no dermal melanophages, compatible with café-au-lait spot (Fig. 2). We didn't perform further diagnostic procedures. By clinical and histological finding, we diagnosed it congenital café-au-lait spot and sequentially occurred nevus spilus.

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## DISCUSSION

According to the previous report<sup>3</sup>, some melanocytic nevi have appeared in association with several congenital syndromes and certain cases which have only skin lesions, not associated with congenital syndrome, appeared by segmental and asymmetric,



**Fig. 1.** Café-au-lait spot and nevus spilus on the upper trunk and right arm alongside Blaschko's lines.



**Fig. 2.** A basal hyperpigmentation with rete ridge elongation compatible with café-au-lait spot (H&E stain,  $\times 100$ ).

by Blaschko's lines<sup>4</sup> or by trigeminal dermatome<sup>5</sup>. However, these cases are lentiginosis or have some histologic finding of junctional nevus, not café-au-lait spot nor nevus spilus.

In addition, one case has been reported that café-au-lait spot, nevus spilus and melanocytic nevi aggregated alongside Blaschko's lines concurrently occurred at birth<sup>6</sup>. The patient had speckled light brown pigmented areas with multiple darker brown macules (nevi spili) characteristically distributed over the left side of his face, cervical region, chest and arm. Clinically, in addition to the large nevi spili described, café-au-lait spots on the face and smaller ones on the arm were present, and multiple flat light to dark brown plaques could be detected. Histological examination at the brown plaques revealed that

normal or papillomatous epidermis, nests of melanocytic cells without cytological atypia in the dermis. Melanocytic cells were also found more deeply around appendages and neurovascular bundles, indicating congenital melanocytic nevi. In our case, it is similar with the previous case that she has only skin lesions and isn't associated with any congenital syndrome or familial history. However, it is different in the fact that the café-au-lait spot at the trunk is congenital but the nevus spilus on the right arm and upper chest was newly developed and became larger in later life following Blaschko's lines.

Nevus spilus may be postulated to represent a localized defect in neural crest melanoblasts and becomes evident during infancy or early childhood rather than at birth<sup>2</sup>. Although the exact nature of Blaschko's lines is unknown, there is a general consensus that they reflect the lines of embryonal development of the epidermis and epidermal derivatives, including melanoblast clones, and determine the distribution pattern of many congenital and acquired skin diseases<sup>7</sup>. The authors of previous cases explained that the distribution of the pigmented lesions alongside Blaschko's lines may suggest a clonal outgrowth of cells carrying an altered gene that is responsible for the development of various melanocytic lesions. This clone might originate from a somatic mutation occurring at an early stage of embryogenesis, when neural structures have already been formed<sup>6</sup>. Hence the development of the acquired nevus spilus along Blaschko's lines in our case can be thought of as a multistep process involving a primary genetic defect coupled with a secondary intrinsically (e.g. growth factor) or extrinsically (e.g. UV light) mediated event leading to the development of the lesion<sup>2</sup>.

Therefore, hereby we report an unusual case of congenital café-au-lait spot and nevus spilus which sequentially occurred in later life by linear unilateral distribution, following Blaschko's lines.

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