

Naevus Lentiginosus Linearis: A Distinct Skin Disorder

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Sir,

Congenital naevi of the melanocytic system include numerous types, which differ in their clinical appearance, pattern of distribution, and histopathological features (1). Examples are large congenital melanocytic naevus, macular naevus spilus, papular naevus spilus, café-au-lait macules of neurofibromatosis 1, café-au-lait macules arranged in broad bands as noted in McCune-Albright syndrome, partial unilateral lentiginosis, naevus achromicus (naevus depigmentosus), phylloid hypermelanosis, and phylloid hypomelanosis (1–3).

We describe here two patients with a systematized pigmentary naevus that differed from all naevi reported so far.

CASE REPORTS

Case 1

A 13-year-old girl presented with a pigmentary naevus present since birth. Her family history was non-contributory. Several narrow bands following Blaschko's lines involved the right side of her body with a strict midline separation (Fig. 1 A, B). These lesions consisted of multiple tightly packed lentigines. On careful palpation, no papules could be found. The background of the lentigines was neither hyper- nor hypopigmented. In addition to the linear pigmentary lesions, the girl had mild, symmetrically distributed freckling of the forehead, the nose and the cheek. No extracutaneous anomalies were noted. In particular, neurological and ophthalmological examinations gave normal results.

Histopathological examination of two specimens obtained from the lentiginous lesions showed pronounced hyperpigmentation of the basal cell layer (Fig. 2A).

Case 2

A 36-year-old man presented with a congenital linear pigmentary disturbance involving his face. A band consisting of tightly packed lentigines was running in a reverse S figure from the left retroauricular region to the eye (Fig. 1C). No other member of his family was known to have a similar skin lesion. The patient had no associated extracutaneous anomalies.

On histopathological examination, the basal and suprabasal keratinocytes were found to be hyperpigmented. In some specimens the amount of melanocytes was normal (Fig. 2B), whereas in other sections their number was definitely increased, especially in the apical regions of rete pegs (Fig. 2C).

DISCUSSION

These patients had an unusual congenital pigmentary disorder that, to our knowledge, did not correspond to any of the various types of pigmentary naevi known so far (Table I). In particular, the tightly packed lentigines did not show any abnormal background in the form of hyper- or hypo-pigmentation.

The disorder follows Blaschko's lines and therefore clearly reflects cutaneous mosaicism (3). The condition differs from naevus spilus maculosus (2) by the absence of background hyperpigmentation and by the

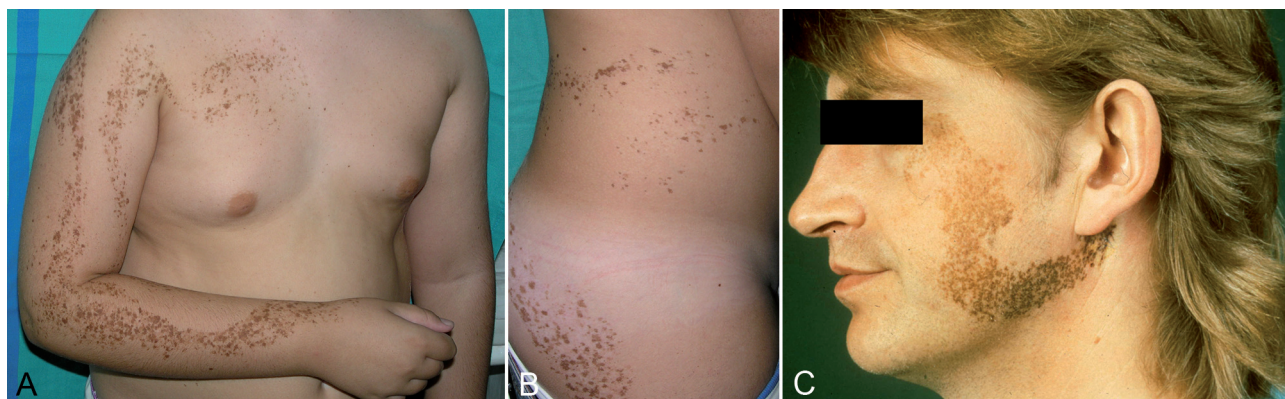


Fig. 1. Tightly packed lentigines distributed in a band-like pattern involving: (A) the right side of the thorax and the right arm; (B) the right flank (case 1); and (C) tightly packed lentigines arranged in a linear pattern (case 2).

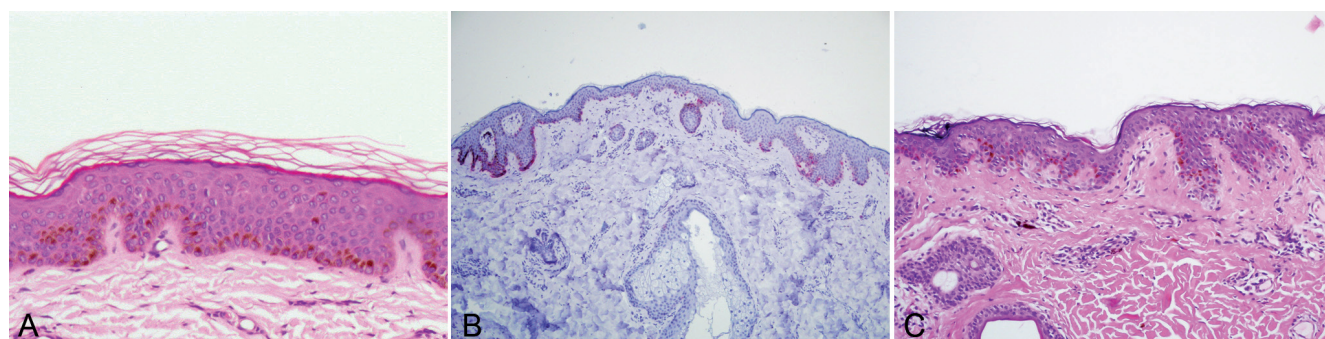


Fig. 2. (A) Pronounced hyperpigmentation of basal and suprabasal keratinocytes (haematoxylin and eosin (H&E) staining; case 1). (B) A lentigo specimen of case 2 showing normal numbers of melanocytes over large areas, stained with the pan-Mel marker recognizing HMB-45 and Mart-1 antigens as well as tyrosinase (immunoperoxidase; antibody from Dako Co., Glostrup, Denmark), whereas in other areas the amount of melanocytes is found to be increased, especially in (C) the apical areas of the rete pegs (H&E staining).

arrangement along Blaschko's lines. The same holds for naevus spilus papulosus that differs, in addition, by the presence of papular speckles (2). Contrasting with the Blaschko-linear arrangement as noted in the present cases, partial unilateral lentiginosis that is also called "agminated lentigines" (1) and that is considered by some authors to represent a mosaic form of neurofibromatosis 1 (4–6) is arranged in a checkerboard pattern (7). Naevus achromicus with secondary lentigines (8) can be excluded in our patients because the lesions did not show any hypopigmentation.

Table I. Distinguishing clinical criteria of different types of naevi associated with lentigines

Disorder	Pattern of distribution ^a	Abnormal background pigmentation
Macular naevus spilus	Checkerboard	Café-au-lait background
Papular naevus spilus	Checkerboard	Café-au-lait background
Partial unilateral lentiginosis (agminated lentigines)	Flag-like or checkerboard	No
Naevus achromicus with secondary lentigines	Flag-like or Blaschko lines	Hypopigmented background
Linear lentiginous naevus	Blaschko lines	No

^aThe terms "zosteriform" or "dermatomal" are misnomers and therefore should be avoided.

In conclusion, the pigmentary disorder described here may represent a new entity, for which we propose the name naevus lentiginosus linearis (linear lentiginous naevus).

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