A Finely Wrinkled, Band-like Naevus Associated with Hyper- and Hypopigmented Spots: a New Entity?

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Sir,

We report here a case of a Japanese man with an unusual congenital band-like naevus, consisting of a finely wrinkled skin area with a mosaic of hyper- and hypopigmented spots involving only the right side of his chest. To our knowledge, there are no previous reports of this presentation of congenital naevus.

CASE REPORT

A 36-year-old Japanese man presented with a band-like pigmented area on only the right side of his chest, which had been present since birth. As there had been no particular change in either its colour or size, he had not consulted a dermatologist previously. There was no family history of a similar skin change.

On his first visit, he had a well-suntanned skin due to his hobby of marine sports, together with a roughly S-shaped, band-like, and mildly pigmented, lesion localized only on the right side of the mid-chest (Fig. 1a). Although the skin texture appeared rough due to the presence of fine wrinkles, the lesions were in fact smooth on palpation. On closer inspection, the mildly pigmented plaque appeared more wrinkled than the surrounding normal skin, presenting a reticular pattern dotted with hyper- and hypopigmented spotty areas (Fig. 1b). The spotty changes were composed of variously sized, darkly hyperpigmented, hypopigmented or almost normal looking, finely wrinkled areas, which did not appear to conform to any of the so-far reported cases of congenital skin lesions. He had no other abnormalities in the skin, hair or teeth, and no clinically observable sexual ambiguity or family history of similar skin changes.

Histologically, in the darkly hyperpigmented spot, we found atrophic epidermis and basal hypermelanosis that was prominently noted at the tips of elongated rete ridges whose covering stratum corneum was somewhat thickened and compact containing scattered melanin granules. Moreover, there were scattered melanophages among a mild mononuclear cell infiltrate at the perivascular portion of the upper dermis. Neither a nest of melanocytes nor sclerosis of the dermis was detectable (Fig. 2a). Other mildly hyperpigmented portions showed thin epidermis, mild basal hypermelanosis and a few melanophages in the upper dermis. On the other hand, the hypopigmented spot revealed a thin epidermis covered by rather thick and compact stratum corneum with no changes suggestive of scar or amyloid deposition in the dermis (Fig. 2b). There were neither hair follicles nor eccrine sweat glands in the specimens. He did not require any further treatment for his skin lesion.

DISCUSSION

To our knowledge, the skin changes found in this patient are unique among the types of congenital naevus described to date. Its clinical feature had been stable since birth, showing a well-defined, band-like, finely wrinkled, mildly pigmentary skin area confined to the right side of the chest. On a closer examination, this consisted of hyper-, hypo-, and even depigmented spotty areas. If there had been any telangiectatic component, we could have described it as poikilodermatous. Although there was no clinical sign of inflammation, histologically we found that the pigmented areas showed mild hyperkeratosis covering an atrophic epidermis associated with focal basal hypermelanosis and pigmentary incontinence, suggestive of the presence of a preceding mild inflammatory event involving the epidermis and upper dermis.

Ardelean & Pope (1) reported a male case of Bloch-Sulzberger syndrome whose skin lesions tended to be more localized, such as unilateral involvement, than that of female cases. But our patient differed, in that there had been neither a remarkable preceding inflammatory stage nor evidence of hereditary occurrence such as that noted in incontinentia pigmenti. However, we cannot completely exclude the presence of a preceding inflammatory stage occurring during the foetal stage. The total lack of skin appendages in the lesions suggested the possibility of such a scar. However, in general, the pigmentation found in incontinentia pigmenti tends to disappear by adolescence.

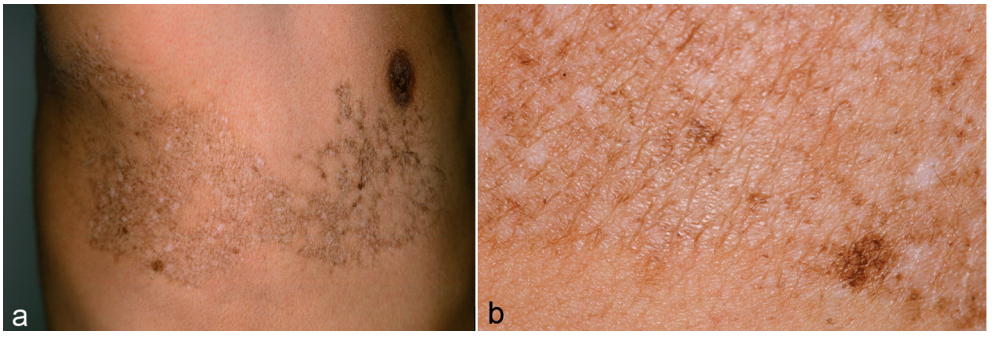

Fig. 1. Band-like, reticular and mosaic-like hyperpigmented lesion located only on the right side of the chest, consisting of reticular and spotty two-coloured hyperpigmentation and spotty hypopigmentation.

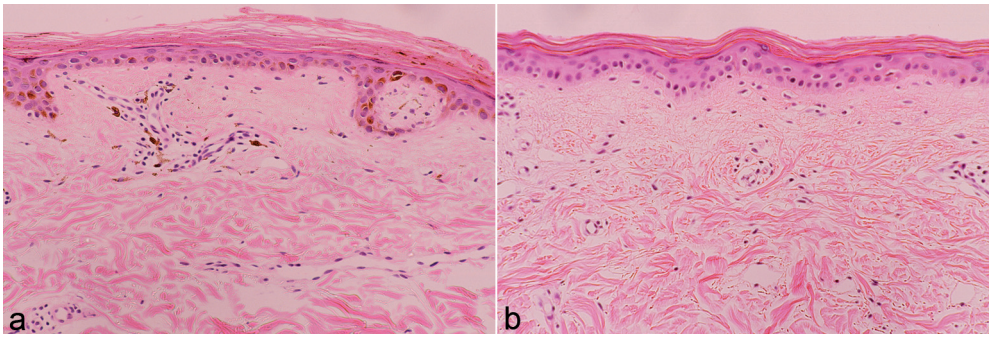

Fig. 2. Histopathology of the biopsy specimen. (a) Atrophic epidermis, basal hypermelanosis and incontinentia pigmenti were seen in the dark hyperpigmented spot. (b) Only a thin epidermis without scarring was observed in the whitish hypopigmented spot. (Haematoxylin and eosin (H&E) stain; original magnification ×40).

Blaschko recognized a common pattern of skin involvement in congenital or acquired linear dermatoses from his observation of 175 patients, and described the clinical and pathophysiological aspects of Blaschko's lines (2). Although we were not completely sure whether the present skin lesion was distributed according to Blaschko's lines, in our differential diagnosis we also considered the possibility of several dermatoses that follow Blaschko's lines, as listed in Table I. However, we could exclude almost all of these due to their clinical features and histopathological changes. In their description of Blaschko's lines, as well as those skin diseases that follow these lines in detail, Bolognia et al. (2) described that the cutaneous lesions that follow Blaschko's lines constitute an important clue to reach a final diagnosis, since they seem to reflect the path of embryonal development of the cutaneous structures. Happle and coworkers (3, 4) hypothesized that mosaicism was responsible for all of the abnormal skin changes developing along Blaschko's lines. Supporting the hypothesis, Taibjee et al. (5) reported that 88% of cases of pigmentary anomalies developing along Blaschko's lines had chromosomal abnormalities, which were overlapped with one or more pigmentary genes. In addition, they speculated that chromosomal abnormalities invisible at the gross karyotypic level, such as microdeletions or point mutations, might be present in other cases (5). There is accumulating evidence indicating that both hypomelanosis of Ito and related disorders, such as linear and whorled nevoid hypermelanosis, are all due to mosaicism of a variety of chromosomal abnormalities, i.e. chimerism, chromosomal mosaicism, somatic mutations, and half-chromatid mutations (3–10). Thus, this group of disorders has been suggested to be "pigmentary mosaicism" in general (5).

In an analysis of 54 children with pigmentary anomalies present along Blaschko's lines, Nehal et al. (6) found that 36 patients (67%) had hypopigmentation, 13 (24%) had hyperpigmentation, and 5 (9%) had a combination of hyperpigmentation and hypopigmentation. In addition, there have been some cases of pigmentary mosaicism with both hyperpigmentation and hypopigmentation. However, all of these reported cases (3–10) displayed hyperpigmentation that was present apart from hypopigmentation in each line or macule, unlike our present patient.

Unfortunately, we could not perform any molecular analysis in this naevus to exclude incontinentia pigmenti. However, to the best of our knowledge, such a clinical feature consisting of four types of pigmentary changes, i.e. a mixed pattern of darkly hyperpigmented, lightly hyperpigmented, hypopigmented, and normally pigmented skin, involving only one localized area has not been reported previously.

Table I. *Similar skin conditions that follow Blaschko's lines reported in the literature to date (1–10)*

Segmental naevus depigmentosus
Incontinentia pigmenti (Bloch-Sulzberger syndrome)
Reticulate hyperpigmentation of Iijima
Linear atrophoderma of Moulin
Linear and whorled nevoid hypermelanosis
Linear scleroderma

REFERENCES

1. Ardelean D, Pope E. Incontinentia pigmenti in boys: a series and review of the literature. Pediatr Dermatol 2006; 23: 523–527.
2. Bolognia JL, Orlow SJ, Glick SA. Lines of Blaschko. J Am Acad Dermatol 1994; 31: 157–190.
3. Happle R. Mosaicism in human skin. Arch Dermatol 1993; 129: 1460–1470.
4. Happle R, Barbi G, Eckert D, Kennerknecht I. "Cutis tricolor": congenital hyper- and hypopigmented macules associated with a sporadic multisystem birth defect: an unusual example of twin spotting? J Med Genet 1997; 34: 676–678.
5. Taibjee SM, Bennett DC, Moss C. Abnormal pigmentation in hypomelanosis of Ito and pigmentary mosaicism: the role of pigmentary genes. Br J Dermatol 2004; 151: 269–282.
6. Nehal KS, PeBenito R, Orlow SJ. Analysis of 54 cases of hypopigmentation and hyperpigmentation along the lines of Blaschko. Arch Dermatol 1996; 132: 1167–1170.
7. Ohashi H, Tsukahara M, Murano I, Naritomi K, Nishioka K, Miyake S, et al. Pigmentary dysplasias and chromosomal mosaicism: report of 9 cases. Am J Med Genet 1992; 43: 716–721.
8. Niessen RC, Jonkman MF, Muis N, Hordijk R, van Essen AJ. Pigmentary mosaicism following the lines of Blaschko in a girl with a double aneuploidy mosaicism: (47,XX,+7/45,X). Am J Med Genet 2005; 137A: 313–322.
9. Lipsker D, Flory E, Wiesel ML, Hanau D, de la Salle H. Between light and dark, the chimera comes out. Arch Dermatol 2008; 144: 327–330.
10. Loomis CA. Linear hypopigmentation and hyperpigmentation, including mosaicism. Semin Cutan Med Surg 1997; 16: 44–53.