Dots and Lines: A Dermoscopic Sign of Regression of Longitudinal Melanonychia in Children

Yozo Murata, MD; Kimiko Kumano, MD

It is not easy to predict the clinical course of longitudinal melanonychia (LM) in children because few prospective studies have been conducted. In our prospective study 15 Japanese children with LM were followed for more than 2 years. Eight patients demonstrated gradual fading of LM. Clinical features such as the patient's sex and the site, age of onset, duration, color, and width of the melanonychia were not significantly associated with the outcome. Dots distributed along melanotic lines, a finding we referred to as dots and lines, can be a dermoscopic sign of regression of melanonychia in children with LM. In this study, the presence of dermoscopically observed dots was significantly related with regression of melanonychia (P=.019; odds ratio, 18.0).

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The etiology of longitudinal melanonychia (LM) in children has been debated. Histopathologically, benign melanocytic hyperplasia (ie, lentigines, nevi) is the cause of 77.5% of cases of pediatric LM.[1] The progression of nail matrix nevi to melanoma is rare[2]; however, 2 children with malignant melanoma of the nails presenting with LM were reported in 2008.[3]

Clinically, Hutchinson sign with variation in the width and color of the pigmented band is believed to be alarming; however, because these findings are not unusual features of nail matrix nevi in children,[2] biopsy procedures, which have potential adverse effects, should not be conducted in all pediatric patients. Dermoscopic patterns for evaluation of nail pigmentation have been described, but their accuracy in the diagnosis of subungual melanoma has not been well-established.[2] Because gradual fading of LM is common in children, careful observation of the clinical course is recommended.[2,4,5]

We prospectively studied 15 Japanese children 13 years or younger with LM. They had been followed for more than 2 years to evaluate clinical features and dermoscopic findings of dots and lines to determine if dots and lines were related to future regression of LM.

Methods

All patients 13 years or younger who had developed LM before 10 years of age and had presented for treatment between December 1998 and January 2008 were included in the study. Clinical features were recorded such as the patient's sex and the site, onset, duration, color, and width of the melanonychia. Dermoscopic findings of pigmented dots distributed along melanotic lines, referred to as dots and lines, also were noted. All patients had been followed for more than 2 years without treatment or nail biopsy. For statistical analysis, the odds for regression of LM were compared with clinical factors using a χ^2 test to calculate *P* values ($\alpha=.05$) and odds ratios.

Results

The study included 9 boys and 6 girls, with the age at presentation ranging from 1 to 13 years (mean, 7 years). The age at initial onset of melanonychia ranged from 3 months to 10 years. Among these patients, a total of 10 fingernails and 5 toenails were affected, with the thumb and great toe being the most common sites of melanonychia (6/15). The width of melanonychia ranged from 1 to 9 mm; some patients

From the Department of Dermatology, Hyogo Cancer Center, Kitaoji-cho, Akashi, Japan.

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Correspondence: Yozo Murata, MD, Department of Dermatology, Hyogo Cancer Center, 13-70, Kitaoji-cho, Akashi, 673-8558, Japan (YZMurata@aol.com).

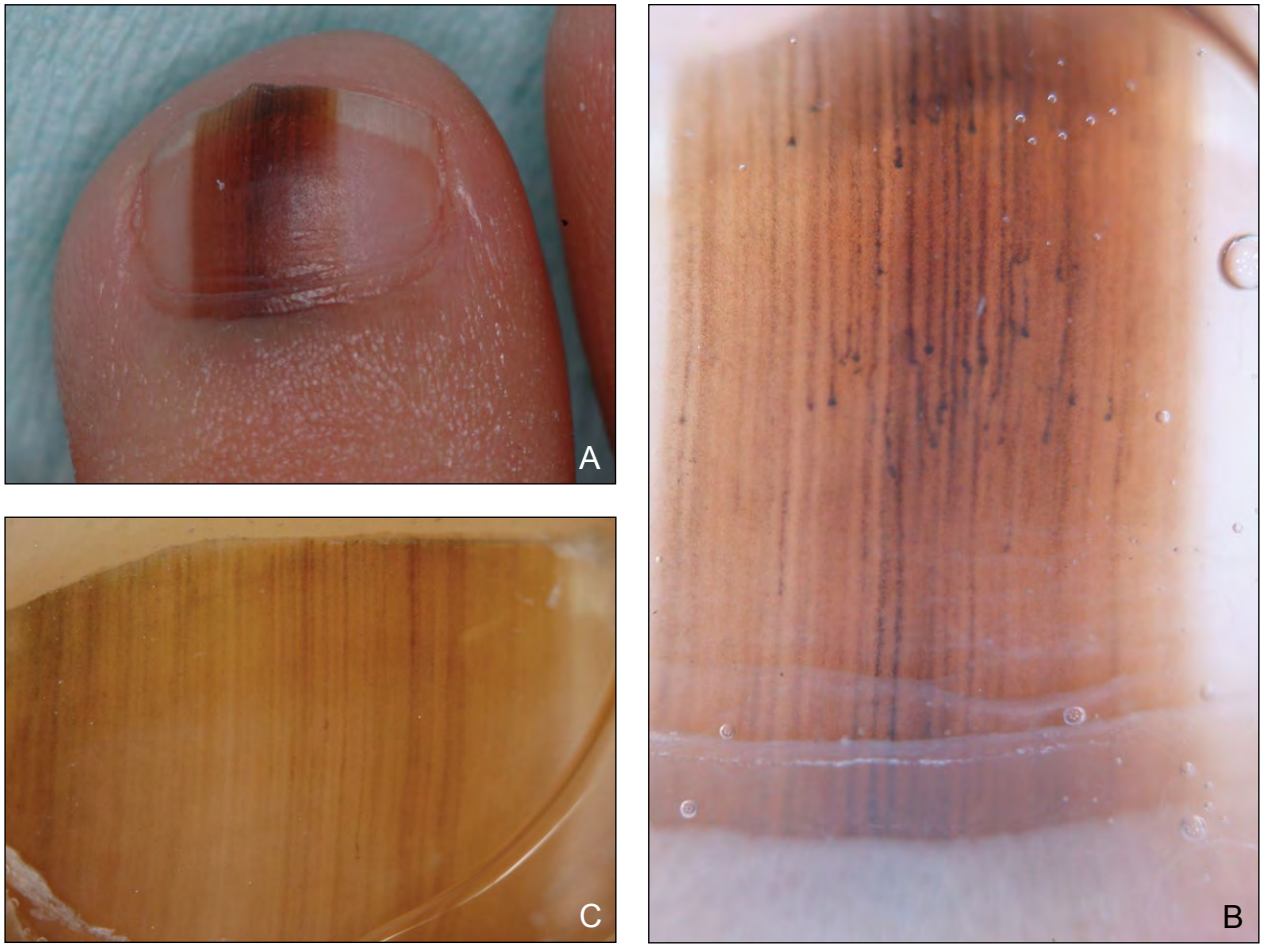

Figure 1. Longitudinal melanonychia of the left third toenail in a 6-year-old girl. Note the dark melanotic band, which was first discovered by the patient's guardians when she was 2 years old (A). Dermoscopy of the nail showed a dots and lines pattern with dots forming a boundary between the darkly pigmented distal line and lighter pigmented proximal line (B). Dermoscopy 30 months after the initial visit showed sparsely distributed light brown lines and only a few remaining dots (C).

showed nails with complete melanonychia. The color of these bands ranged from light brown to dark brown or black.

Dermoscopic examination showed regular melanotic lines in all patients. In some patients, interruption, curving, or branching of the lines was observed. In 7 patients, blackish dots were noted and were similar in size and shape (round to oval). No purple or red color was noted in these dots, and there were no filamentous distal ends. The dots were distributed in a vaguely longitudinal fashion along the pigmented lines. They moved distally with time. In some patients, the dots formed shallow pits rimmed by pigment at the periphery; in other patients, the dots formed the boundary between darkly pigmented distal lines and lighter pigmented proximal lines. Some lines were observed to be suddenly disrupted by the presence of dots.

Patients returned for follow-up visits every 3 to 6 months for a total of 24 to 123 months (mean, 54 months). Eight patients showed gradual fading of LM (2 patients with complete fading; 6 patients with partial fading)(Figure 1) and others remained unchanged. A univariate statistical analysis of 7 clinical factors—sex, site, age at onset, period from onset to consultation, width, color, and presence or absence of dots and lines—was used to detect the factors with a significant effect on the occurrence of spontaneous regression of LM in this study (Table). The first 6 factors in this analysis were not significantly associated with regression of melanonychia. Notably, 6 of 7 patients with dots and lines showed gradual fading of LM, while 2 of 8 patients without dots showed gradual fading. This difference was statistically significant (P=.019; odds ratio, 18.0). Dots disappeared over time in most patients.

Univariate Analysis of Regression of Longitudinal Melanonychia in Children

Clinical Factor	Regression		*P* Value	Odds Ratio
	Observed	Not Observed		
Sex				
Male	5	4	.83	1.3
Female	3	3		
Site				
Fingernails	5	5	.71	0.67
Toenails	3	2		
Age at onset				
≤2 y	4	1	.14	6.0
>2 y	4	6		
Period from onset to consultation				
≤2 y	4	3	.78	1.3
>2 y	4	4		
Width				
≤5 mm	3	4	.45	0.45
>6 mm	5	3		
Color				
Dark	5	4	.83	1.3
Light	3	3		
Dots and lines				
Present	6	1	.019	18.0
Absent	2	6		

All patients who were included in this study continued to grow and develop normally without any signs of disease except for LM; none of the patients developed subsequent destruction of the nail or tumor formation that might be suggestive of malignant changes.

Comment

Dots were observed as small, round to oval, black structures that were similar in size. They were randomly distributed but tended to vaguely follow the melanotic lines.

These dots were not considered to be hematomas because they did not show a red periphery or filamentous distal end. Some researchers have noted these black dots in children with LM and have ascribed them to accumulation of pigment in the nail plate.[2] We believe these black dots represented an accumulation of melanin pigment and also likely derived from a cluster of nevus cells that migrate upward from the dermoepidermal junction. This conclusion was based on several factors.

First, in some cases the dots form shallow pits with black rims. The mechanism of this pit formation

may be analogous to psoriatic nail pitting. In psoriasis, small columns of pathologic parakeratotic nail fall off the upper surface of the nail plate to produce a pit.[6] In melanocytic nevi of the proximal nail matrix, an entire junctional nest of melanocytes may migrate upward to the nail plate where the cohesively nested melanocytes are not adhesive to surrounding corneocytes. These nested melanocytes may be separated and drop out to form pits if located on the upper surface of the nail. If the dots were a simple accumulation of melanin granules, they would not form pits because the granules are distributed within the nail corneocytes and do not disturb the tight connection of the corneocytes.

Second, if the dots were accumulations of melanin pigment derived from matrical melanocytes, the sizes of major axes of dots should vary according to the time period of melanin delivery from matrical melanocytes. Dots are not slender; they are round to oval in shape. When whole cells from a nevus cell nest are cast off, a round spot will form, not a slender structure, as the casting off is a solitary event for the one nest.

Third, in some of our patients the dots formed a boundary between darkly pigmented distal lines and lighter pigmented proximal lines (Figure 1B). This phenomenon can be explained as the result of a decrease in the number of melanocytes after the casting off of a nevus cell nest. In the case of malignant melanoma, the casting off of a melanocytic cell nest might not necessarily mean a decrease of melanin-producing cells because these cells with self-duplicating potency can proliferate and increase in number; however, for benign melanocytic nevi, a duplicated melanocytic cell nest may not be present after casting off.

Fourth, the dots were the same size as the nevus cell nests, which were histopathologically measured. In our study period, we had an opportunity to histopathologically observe a cluster of melanin pigment in the nail plate of an adult with LM resulting from a melanocytic nevus of the nail matrix (Figure 2). The cluster was distinct from surrounding corneocytes, showing round nuclei and densely packed melanin granules. The size of the cluster was similar to the junctional melanocytic nest. The cluster measured 0.085 mm with a micro-oculometer. Calculating the size on dermoscopic photographs with a scale, the dots measured 0.07 to 0.15 mm. Their relative similarity in size supported the idea that the dots were derived from casting off of the junctional melanocytic cell nests of the nail matrix. In ordinary melanocytic nevi on sites other than the nail apparatus, small, deep, black dots often are dermoscopically observed and sometimes can be histopathologically identified as clusters of cast-off nevus cell nests.

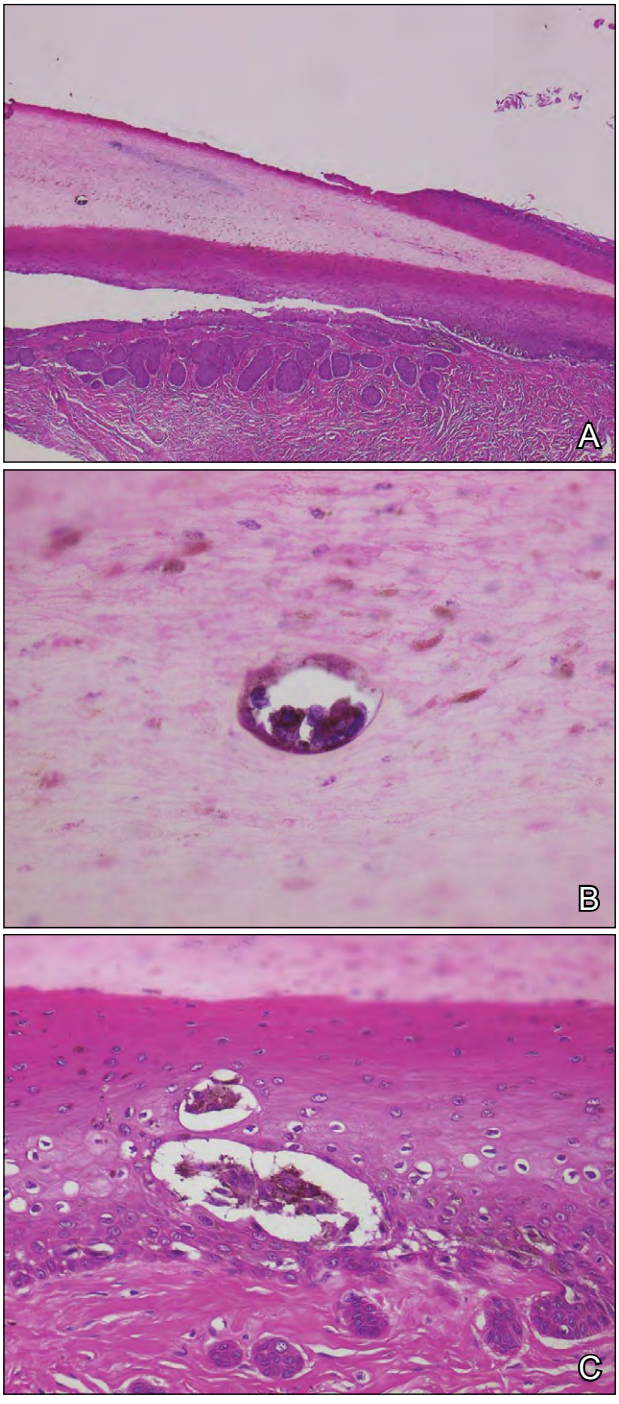

Figure 2. Biopsy specimen of melanonychia from an adult patient with a compound nevus with junctional melanocytic nests at the right of the slide (A)(H&E, original magnification ×100). The cluster of melanocytes was composed of round nucleated cells with abundant melanin granules and was distinct from surrounding corneocytes, which have parakeratotic slender nuclei and sparse melanin granules (B)(H&E, original magnification ×400). In the nail matrix, a melanocytic nest can be seen in the mid to upper spinous layer of the epidermis (C)(H&E, original magnification ×200).

CONTINUED ON PAGE 301

CONTINUED FROM PAGE 296

Because epidermal turnover time has been estimated to be 52 to 75 days,[7] a cluster of cast-off nevus cell nests can be expected to fall off the skin within a few months, and subsequent clinical changes may be overlooked without careful photographic examination in short-term follow-up visits; however, because fingernails grow approximately 1 cm every 3 months and toenails at one-third of this rate,[8] pigmentary abnormalities in the nail plate can be observed over a longer period of time. When managing patients with LM, physicians must recognize that they are just seeing a snapshot of a 4-dimensional event.

After many junctional melanocytic nests are cast off, there should be a decrease in the number of melanin-producing cells, resulting in the regression of LM. In our study, 6 of 7 patients with dots and lines showed gradual fading of LM in a follow-up period of more than 2 years, while only 2 of 8 patients without dots showed gradual fading. The difference between these 2 groups was statistically significant (P=.019; odds ratio, 18.0).

Conclusion

Dots and lines may be an indicator of spontaneous fading of LM in children, though this pattern may appear suspicious and may be misinterpreted as a warning sign of melanoma.

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