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CASE REPORT

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In Situ Melanoma of the Nail Unit in Children: Report of Two Cases in Fair-Skinned Caucasian Children

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Abstract: **Nail melanoma in children is rarely reported in the literature, and all of the published cases were diagnosed in dark-skinned phototypes or in Asians. We report two cases of in situ nail matrix melanoma presenting as longitudinal melanonychia (LM) in fair-skinned children of Italian origin. Nail plate dermatoscopy revealed a brown background with lines of irregular color, spacing, and thickness in both cases. Histopathology of the excised lesions showed melanoma in situ. Clinical, dermatoscopic, and pathological criteria that permit clear differentiation of benign melanocytic activation or proliferation from nail matrix melanoma are not established for children. The presence of a pigmented band of a single nail in a child usually represents a problem for clinicians, because the clinical and dermatoscopic features that are considered possible indicators of nail unit melanoma in adults are frequently observed in benign melanocytic hyperplasia and nevi in children. There is therefore the need to find parameters useful for clinical and dermatoscopic diagnosis in childhood nail pigmentation and to reach a consensus on management of children with a band of LM.**

Longitudinal melanonychia (LM) is the presence of a pigmented band due to a brown to black pigment within the nail plate, commonly melanin. When it occurs as a solitary lesion, it poses a diagnostic dilemma, because nail matrix melanocyte activation or benign (lentigo, nevus) or malignant (melanoma) proliferation of nail matrix melanocytes may cause it (1,2).

Longitudinal melanonychia in children is usually benign, in most cases due to a junctional nevus of the nail matrix. Nail melanoma in children is rarely reported

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in the literature, and all of the published cases were diagnosed in dark-skinned phototypes or in Asians (3,4).

We report two cases of in situ nail matrix melanoma presenting as LM in fair-skinned children of Italian origin.

CASE REPORT

Case 1

A 6-month-old boy (skin phototype 1) presented with a band of LM of the first right toe present since birth. The child was in good health. He did not have any melanocytic skin nevus. The band was dark brown and, at the first evaluation, had a triangular shape with the base at the nail matrix. At a follow-up visit 2 months later, the band had acquired a rectangular shape with parallel margins. It was irregularly deep brown, with a pale central part and darker peripheral lines. The nail plate showed distal splitting and fissuring. The periungual skin was not involved, but the dark pigmented band of the nail plate was visible through the transparent cuticle (pseudo-Hutchinson's sign). Nail plate dermatoscopy showed a dark-brown longitudinal band formed by lines with irregular color, spacing, and width (Fig. 1A). The family history for melanoma was negative, as was the history of trauma to the right big toe and pigmentation-related disorders.

It was decided to excise the entire lesion. The pathology showed a poorly circumscribed proliferation of heavily pigmented melanocytes in the epidermis with various degrees of pagetoid spread (Fig. 1B). Melanocytes were characterized by atypical hyperchromatic and polymorphic nuclei and atypical mitoses (Fig. 1C). A diagnosis of melanoma in situ was made, and a re-excision with wider surgical margins was performed. The histopathology showed no signs of residual melanocyte proliferation.

Case 2

An 11-year-old right-handed girl (skin phototype 2) presented with a LM of the second right fingernail that had been present since the age of 1 year. The child was in good health. She had a nevus count of < 10 and did not have any clinically suspicious melanocytic nevi.

The band involved most of the nail, was pale brown in color, and contained several longitudinal brown–black pigmented lines. Follow-up after 8 months revealed enlargement of the band and development of new black longitudinal lines. Nail plate dermatoscopy revealed a pale brown background with lines of irregular coloration, spacing, and thickness (Fig. 2A). Black dots were

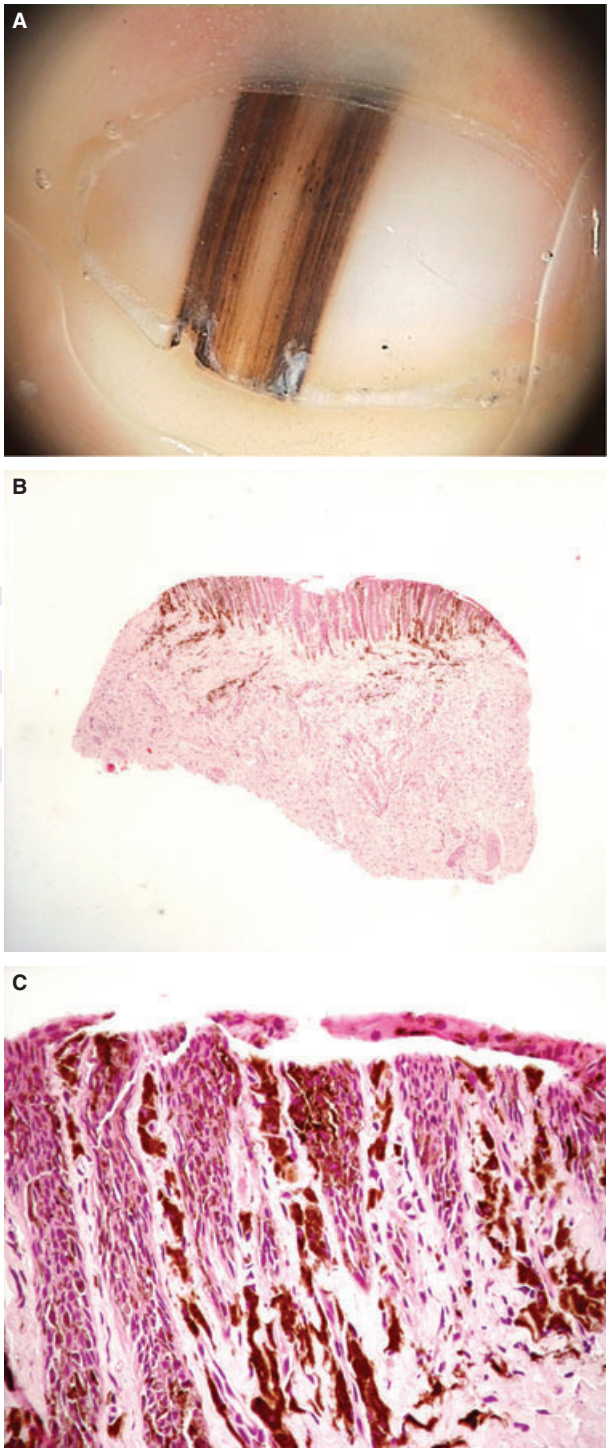

Figure 1. (**A**) Case 1: 6-month-old boy with a band of longitudinal melanonychia of the first right toe. Dermatoscopy shows a dark-brown longitudinal band with lines exhibiting irregular coloration, spacing, and thickness. (**B**) Low-power view of the heavily pigmented matrix lesion. (**C**) High-power view of the mainly lentiginous proliferation of large atypical melanocytes.

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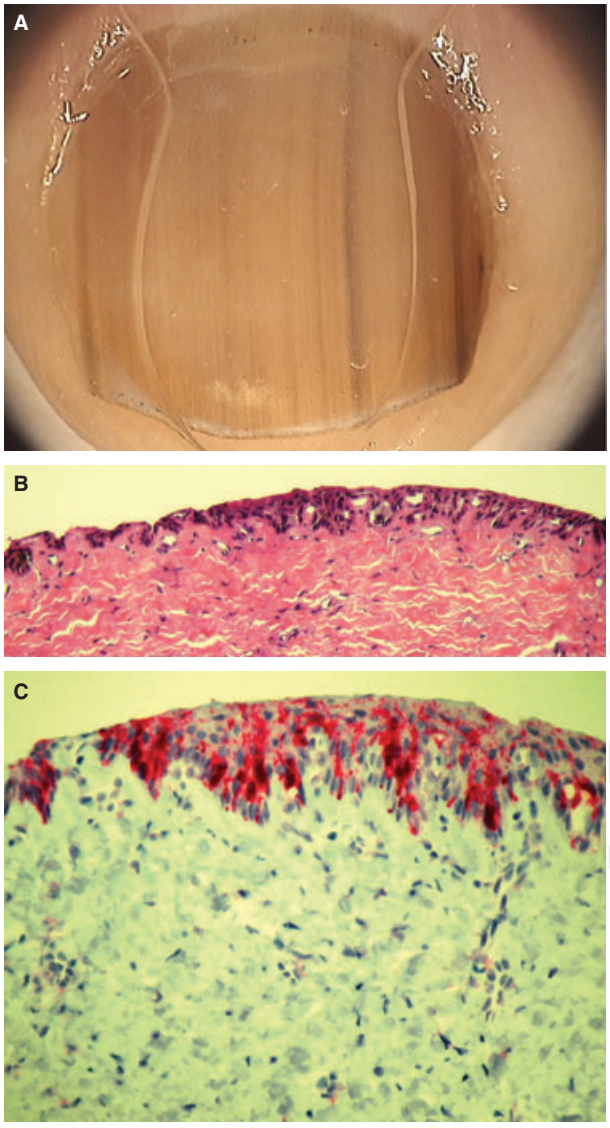

Figure 2. (**A**) Case 2: 11-year-old girl with a longitudinal melanonychia of the second right fingernail. Dermatoscopy shows a pale brown background with lines of irregular coloration, spacing, and thickness. (**B**) Scanning view of large irregularly spaced melanocytes. (**C**) MelanA stain reveals abnormal positive melanocytes.

evident in the proximal nail fold cuticle. We decided to excise the lesion completely with a tangential nail matrix biopsy. Intraoperatively, the entire matrix width was shown to be involved. Histopathology showed an intraepithelial melanocyte hyperplasia without nest formation and with striking cellular atypia (Fig. 2B), such as large irregular and hyperchromatic nuclei that were positive for protein S100, melanA, and HMB45 (Fig. 2C). Many of the atypical melanocytes were in the suprabasal epithelium.

A diagnosis of melanoma in situ was made. The nail regrew unpigmented to approximately 80% within 4 months. After discussion with the parents, we decided to remove the entire nail apparatus. The surgical margin of the tangential excision was 2 mm and of the complete nail apparatus removal was more than 5 mm. Complete degloving of the finger tip was not performed because it would not only not have increased the safety margin where it is important, but would also have left a finger tip without soft tissue, which would have had to be covered with a wrap-around full-thickness skin graft, which is functionally inferior because it lacks all sensory function of the finger tip and is esthetically inferior, so the finger pulp was left in place. A single tiny focus of abnormal melanocytes was found histopathologically at the undersurface of the proximal nail fold close to the cuticle, indicating that the first excision left a part of the tumor in place.

DISCUSSION

Longitudinal melanonychia in children younger than 12 is not common in any race (5), especially in the fair-skinned Caucasian population. The presence of a pigmented band in a single nail of a child usually represents a delicate problem for clinicians because clinical, dermatoscopic, and pathologic criteria that permit clear differentiation of benign melanocytic activation and proliferation from nail matrix melanoma are not established for children (6), and the clinical (7) and dermatoscopic features that are considered possible indicators of nail unit melanoma in adults are sometimes observed in benign melanocytic hyperplasia in children. Worrisome features in adults include bands that are not homogeneous in color, with blurred lateral borders, with irregular and not parallel lines upon dermatoscopy, presence of nail plate fissuring or splitting, rapidly enlarging streaks, increase or decrease of the pigmentation over time, bands with a triangular shape, and presence of pigmentation of the periungual skin (8). These features are commonly found in childhood melanonychia because of nail matrix melanocyte activation, lentigo, or nevus, and their finding in children is not considered an indicator for surgical excision of the lesion (9–11). Our cases had almost all of the features that are worrisome in adults, but these may, at least in part, be seen in children. This is what makes the clinical and dermatoscopic diagnosis so difficult.

Moreover, the differential diagnosis between benign melanocytic hyperplasia and in situ melanoma of the nail matrix is often a serious problem even for the pathologist, because the few studies in this field have only been performed in adults (12,13). The histopathological diagnosis of subungual melanoma in children is difficult. Nail matrix nevi in children often present a mild

degree of transepidermal melanocyte migration and some cellular atypia (9,10). Differential diagnosis between in situ melanoma and nevus of the nail matrix is based on the presence, in the former, of a large number of atypical melanocytes, with single melanocytes prevailing over nests, and pagetoid spread (9,10,12). Differentiation between in situ melanoma and benign melanocytic hyperplasia of the nail matrix is even more difficult in children, because qualitative and quantitative parameters, such as number of melanocytes per mm stretch of normal nail matrix epithelium, have been assessed only in adults (12–17). The diagnosis of melanoma in situ of the nail matrix in adults is based on quantitative parameters, such as a high density of melanocytes per mm (>40), and qualitative parameters, such as melanocyte confluence, pagetoid spread, and cellular atypia with multinucleated cells (17). The final diagnosis of in situ melanoma of the nail matrix in our two Caucasian children was made after long discussions on the pathological specimens by pathologists of different countries involved in the diagnosis of nail disorders and melanoma. Both cases presented the criteria proposed by Ackerman (13–16) and confirmed by Amin (17) for the histopathological diagnosis of melanoma in situ in adults. On the other hand, it had been stressed that the same criteria apply for the diagnosis of melanoma in adults and children (14).

We report these cases because they are the first reported cases of melanoma of the nail unit occurring in Caucasian children with fair skin and because the clinical and dermatoscopic features were similar to what is commonly seen in a great number of bands of LM in children, in which the pathology reveals nail matrix nevi. Only one of the 10 previously reported cases of nail melanoma in dark-skinned children was evaluated with dermatoscopy, which showed a dark-brown background with irregular parallel lines. Melanonychia was the presenting symptom in eight of the 10 patients and in all cases in which the band was excised because it was growing. It is therefore necessary to find parameters useful for clinical and dermatoscopic diagnosis in childhood nail pigmentation and to have a consensus on management of children with a band of LM. There are still different opinions on whether a single band of LM with clinical and dermatoscopic features that suggest melanocyte hyperplasia in a child should be excised. Our two cases indicate that the "wait and see" policy that the results of previous studies on LM in children have suggested is not appropriate (9,10) and may produce delayed diagnosis of melanoma.

Case 2 also raises the problem of the correct surgical approach in malignant LM, because the second surgical procedure with complete removal of the nail apparatus showed that a focus of atypical melanocytes in the dorsal surface of the proximal nail fold remained even though the regrown nail did not exhibit any pigmentation clinically, dermatoscopically, or histopathologically. This indicated that the tangential nail biopsy (shave excision) did not remove all nail matrix melanocytes giving rise to the pigmentation even though the surgical margins were clear with the tangential excision, and the newly diagnosed melanocyte focus was not contiguous with the former excision margin. This reinforces our approach of removing the entire nail apparatus in the case of ungual melanoma, although some of the cases of LM undergoing tangential nail matrix biopsy show a recurrence of the pigmentation, even if lighter and thinner than the previous one, when the nail plate regrows. This is probably due to too narrow a margin, which can, however, be extended without the risk of nail dystrophy. Total removal of the source of the pigment is mandatory for a correct pathological diagnosis and for a good patient outcome.

A final problem remains unsolved: are these melanomas detected in children (aged 3–4) after removal of pigmentations present since birth or soon after, are they malignant from the beginning, or do they arise from an initial benign melanocyte hyperplasia? Only accumulating evidence on pathologic studies of childhood pigmentation will answer these questions.

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AUTHOR CONTRIBUTIONS

Antonella Tosti, Bianca Maria Piraccini, Anna Cagalli, and Eckart Haneke had full access to all of the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Tosti, Piraccini, Cagalli, Haneke. Acquisition of data: Tosti, Piraccini, Cagalli, Haneke. Analysis and interpretation of data: Tosti, Piraccini, Cagalli, and Haneke. Drafting of the manuscript: Tosti, Piraccini, Cagalli, and Haneke. Critical revision of the manuscript for important intellectual content: Tosti, Piraccini, Cagalli, and Haneke Obtained funding: no funding was obtained. Administrative, technical, or material support: Tosti, Piraccini, Cagalli, and Haneke. Study supervision: Tosti, Piraccini, Cagalli, and Haneke.

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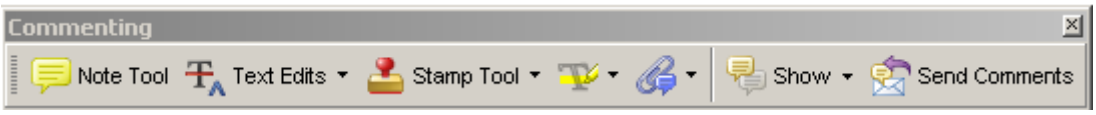

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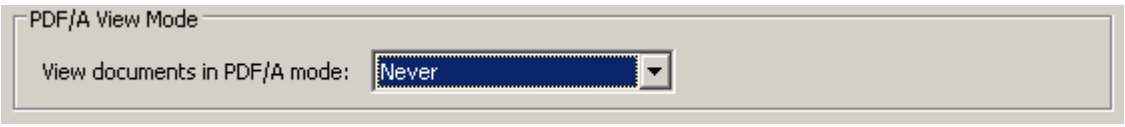

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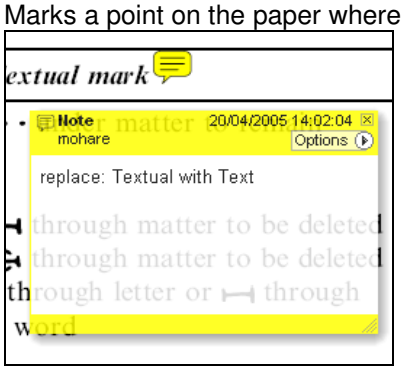

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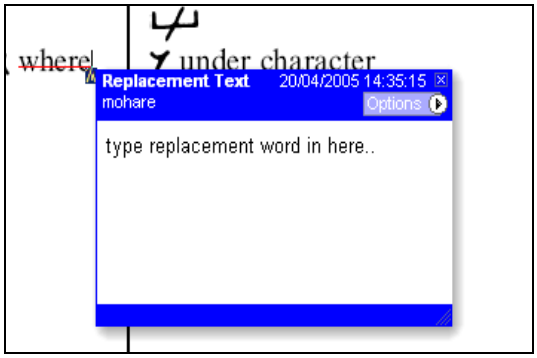

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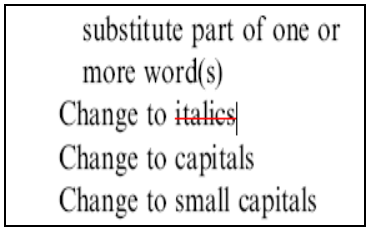

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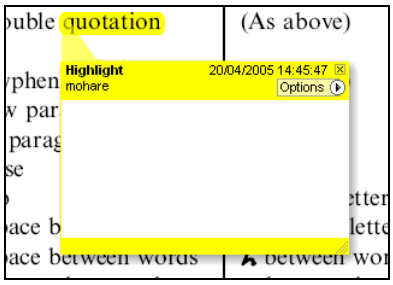

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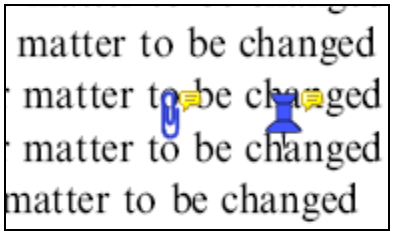

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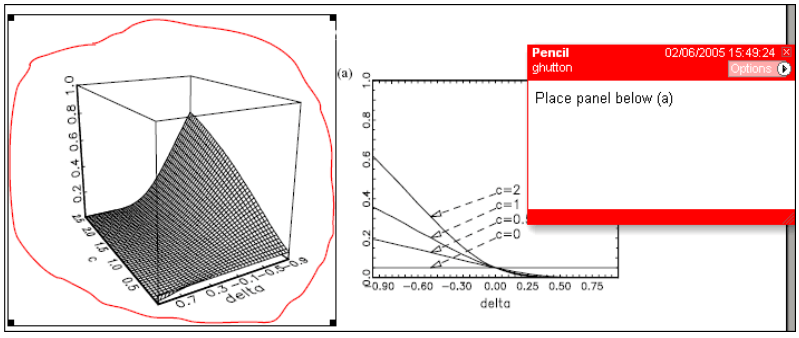

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