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Heffi Anindya Putri
Dermatology and Venereology Department, Medical Faculty, Andalas University, Padang, Indonesia

Ennesta Asri
Dermatology and Venereology Department, Dr. M. Djamil Hospital, Padang, Indonesia

Qaira Anum
Dermatology and Venereology Department, Dr. M. Djamil Hospital, Kota Padang, Indonesia

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Case Report

A case of longitudinal melanonychia in a child: Benign or malignant?

Heffi Anindya Putri[1], Ennesta Asri[2], Qaira Anum[2]

[1]Dermatology and Venereology Department, Medical Faculty, Andalas University,
Jl. Perintis Kemerdekaan, Sawahan, Kecamatan Padang Timur, Kota Padang, Indonesia
[2]Dermatology and Venereology Department, Dr. M. Djamil Hospital,
Jl. Perintis Kemerdekaan, Sawahan, Kecamatan Padang Timur, Kota Padang, Indonesia

Email: heffi_anindya@yahoo.com

Abstract

Background: Longitudinal melanonychia (LM) describes a longitudinal brown-to-black band of discoloration affecting a finger, thumb, or toenail. Discoloration of the nails can be a sign of benign or malignant nail disorders. Although nail melanoma rarely occurs in children, health practitioners should be aware of melanonychia. Dermoscopy and histopathology are helpful in the diagnosis.
Case Illustration: A case of LM in a thirteen-year-old girl was reported with a complaint of brownish-black discoloration, sometimes painful, on her right thumb since 1 year ago. She had no history of trauma, nail damage, or bleeding tendency. Dermatological examination found a homogeneous brownish-black band on the right thumbnail. Dermoscopy revealed regular, homogeneous black linear pigmentation and pigment through the translucent cuticle and proximal nail fold (pseudo-Hutchinson's sign). Punch biopsy was done on proximal nail fold reaching nail matrix. Histopathology revealed increased melanocytes, some nests of melanocytes in rete Malpighi, and no atypical cells. She was diagnosed with nail matrix nevus. Five months after the punch biopsy, the nail grew well without onychodystrophy.
Discussion: Nail matrix nevus is a benign LM. Diagnosis is based on anamnesis, physical examination, dermoscopy, and histopathology. Dermoscopy of the nails is a useful, quick, non-invasive, and highly effective tool that may help differentiate benign or malign melanonychia.
Conclusion: Histopathology is important to determine whether the pigmented lesion is benign or malignant.

Keywords*: melanonychia striata on a child*

Background

Longitudinal melanonychia (LM) or melanonychia striata is a brown-to-black band that develops in the nail running from the proximal nail fold to the free margin of the nail plate. It arises as a result of the deposition of pigment in the nail plate from increased pigment production within the nail matrix.[1,2]

The publication of the epidemiology of melanonychia in children has been scarce. The fingernails are more frequently affected than the toenails, and among the fingernails, the first digit is the most frequently affected. The mean age at onset is 3 years. The prevalence increases after puberty. No gender differences are reported among children. Most cases of longitudinal melanonychia are nevi. Nail melanoma has an incidence of 0.7%-7% of all cases of melanoma.[3,4]

The etiologies of melanonychia may be divided into two broad categories: melanocytic activation and melanocytic hyperplasia. Melanocytic activation (also termed melanocytic stimulation or functional melanonychia) describes the process by which melanonychia results from an increased melanotic pigmentation of the nail matrix epithelium and nail plate without a concurrent increase in the number of melanocytes. Common causes of melanocytic activation are race, underlying syndromes, or drugs. In dark-skinned populations, such as African–Americans

and people from the Caribbean, 77% of children have melanonychia caused by melanocytic activation, especially racial activation, which involves multiple nails. Melanonychia can represent a sign of a syndrome. The syndromes that involve melanonychia in children are Peutz–Jeghers syndrome and Laugier–Hunziker syndrome. Peutz-Jeghers syndrome, pigmented macules on the oral mucosa, lips, fingers, and toes, and melanonychia are associated with intestinal polyposis, with a possible malignant degeneration. In Laugier–Hunziker syndrome, LM affects several nails and is associated with pigmented macules of the lips, mouth, esophageal mucosa, and genitalia. Other causes of melanocytic activation in children are trauma, irradiation, gold therapy, cytotoxic agents, arsenic intoxication, hemochromatosis, Addison's disease, or vitamin B12 deficiency.[5]

Benign melanocytic hyperplasia is subdivided into two categories: lentigines when nests of melanocytes are absent, and nevi when at least one melanocytic nest is present. While lentigines are observed more often than nevi in adults, nevi are found more often in children. Subungual melanoma rarely occurs in children, and benign melanocytic hyperplasia constitutes 77.5% of childhood melanonychia cases .[6]

The diagnosis approach in melanonychia in children is to combine personal/ family history together with clinical and dermoscopic features. Dermoscopy is a non-invasive technique that enhances the clinical evaluation of LM and has been demonstrated to help improve the clinical decision-making regarding whether to perform a biopsy. The best way to manage these patients is to repeat a regular and accurate follow-up that includes global photography and dermoscopy. A dermatologist should advise immediate excision of pigmented lesions with alarming clinical features, such as a band that enlarges to involve the whole nail plate and or darkens, and or bands with irregular borders and spacing, and or a thick pattern of lines and areas of disrupted parallelism. The decision to perform a biopsy on a child with melanonychia is taken when the parents are very anxious.[3]

Case Illustration

A 13-year-old Indonesian girl had a chief complaint of brownish-black longitudinal discoloration on her right thumb fingernail that sometimes felt painful since 1 year ago. Initially, 1 year ago, the patient saw a black spot slowly growing from the base to the tip of the nail, forming a straight line on her right thumb fingernail. The patient's parents were worried because online media suggested that the abnormalities in the patient's nails were caused by malignancy. There was no pain, swelling, warmth, tenderness, or redness on the skin around the nails. History of nail trauma and bleeding was denied; history of malignancy, radiotherapy, chemotherapy, or phototherapy was denied. There was no history of blackish discoloration with progressive changes on other nails or body areas. History of nevi with progressive changes on other body areas was denied. History of taking medications such as antimalarials, minocycline, and phenytoin was denied. History of pigmented macules on the oral mucosa, lips, fingers, toes, or genitalia was denied. There was no history of painful blackish discoloration with progressive changes and easy bleeding on nails or other body areas in her family.

General physical examination was normal with Fitzpatrick's skin type IV. Dermatological examination revealed blackish discoloration with a longitudinal line, no hemorrhagic and granulation tissue, no onycholysis, and no transversal line. The skin around the nail was normal. Hutchinson's sign was negative. The band was 0,1mm wide on the right thumb fingernail.

Dermoscopy image revealed regular, homogeneous black linear pigmentation and pigment, visible through the translucent cuticle and proximal nail fold (pseudo-Hutchinson's sign). Histopathological examination revealed epidermal tissue with a surface consisting of keratin mass (nail plate), an increased number of melanocytes, and some nests of melanocytes in rete Malpighi. There were no atypical cells in this preparation.

We diagnosed this patient with nail matrix nevus and explained to the patient and her parents that her nail condition is benign and to be cautious for nail discoloration, especially if the color is not homogeneous, or irregular with onychodystrophy and bleeds easily. There was no specific treatment for the disease at our dermatology and venereology department. We followed up on the biopsy wound 3 times. After 5 months from punch biopsy, the nail grew well without onychodystrophy or black discoloration.

Discussion

This study reported a case of longitudinal melanonychia caused by nail matrix nevus in a 13-year-old girl, with a history of black discoloration line on her right thumb since 1 year ago and sometimes felt painful over the last month. History of nail trauma, nail bleeding, malignancy, radiotherapy, chemotherapy, phototherapy, and history of medications such as antimalarials, minocycline, phenytoin, and

chemotherapeutic agents was denied. There was no history of malignancy in her family.

Melanonychia in childhood is usually due to benign melanocytic lesions (e.g., melanocyte activation, lentigo, or melanocytic nevus). Although rare, cases of pediatric subungual melanoma *in situ* have been reported in the literature. Less common causes of melanonychia in children and adults include drugs, such as antimalarials, minocycline, phenytoin, and chemotherapeutic agents, which affected multiple nail digits.[7]

Tan et al.[7] in Singapore reported that out of 14 childhood LM biopsies, no diagnosis of subungual melanoma was found. Seven cases were diagnosed as nevus, 5 cases as lentigo, and 2 cases as melanocyte activation. Study literature by Bonifazi[4] in Italia revealed 11 case reports of *in situ* nail melanoma presenting as longitudinal melanonychia in children during 1988-2014.[4]

On physical examination, we excluded malignancy first. In subungual melanoma, there are signs of brown to black discoloration or growth in the nail bed. A widening, dark, or irregularly pigmented longitudinal nail streak with or without nail dystrophy and nail plate elevation may be seen. Hutchinson's sign (pigmentation on the proximal nail fold) may be noted.[8] This patient had a longitudinal line on the regular nail with a homogeneous black color, no onychodystrophy, no onycholysis, and no Hutchinson's sign. Furthermore, the alphabet of nail melanoma (ABCDEF) was used to assist in the risk assessment of subungual melanoma (Table 1).[9] Among the 6 alphabets of nail melanoma, only the digit involved (D criteria) was positive in this patient.

Table 1. The alphabet of nail melanoma

	Subungual melanoma	Patient
A : Age	50-70 years old African, Japanese, Chinese, and Native Americans are dominant	13 years old Indonesian
B : Band or strip	Brown-black band often >3mm with an irregular border	Black band, 1mm, regular border
C : Change	Change in size and growth rate or failed improvement with adequate treatment of the alternative cause	No change in size, the patient has never been treated
D : Digit involved	The thumb is involved more frequently than the index finger and more frequently on one finger	Right thumb fingernail
E : Extension	Extension of discoloration, Hutchinson's sign	No Hutchinson's sign
F : Family or personal history	Family history of melanoma	No family history of melanoma

Taken from Levit et al.[9] The ABC rule for clinical detection of subungual melanoma. J Am Acad Dermatol. 2000;42(2):269–74.

Dermoscopy (onychoscopy) helps distinguish subungual melanoma from benign melanocytic pigmented lesions. Some characteristics suggestive of melanoma include irregular bands of brown or black pigmentation, unparallel lines, ill-defined borders, width >3 mm, nail dystrophy, nail fold ulceration, and Hutchinson's sign.[5,10] Onychoscopy examination on this patient revealed regular, homogeneous black linear pigmentation and pigment visible through the translucent cuticle and proximal nail fold (pseudo-Hutchinson's sign), indicating the lesion was benign. Dermoscopy on nail matrix nevus revealed a regular pattern of longitudinal parallel micro lines and brown-black background to the pigmentation according to Fitzpatrick's skin type. The darker the skin color, the darker the longitudinal pigmentation. Nail apparatus lentigines and melanocytic activation such as ethnic-type pigmentation and drug-induced pigmentation were significantly associatedwith a homogeneous grayish coloration of the background, the presence of thin longitudinal gray lines, and regular coloration, thickness, spacing, with and without pseudo-Hutchinson's sign.[5,6]

Histopathology is useful in differentiating benign and malignant lesions. A punch biopsy is performed on lesions <3mm in diameter and on the proximal longitudinal band to reach the nail matrix. Classification of histologic diagnosis of nail matrix biopsy is divided into:

1. Melanocyte activation, an increase in melanin deposition in the epidermis but no increase in melanocyte numbers.
2. Lentigo, an increase in melanin deposition and number of single melanocytes at the basal epidermis with an absence of atypia.
3. Nevus, an increase of single and nests of melanocytes at the basal epidermis with the absence of atypia.
4. Atypical melanocytic hyperplasia, melanocyte proliferation with mild cytological and architectural atypia.
5. Melanoma *in situ*, melanocyte proliferation with worrying architectural and cytological features such as moderate to marked cytological atypia, crowding of melanocytes, and pagetoid spread.
6. Invasive melanoma, melanocyte proliferation with architectural and cytological features of atypia, and invasion beyond the epidermis.[7,11,12]

Histopathology of this patient revealed melanocytic nevus with a surface consisting of keratin mass (nail plate), increased melanocytes, and nests of melanocytes in rete Malpighi on the epidermis. There were no atypia cells in this preparation. This condition indicates a benign lesion.

There is no specific therapy for nail melanocytic nevus. This patient was diagnosed with nail matrix nevus and the lesion regressed completely five months after punch biopsy. A retrospective study by Lee et al.[13] in Seoul on-the clinicopathology features of 20 nail matrix nevi cases found that 2 out of 20 cases of melanonychia disappeared after nail matrix biopsy. However, there was no further explanation. There is no publication regarding punch biopsy as a therapy for nail nevus melanocytic, but in cutaneous acquired melanocytic nevi, complete removal of nevi is one of the frequently used therapies. Leaving a partially excised nevus is fraught with potentially concerning consequences of repigmentation, regrowth, or both (recurrent melanocytic nevus).[14]

Nail biopsies on children carry a risk of permanent nail dystrophy. Tan et al.[7] in Singapore reported that 3 out of 14 patients who were biopsied developed permanent nail dystrophy. Lee et al.[13] in Seoul revealed that 12 out of 20 patients had nail dystrophy after biopsy. The lesion biopsied in this patient regressed without onychodystrophy.

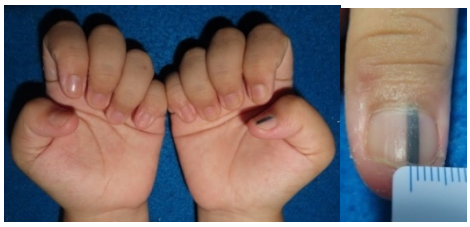

Figure 1. Longitudinal band on right thumb fingernail

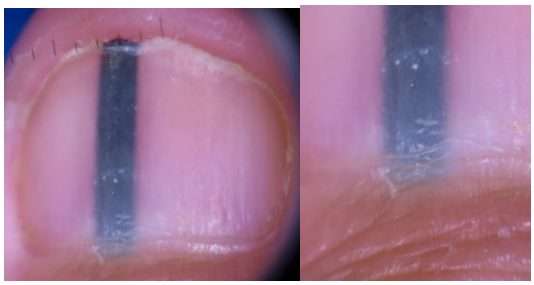

Figure 2. Dermoscopy image revealed regular, homogeneous black linear pigmentation and pigment through the translucent cuticle and proximal nail fold (pseudo-Hutchinson's sign).

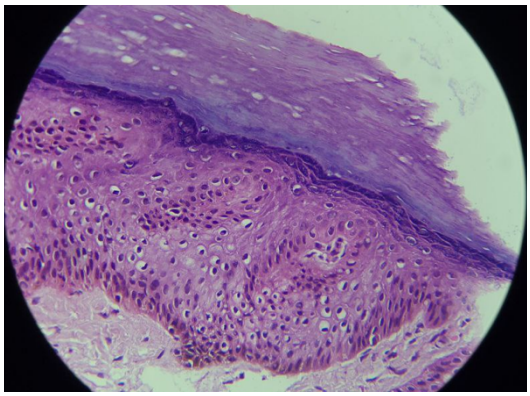

Figure 3. Histopathology showed epidermal tissue with a surface consisting of keratin mass, some nests of melanocytes in rete Malpighi, and increased melanocytes.

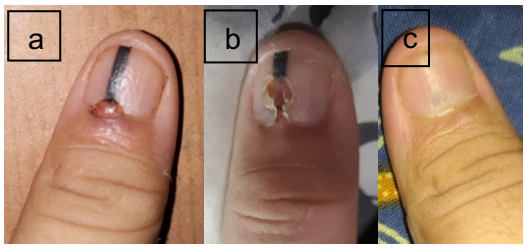

Figure 4. The wound after the biopsy a.) Two weeks. b.) Two months. c.) Five months.

Conclusion

This case reported a benign longitudinal melanonychia caused by nail matrix nevus in a 13-year-old girl with black discoloration, sometimes painful, on her right thumb. The diagnosis was based on anamnesis, physical examination dermoscopy, and histopathology. There was black coloration, regular pattern of longitudinal lines, regular thickness, coloration, and parallelism with pseudo-Hutchinson's sign on dermoscopy. There were some nests of melanocytes and no atypia cells on histopathological examination thus showing a benign lesion. A biopsy is useful in differentiating benign and malignant lesions.

References

1. Cooper C, Arva NC, Lee C, et al. A clinical, histopathologic, and outcome study of melanonychia striata in childhood. J Am Acad Dermatol. 2015;72(5):773–9.
2. Haneke E. Nail Disorder. In: Kang S, Amagai M, Bruckner AL, et al., editors. Fitzpatricks dermatology in general medicine. 9th ed. New York: McGraw-Hill Companies; 2019. p. 1568–608.
3. Piraccini BM, Alessandrini A, Dika E. Melanonychia in Children. In: A.Tosti, Chiaccio N Di, editors. Melanonychias. 1st ed. Switzerland: Springer International Publishing; 2017. p. 85–96.
4. Bonifazi E. Longitudinal melanonychia in childhood. Eur J Pediatr Dermatology. 2017;27(4):215–23.
5. Chiacchio N Di, Farias DC de, Piraccini BM, et al. Consensus on melanonychia nail plate dermoscopy. An Bras Dermatol. 2013;88(2):309–13.
6. Jefferson J, Oregon PR. Melanonychia. Hindawi dermatology Res Pract. 2012;1–8.
7. Tan WX, Wang DY, Seghers AC, Koh MJA, Goh SG, Lee SS. Should we biopsy melanonychia striata in Asian children? A retrospective observational study. Pediatr Dermatol. 2019;36(6):864–8.
8. Hassel JC, Enk AH. Melanoma. In: Kang S, Amagai M, Bruckner AL, Enk AH, Margolis DJ, Mcmichael AJ, et al., editors. Fitzpatricks dermatology in general medicine. 9th ed. New York: McGraw-Hill; 2019. p. 1982–2011.
9. Levit EK, Kagen MH, Scher RK, Grossman M, Altman E. The ABC rule for clinical detection of subungual melanoma. J Am Acad Dermatol. 2000;42(2):269–74.
10. Tosti A, Piraccini BM, Cagalli A, Haneke E. In situ melanoma of the nail unit in children: Report

of two cases in fair-skinned caucasian children. Pediatr Dermatol. 2012;29(1):79–83.
11. Güneş P, Göktay F. Melanocytic lesions of the nail unit. Dermatopathology. 2018;5(3):98–107.
12. Ren J, Ren M, Kong YY, Lv JJ, Cai X, Kong JC. Clinicopathological diversity and outcome of longitudinal melanonychia in children and adolescents: Analysis of 35 cases identified by excision specimens. Histopathology. 2020;77(3):380–90.
13. Lee JH, Lim Y, Park JH, et al. Clinicopathologic features of 28 cases of nail matrix nevi (NMNs) in Asians: Comparison between children and adults. J Am Acad Dermatol. 2018;78(3):479–89.
14. Cuda JD, Moore RF, Busam, J. K. Melanocytic Nevi. In: Kang S, Amagai M, Bruckner AL, et al., editors. Fitzpatricks dermatology in general medicine 9th edition. 9th ed. New York: McGraw-Hill Companies; 2019. p. 1951–7.