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Acquired blue nevus of the nail bed

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Abstract

Blue nevi are benign proliferations of melanin-producing dendritic melanocytes located in the dermis. These nevi tend to occur mostly on the skin, predominantly on the head and neck, dorsal aspects of the distal extremities, and the sacral area, but can also occasionally appear on mucosal surfaces. Blue nevi of the nail apparatus are uncommon. The majority originate in the nail matrix where there is a higher density of melanocytes. Herein we report on a 47-year-old man with a rare common blue nevus of the nail bed, an area with low melanocyte density. A thorough review of the English language literature found no documented cases of acquired blue nevi originating in the nailbed of the toe.

Keywords: blue nevus, nail apparatus, benign melanocytic lesions, subungual melanoma

Introduction

Blue nevi are dermal melanocytic proliferations that are histologically characterized by dermal spindle-shaped and pigmented bipolar dendritic cells [1,2]. This generally benign skin tumor can be congenital or acquired. The nevi usually present as gray-blue or brown-blue solitary nodules or plaques, often found on the skin of the head and neck, dorsum of distal extremities, and pre-sacral regions and occasionally in the oral cavity [3] and mucosal surfaces. Nevi on the nail apparatus, however, are quite rare with only eleven cases currently documented in the literature, only two of which document cases in the nail bed [4,5]. Although blue nevi are typically benign, there have

been reports of malignant degeneration particularly of the cellular variant [1,2,6]. Nonetheless, careful examination and evaluation should be performed in order to differentiate between a blue nevus and a subungual malignant melanoma, which carries a poor prognosis. We report here on a 47-year-old Japanese man with an acquired blue nevus in the nail bed of his right first toe and discuss clinical and pathologic characteristics of this rare finding.

Case Synopsis

A 47-year-old Japanese man with a family history of acral lentiginous melanoma presented to clinic with a blue macule under the right first toenail, which had been present for one year. He denied any pain or tenderness and reported that the lesion had been stable in size. Physical examination showed a 2 mm blue circular macule on the nail bed immediately distal to the lunula of the right first toe (**Figure 1**).



Figure 1. Well-demarcated blue spot on right first toenail.

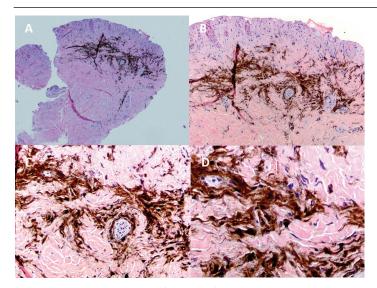


Figure 2. Histopathology of biopsy of blue nevus. (A) Hematoxylin-eosin stain; original magnification x10 (B) Hematoxylin-eosin stain; original magnification x20 (C) Hematoxylin-eosin stain; original magnification x40 (D) Hematoxylin-eosin stain; original magnification x100

Dermoscopy showed a homogenous blue blotch. A punch biopsy of the nail bed revealed dendritic melanocytes containing fine melanin and numerous polygonal macrophages containing abundant coarse melanin throughout a fibrotic reticular dermis (**Figure 2**). A MART-1 immunohistochemical stain was positive for dermal melanocytes and a Fontana-Masson stain was positive for dermal pigment (**Figure 3**). The patient was diagnosed with an acquired common blue nevus on the nail bed, which was completely removed by punch biopsy. No recurrence of the blue nevus was noted on follow-up.

Case Discussion

A blue nevus is a pigmented lesion caused by proliferating melanocytes in the dermis, usually presenting as a single, small dark blue or blue-black macule or dome-shaped papule. The unique blue color of these nevi is attributed to the Tyndall effect, which causes the skin to reflect shorter wavelengths of the blue spectrum [2].

The predominant theory regarding the pathogenesis of blue nevi is that they arise from aberrant dendritic melanocytes that fail to completely travel from the dermis to epidermis during embryologic migration of melanocytes [7]. Additionally, inflammation or other insults may play a role in inducing proliferation of the aforementioned aberrant melanocytes, resulting in acquired blue nevi [8].

Melanocytic nevi of the nail apparatus are extremely rare simply because the number of melanocytes in the nail are much lower than the number in the skin. For comparison, the epidermis has a melanocyte density of about 1150/mm2 whereas the nail matrix has about 200/mm2. What makes our case truly unique is that our patient's nevus involved the nail bed, where melanocytes are even rarer, often completely absent or having a density of only 50/mm2. Furthermore, nail bed melanocytes are, by nature, entirely dormant, meaning they produce no pigment [9].

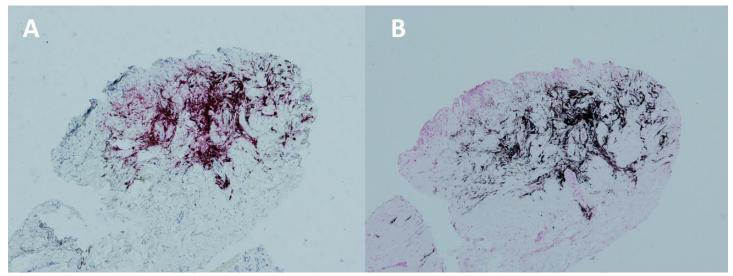


Figure 3. Histopathology of biopsy of blue nevus. (A) Fontana-Masson silver stain showing that the lesion is staining positively for dermal melanin pigment; original magnification x10 (B) MART1 (Melan-A) stain to highlight dermal melanocytes of the lesion; original magnification x10

Given the quiescent nature of nail bed melanocytes and their sheer low number, it should come as no surprise that melanocytic nevi are rarely observed in clinical practice. Our report of a nail bed nevus is only the third such report in the literature, and the sole case of an acquired blue nevus of the toenail bed (**Table 1**). **Table 1** illustrates that common blue nevi are the most common type found on the nail apparatus with five such documented cases. Blue nevus is often found as a single nevus; multiple nevi should raise concerns for Carney complex, or NAME/LAMB syndromes [10]. Only one case of subungual blue nevus has been associated with a syndrome [11].

Despite the generally benign nature of blue nevi, malignant transformation has been reported, particularly in the compound variant with dermal-epidermal junction involvement [6,12]. Malignant blue melanoma is a rare form of aggressive melanoma that can arise in a common/cellular blue nevus or at sites of previous excision. A malignant blue nevus is characterized by tumor necrosis, high grade epithelioid cell atypia, pleomorphic nuclei, and high mitotic activity [13]. As such, subungual blue nevi require a detailed work-up and thorough consideration of the differential diagnosis, including subungual malignant melanoma, which represents between 0.7 and 3.5% of all cutaneous melanoma

Table 1. Reported cases of blue nevi of nail apparatus

Source	Age	Gender	Location	Clinical findings	Histology	Onset	Syndrome?	Recurrence
Vidal et al. [20]	20	M	Right foot, first toe	Blue-black nodule at distal edge of nail plate with hypony- chium involvement	Common blue nevus (no junction- al compo- nent)	Congenital	No	NR
Soyer and Kerl [11]	4	F	Left foot, first toe	N/A	Combined blue nevus	Congenital	Yes - Klip-	N/A
Salasche and Garland [21]	NR	NR	Thumb	Longitudinal mela- nonychia	NR	NR	NR	NR
Kim et al. [4]	44	F	Right hand, thumb	Longitudinal mela- nonychia and blue discoloration in lunar area; involved prox- imal nail bed and distal nail matrix	Common blue nevus	Acquired	No	NR
S m i t h , Morgan, and Bettencourt [22]	61	M	Right hand, third finger		Common blue nevus	Acquired		No – 2 years
Dalle et al. [23]	34	F	Right hand, second finger	Blue gray macule in middle of lunula; superficial linear erosion of nail plate extended from distal part of tumor process to the distal nail plate	Common blue nevus	Acquired	No	No – 2 years
Causeret et al. [24]	42	F	Left foot, first toe	Ovoid-shaped blue pigmentation of distal nail matrix area	Cellular blue nevus	Acquired	No	No – 1 year

Source	Age	Gender	Location	Clinical findings	Histology	Onset	Syndrome?	Recurrence
Moulonguet- Michau and Abimelec [25]	52 42	F M	1) Left first toe 2) Right hand, thumb	Blue-black macule Blue-black macule	Cellular blue nevus (?) Cellular blue nevus (?)	Acquired Acquired	No No	No – 3 years No – 2 years
Naylor et al. [26]	40	F	Left hand, fourth finger	Dark blue dyschro- mia under proximal nail plate, overlying proximal nail bed and extending under proximal nail fold	Combined blue nevus – arising from directly beneath the nail bed and matrix epithelium, forming nodule in the dermis	Acquired	No	No – 12 months
Gershtenson et al. [5]	21	F	Right foot, second toe	Irregularly shaped, sharply delineated blue-black plaque extending to the cuticle, proximal and lateral nail fold, and adjacent nail bed and hyponychium	Cellular blue nevus	Congenital	No	No – 1 year
Our case	47	М	Right foot, first toe	Blue circular macule on nail bed	Common blue nevus	Acquired	No	No

[14]. However, the incidence is significantly higher in the Japanese population, estimated to be as high as 23% [15]. This was of particular concern in our patient given his Japanese ethnicity and family history of acral lentiginous melanoma. Clinical signs such as enlargement of the nevus, nail dystrophy, or periungual extension should raise concern for possible subungual melanoma [5]. Other potential causes for similar appearing lesions include argyria [16-18], glomus tumor, Wilson disease, iatrogenic pigmentation by drugs such as minocycline or zidovudine, and occupational exposures [19]. Once the diagnosis of subungual blue nevus is confirmed, treatment options may include complete excision, as performed in our patient. However, excision of larger nevi does increase the risk of nail dystrophy or even functional impairment of the affected digit [5].

Conclusion

A blue nevus is a unique neoplasm caused by the dermal proliferation of melanocytes. Although they can be located anywhere on the body, blue nevi of the nail bed are extremely rare and can be mistaken for glomus tumors or drug-induced pigmentation. Blue nevi have an excellent prognosis, but providers

must biopsy to rule out the possibility of a subungual melanoma.

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