JAMA Dermatology Clinicopathological Challenge

A Case of Pigmented Longitudinal Melanonychia

Lindsey West, BA; Lisa Garner, MD

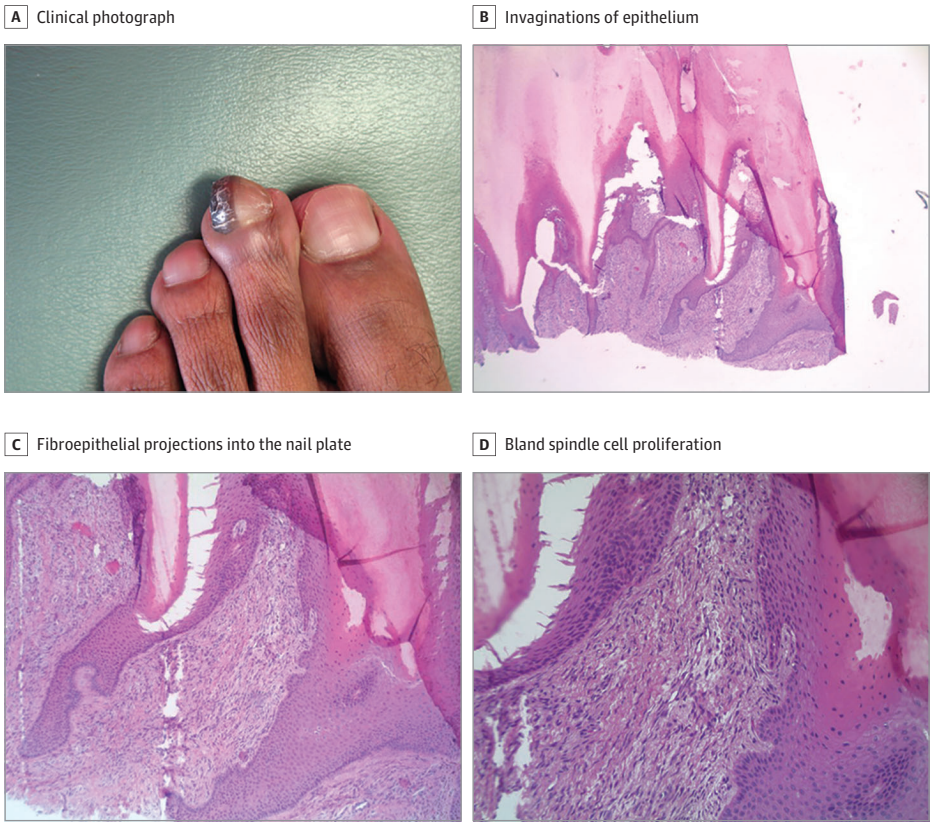

Figure. A dark brown longitudinal pigment band on the second toenail with pigment extends to the proximal nail fold (A). Histopathologic examination of the specimen with hematoxylin-eosin stain is seen at original magnification of ×40 (B), ×100 (C), and ×200 (D).

A man in his 40s presented to the dermatology clinic for evaluation of a new pigmented lesion on his left second toenail. The lesion first appeared 12 months before presentation and was progressively growing. This was his first time presenting to a physician for this problem, and no prior treatments had been attempted. The lesion was asymptomatic. He was otherwise well and denied any fevers or weight loss.

Clinical examination revealed a dark brown longitudinal pigment band covering the full length of the toenail of the left second digit with corresponding nail thickening over the pigmented band. The pigment was the same width throughout the band and extended to the proximal nail fold (**Figure**, A). No other nail or skin lesions were found at the time of examination. A biopsy specimen was obtained from the nail plate and nail matrix and stained with hematoxylin-eosin (Figure, B-D).

WHAT IS YOUR DIAGNOSIS?

A. Pigmented onychomatricoma

B. Subungual melanoma

C. Pigmented squamous cell carcinoma in situ

D. Pigmented onychopapilloma

Diagnosis

A. Pigmented onychomatricoma

Microscopic Findings and Clinical Course

Histologic examination of the nail plate and nail matrix showed bland spindle cell proliferation within the nail matrix with invaginations of the epithelium. No melanocytic lesions were observed, and immunostaining was negative for S100 and CD34. Intracorneal hemorrhage and adjacent verrucous hyperkeratosis were present.

Based on the histologic findings of villous fibroepithelial projections into the nail plate without melanocytic proliferation or keratinocyte atypia, a diagnosis of pigmented onychomatricoma was made. The nail was surgically removed to fully excise the lesion after the original biopsy. At approximately 3 months of follow-up, the patient was doing well and without evidence of recurrence of the lesion.

Onychomatricoma is a rare benign nail matrix tumor, with fewer than 80 cases reported since it was first described in 1992 by Baran and Kint.[1,2] The classic findings include a longitudinal yellow and thickened nail plate segment with transversal curvature of the discolored segment, with filamentous projections of the nail matrix creating visible cavities at the free end of the nail plate.[3] Diagnosis of onychomatricoma involves a combination of gross appearance, dermoscopy, and histopathologic findings.[2] Under dermoscopy, honeycomblike cavities are seen at the free end of the nail plate.[4] Histopathologic findings include 2 distinct regions of tumor. The proximal zone, involving the nail matrix beneath the proximal nail fold, is characterized by an epithelial proliferation that appears to invaginate into the dermis, where bland spindle cell proliferation is also found. The distal zone, defined by the borders of the lunula, has the appearance of fibroepithelial projections consisting of matrix epithelium overlaying a loose spindle cell stroma.[5]

Pigmented onychomatricomas represent a rare variant of this rare disease, with only 8 cases previously reported total in the literature.[6,7] They present as a single-digit longitudinal melanonychia with a broad differential diagnosis, including subungual melanoma, pigmented squamous cell carcinoma in situ, pigmented onychopapilloma, and subungual hematoma.[6,7] Review of 8 published cases of pigmented onychomatricomas[6-8] notes that 3 cases were associated with prior trauma to the affected nail, and 3 cases had a history of treatment of the affected nail for onychomycosis; however, the cause of this lesion is unknown. Biopsy of pigmented onychomatricomas demonstrates epithelial extensions into the dermis and proliferation of dermal spindle cells, the latter of which can stain positive for CD34 immunoperoxidase. Importantly, no evidence of melanocyte proliferation is found, which helps to distinguish this entity from subungual melanoma.[7] Staining with cytokeratin immunoperoxidase can aid in identifying the epithelial component of the tumor.[7] Pigmentation of the nail is thought to be secondary to activation of the melanocytes in the nail matrix.[8]

This case presented with high suspicion for subungual melanoma, and consultation was sought from a nail specialist to obtain an appropriate biopsy. Proper orientation of the specimen for longitudinal sectioning is important to facilitate the correct diagnosis of pigmented onychomatricomas by the dermatopathologist.[7] Long-term follow-up of onychomatricoma in the literature shows that the tumor responds well to complete surgical excision.[9] Long-term prognosis for pigmented onychomatricomas is unknown owing to the small number of case reports and limited follow-up.

ARTICLE INFORMATION

Author Affiliations: Department of Dermatology, University of Texas Southwestern Medical Center, Dallas.

Corresponding Author: Lisa Garner, MD, Department of Dermatology, University of Texas Southwestern Medical Center, 5323 Harry Hines Blvd, Dallas, TX 75390 (lisa@lisagarnermd.com).

Published Online: August 15, 2018. doi:10.1001/jamadermatol.2018.1894

Conflict of Interest Disclosures: None reported.

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