JAMA Dermatology Clinicopathological Challenge

An Unusually Shaped Mass Over the Proximal Nail Fold

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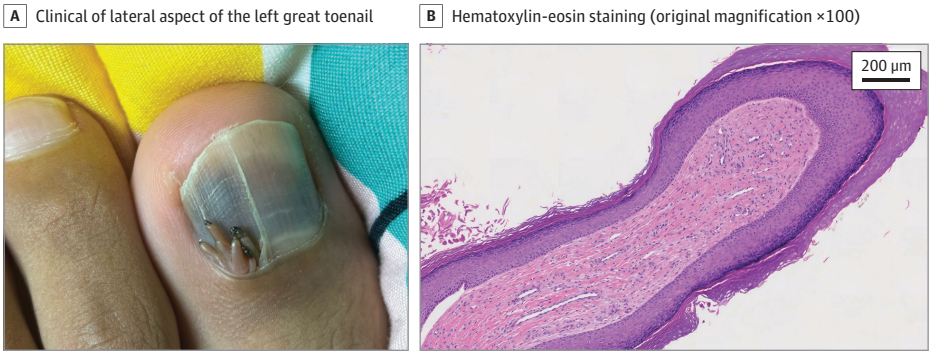


Figure 1. A, Initial clinical presentation over the lateral aspect of the left great toenail displaying 5 distinct keratotic tips, basal collarette, a longitudinal nail split, and an intact surrounding nail plate. B, High-power microscopy on a single projection showing hyperkeratosis and acanthosis with a central, longitudinally-oriented collagen core (hematoxylin-eosin; original magnification ×100).

A 23-year-old male marathon runner presented with an asymptomatic, progressive growth over the lateral aspect of his left great toenail 1 year prior, directly following a marathon. Physical examination revealed a firm, flesh-colored, 0.8 × 0.5 × 0.3-cm^3 solitary periungual nodule beneath the eponychium, featuring 5 distinct fingerlike keratotic projections forming a basal collarette and a longitudinal nail split (**Figure 1**A). The surrounding nail plate remained otherwise normal. The patient had no remarkable medical history, no family history of neurocutaneous disorders, and no other stigmata of tuberous sclerosis. A primary germinal-sparing narrow spindle excision with a partial nail avulsion was performed under digital block, clearing the slender anchoring stalk and collarette with an excision down to the periosteum. The patient showed a completely healed nail bed with no recurrence on follow-up.

WHAT IS YOUR DIAGNOSIS?

- **A.** Koenen tumor
- **B.** Cellular digital fibroma
- **C.** Multibranched acquired periungual fibrokeratoma
- **D.** Verruca vulgaris (periungual warts)

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Diagnosis

C. Multibranched acquired periungual fibrokeratoma

Microscopic Findings and Clinical Course

This multibranched acquired periungual fibrokeratoma (MAPFK) demonstrated typical hyperkeratosis and acanthosis overlying a dense, central core of longitudinally oriented collagen bundles and prominent interspersed thin-walled, dilated microvasculature (Figure 1B). The initial fleshy nodular growth in this case grew to a multibranched exophytic mass with dystrophic changes within 1 year.

Discussion

MAPFK represents a rare topomorphological variant of acquired digital fibrokeratomas[1-3]. MAPFKs are characterized by distinctive multiple filiform keratotic projections arising from a solitary nail unit lesion and a basal collarette.[4] The pathogenesis is strongly associated with localized chronic mechanical friction, which stimulates hyperplasia within the germinal matrix and growth through the perionychium.[5] Differentiating a solitary MAPFK from its clinical mimics is essential to avoid unnecessary radical treatments or missed systemic diagnoses.[5]

Koenen tumors and fibrokeratomas are distinct clinical entities with different aetiopathogenesis, occurrence and progression patterns, topomorphology, gross appearance, and syndromic associations.[5,6] MAPFKs arise sporadically as isolated acral lesions originating from within the nail matrix, remaining deeply implanted in the nail root, and finally emerging from the perionychium.[4-6] The diagnosis of MAPFK was supported by the characteristic clinical morphology and histopathological findings, together with the absence of clinical features or family history suggestive of tuberous sclerosis complex (TSC).[3,5,7]

Despite the clinical differences between Koenen tumors and fibrokeratomas, they share similar histopathological and immunohistochemical features.[5] Immunohistochemistry showed positive

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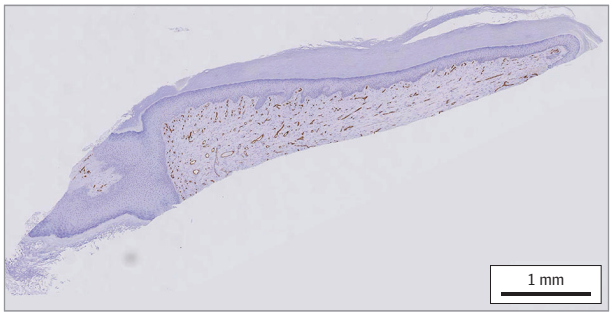


Figure 2. CD34 immunohistochemical staining highlighting focal, prominent microvascular endothelial proliferation within the stromal core.

staining of vessels with CD34 (**Figure 2**) and negative labeling with CD68, desmin, and S100; the latter suggested against a histiocytic process as well as smooth muscle or neural differentiation. The absence of diffuse CD34 positivity suggests against cellular digital fibroma or superficial acral fibromyxoma.[3,5]

Differentials for a solitary MAPFK include Koenen tumors, cellular digital fibroma, and verruca vulgaris. While histopathologically indistinguishable from MAPFK, true Koenen tumors present as part of TSC as multiple discrete, soft, rounded, fleshy digital growths between ages 12 and 14 years, increasing progressively in size and number and resembling fibrokeratomas when hyperkeratotic.[6] These acral lesions are associated with multifocal angiofibromatosis (eg, on the face or back), syndromic involvement (eg, cardiac, neural, or kidney), or positive family history.[6,7] When 2 or more Koenen tumors appear, they fulfill a major diagnostic criterion of TSC, but isolated solitary tumors rarely present as the only evidence.[6,7] Cellular digital fibroma typically present as flat, dome-shaped, or well-circumscribed small nodules lacking the pedunculated protrusion of an acquired digital fibrokeratoma, basal collarette, or microtraumatic causal link. A true cellular digital fibroma is defined by being diffusely and strongly CD34 positive across its highly cellular sheets of spindle cells, while this MAPFK only showed focal CD34 positivity restricted to the microvascular endothelial architecture within its stromal core.[8] Finally, verruca vulgaris are the most common cause of rough, hyperkeratotic periungual masses. Filiform warts can mimic the fingerlike protrusions of an MAPFK. However, dermoscopy reveals tiny black dots (thrombosed capillaries), and histopathology reveals classic human papillomavirus features, like koilocytosis.[3,5,8]

JAMA Dermatology published a landmark index case of a multibranched variant by Moriue et al[4] in 2014. This case expands the global literature by documenting a rare 5-branched presentation following specific athletic repetitive microtrauma with confirmatory immunohistochemistry. Following primary spindle (tapered elliptical) excision down to the periosteum, the anchoring stalk was cleared, successfully preserving the remaining germinal matrix with a uniformly healed proximal nail fold during follow-up at 2 months.[9,10] While the patient subsequently reported a suspected focal tissue alteration at 6 months, surgical excision is curative in 90% of cases,[5] and clinical follow-up and confirmation are required to definitively exclude keloidal scarring or true recurrence.

ARTICLE INFORMATION

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