Onychomatricoma: A Rare Case of Unguioblastic Fibroma of the Fingernail Associated With Trauma

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PRACTICE **POINTS**

- · Onychomatricoma is a rare benign neoplasm of the nail matrix that actively produces a nail plate.
- Onychomatricoma should be in the differential diagnosis of a thickened discolored nail plate with transverse overcurvature.
- · Onychomatricoma has been associated with onychomycosis and trauma to the nail apparatus.

Onychomatricoma (OM) is a rare tumor originating from the nail matrix. Fingernail involvement is twice as common as toenail involvement. Onychomatricoma is the only tumor that actively produces a nail plate. Clinically, it presents with yellow discoloration along the entire nail plate, proximal splinter hemorrhages, and a tendency toward transverse overcurvature of the nail plate with prominent longitudinal ridging. We report a rare case of OM in which the etiology was associated with trauma. Onychomycosis can be present, which may serve as a predisposing factor or be secondary to the deformed nail plate. Histologically, the tumor is fibroepithelial or biphasic with stromal and epithelial components. Onychomatricoma has been further classified into 3 types, including unguioblastoma, unguioblastic fibroma, and atypical unguioblastic fibroma. Our case represents the unguioblastic fibroma type.

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nychomatricoma (OM) is a rare benign neoplasm of the nail matrix. Even less common is its possible association with both trauma to the nail apparatus and onychomycosis. This case illustrates both of these findings.

Case Report

A 72-year-old white man presented to the dermatology clinic with a 26-year history of a thickened nail plate on the right third finger that had developed soon after a baseball injury. The patient reported that the nail was completely normal prior to the trauma. According to the patient, the distal aspect of the finger was directly hit by a baseball and subsequently was wrapped by the patient for a few weeks. The nail then turned black and eventually fell off. When the nail grew back, it appeared abnormal and in its current state. The patient stated the lesion was asymptomatic at the time of presentation.

Physical examination revealed thickening, yellow discoloration, and transverse overcurvature of the nail plate on the right third finger with longitudinal ridging (Figure 1). A culture of the nail plate grew Chaetomium species. Application of topical clotrimazole for 3 months followed by a 6-week course of oral terbinafine produced no improvement. The patient then consented to a nail matrix incisional biopsy 6 months after initial presentation. After a digital nerve block was administered and a tourniquet of the proximal digit was applied, a nail avulsion

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was performed. Subsequently, a 3-mm punch biopsy was taken of the clinically apparent tumor in the nail matrix.

On microscopic examination of the removed tissue, a benign mixed epithelial and stromal proliferative lesion was noted. The basaloid epithelium, lacking a granular layer, arose from the surface epithelial layer and formed a reticulated pattern extending into the stromal component, which was moderately cellular with spindle to fusiform nuclei dissecting between collagen bundles arranged in parallel arrays (Figure 2). The stromal component predominated over the epithelial component in this neoplasm. The nail was preserved in formalin and underwent hematoxylin and eosin staining. It was thickened and grossly showed filiform fibrous projections extending into the nail plate. Histologically, the nail displayed prominent oval clear channels. Periodic acid-Schiff staining was negative for fungal organisms.

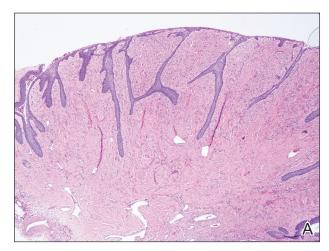
A diagnosis of unguioblastic fibroma—type OM was made. After receiving the diagnosis, expected course, and treatment options, the patient was offered conservative surgical excision but preferred clinical monitoring. At his last visit (6 months after the biopsy), the nail plate distal to the biopsy site had thinning and improvement, while the nail plate

Figure 1. Thickening, yellow discoloration, and transverse overcurvature of the nail plate on the right third finger with longitudinal ridging.

distal to the matrix that was not removed continued to show thickening, yellow discoloration, overcurvature, and longitudinal ridging (Figure 3).

Comment

Onychomatricoma is a rare tumor originating from the nail matrix. The tumor was first described by Baran and Kint¹ in 1992 using the term *onychomatrixoma*, but later the term *onychomatricoma* became more widely used.² Onychomatricomas are more common in adults (mean age, 48 years) and white individuals with no gender predilection.^{3,4} Fingernail involvement is twice as common as toenail involvement.³ Onychomatricoma is the only tumor that actively produces a nail plate.⁴



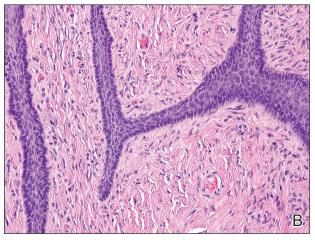


Figure 2. The basaloid epithelium arose from the surface epithelial layer and formed a reticulated pattern extending into the stromal component (A)(H&E, original magnification $\times 2$). At higher magnification, the stromal component was moderately cellular with spindle to fusiform nuclei dissecting between collagen bundles arranged in parallel arrays (B)(H&E, original magnification $\times 10$).

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Figure 3. Six months following the biopsy, the nail plate distal to the biopsy site had thinning and improvement, while the nail plate distal to the matrix that was not removed continued to show thickening, yellow discoloration, and overcurvature with longitudinal ridging.

Clinically, OM presents with yellow discoloration along the entire nail plate and proximal splinter hemorrhages. It has a tendency toward transverse overcurvature of the nail plate with prominent longitudinal ridging.⁴ Trauma has been associated in at least 3 cases reported in the literature, though the association was sometimes weak.^{3,4} Xanthonychia and onychodystrophy of the nail are common.³ Pterygium, melanonychia, nail bleeding, and cutaneous horns have been reported but are rare.³⁻⁵ The tumor typically is painless with no radiographic bone involvement.³ Onychomycosis can be present,³ which may either be a predisposing factor for the tumor or secondary due to the deformed nail plate.⁴

When the nail plate is avulsed and the proximal nail fold is turned back, the matrix tumor is exposed. This polypoid and filiform tumor has characteristic fingerlike fibrokeratogenous projections extending from the nail matrix into the nail plate.³

Histologically, the tumor is fibroepithelial or biphasic with stromal and epithelial components. It has a lobulated and papillary growth pattern with 2 distinct areas that correspond to 2 anatomic zones.³ The base of the tumor corresponds to the proximal anatomic zone, which begins at the root of the nail and extends to the cuticle. This area is composed of V-shaped keratinous zones similar to the normal matrix. If the nail is removed prior

to excision, these areas can be avulsed, leaving clear clefts. The superficial aspect of the tumor corresponds to the distal anatomic zone, which is located in the region of the lunula. This area is composed of multiple digitate or fingerlike projections with a fibrous core and a thick matrical epithelial covering.³ These digitations extend into small cavities in the nail plate, which can be visualized as clear channels or woodwormlike holes in hematoxylin and eosin–stained specimens. A biphasic fibrous stroma also can be observed with the superficial dermis being cellular with fibrillary collagen and the deep dermis more hypocellular with thicker collagen bundles.^{3,4}

An analysis of keratins in the nail matrix, bed, and isthmus showed that OM has the capacity to recapitulate the entire nail unit with differentiation toward the nail bed and isthmus.⁶ It appears that the mesenchymal component has an inductive effect that can lead to complete epithelial onychogenic differentiation.⁶

Due to the histological differences among the described cases of OM in the literature, a new classification based on the spectrum of epithelial to stromal ratio of stromal cellularity and the extent of nuclear pleomorphism was proposed in 2004.⁷ The prominent feature of the unguioblastoma type of OM is epithelial, while the cellular stroma is the prominent feature in the unguioblastic fibroma type. Atypical unguioblastic fibroma refers to a tumor with increased mitotic activity and nuclear pleomorphism among the stroma.⁷

Most OM tumors follow a benign clinical course; however, complete excision is advised to include the normal nail matrix proximal to the lesion, which may prevent recurrence and serves as a primary treatment.

Conclusion

Onychomatricoma is a benign neoplasm of the nail matrix that may be triggered by trauma; however, due to the weak association, further observations and studies should be conducted to substantiate this possibility. Patients with the classic clinical presentation possibly may be spared a nail avulsion and biopsy. Onychomycosis occurs in the setting of OM, and culture and treatment are unlikely to change the appearance or course of this nail condition.

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