Acquired Digital Fibrokeratoma Presenting as a Painless Nodule on the Right Fifth Fingernail

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Case Report
A 53-year-old woman presented for an initial visit to the dermatology clinic for a growth under the right fifth fingernail of 1 year’s duration. She had no history of trauma to the digit or pain or bleeding. She self-treated with over-the-counter wart remover for several months without improvement. She reported no other skin concerns. She had a medical history of rheumatoid arthritis (RA) and basal cell carcinoma of the nose; she was taking methotrexate and adalimumab for the RA. She had a family history of melanoma in her father.

On physical examination, a firm nontender nodule was noted on the distal nail bed of the right fifth fingernail with onycholysis; the nail plate was otherwise intact (Figure 1). All other nails were normal. A plain radiograph of the involved digit showed no bony abnormality. Excisional biopsy of the nodule was performed and analyzed by histopathology (Figure 2). The biopsy specimen showed a benign epidermis that was acanthotic and surmounted by hyperkeratotic scale. The dermis was fibrotic with collagen bundles assuming a vertical orientation to the long axis of the epidermis. The dermis was fibrotic with collagen bundles assuming a vertical orientation to the long axis of the epidermis, typical of a fibrokeratoma. There were no atypical features in the dermal component or epidermis (Figure 2). These findings were consistent with the diagnosis of acquired digital fibrokeratoma (ADF). The patient tolerated excisional biopsy well and had no evidence of recurrence 4 months following excision.

Comment
History and Clinical Presentation—First described by Bart et al in 1968, ADF is a rare benign fibrous tumor localized to the nail bed or periungual area. Typically, it presents as a solitary flesh-colored papule measuring 3 to 5 mm in diameter. It can be keratotic with a surrounding collarette of elevated skin. Acquired digital fibrokeratoma is usually localized to the digits of the hands or feet; when presenting subungually, it is more commonly found arising from the proximal matrix or nail bed of the great toe. Observed nail changes include longitudinal grooves, trachyonychia, subungual hyperkeratosis, and onycholysis.

PRACTICE POINTS
• Acquired digital fibrokeratoma is a benign tumor of the nail bed and periungual area.
• Histopathology shows epidermal acanthosis and hyperkeratosis, and collagen bundles are arranged in a vertical orientation to the long axis of the epidermis.
• Acquired digital fibrokeratoma should be considered in the differential diagnosis of flesh-colored papules on the nail unit associated with longitudinal grooves, trachyonychia, subungual hyperkeratosis, and onycholysis.

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Acquired digital fibrokeratoma is more commonly found in middle-aged men; however, it has been reported among patients of various ages and in both sexes. In a study of 20 cases, the average duration before presenting for medical advice was 28 months. Acquired digital fibrokeratoma arises sporadically; some patients report prior local trauma. Lesions typically do not self-resolve.

**Diagnosis**—The diagnosis of ADF is made using a combination of clinical and histopathological findings. Dermoscopy is helpful and may show homogenous white or milky white structures, likely representing hyperkeratosis, proliferation of capillaries, and an increase in collagen bundles with a surrounding collarette of scale. Histopathology shows acanthosis and hyperkeratosis of the epidermis. Collagen bundles assume a characteristic vertical orientation to the long axis of the epidermis.

Two other histomorphologic subtypes, less common than the type I variant, are the type II variant, in which the number of fibroblasts is increased and the number of elastic fibers is decreased, and the type III variant, in which the stroma are edematous and cell poor. There is an even greater reduction in elastic tissue content in the type III variant than in the type I variant. There is evidence that type II ADFs exhibit more hyperkeratosis clinically than the other 2 subtypes, but from a practical perspective, this subclassification is not conducted in routine practice because it does not have clinical significance.

**Differential Diagnosis**—The clinical differential diagnosis of ADF is broad and includes squamous cell carcinoma, onychomatricoma, onychopapilloma, verruca vulgaris, supernumerary digit, neurofibroma, cellular grooves, trachyonychia, subungual hyperkeratosis, and onycholysis. The affected nail can be painful, depending on the size and location of the tumor.

**FIGURE 1.** Acquired digital fibrokeratoma. A, Physical examination of the right fifth fingernail revealed moderate onycholysis but an otherwise intact nail plate. B, Close view of the right fifth fingernail revealed a firm nontender nodule on the distal nail bed.

**FIGURE 2.** A, Histopathologic analysis of an excisional biopsy showed a verrucous and acanthotic epidermis surmounted by a thick hyperkeratotic stratum corneum (H&E, original magnification ×4). B, Higher magnification showed sclerotic-appearing collagen bundles assuming a vertical orientation to the long axis of the epidermis (H&E, original magnification ×20).
digital fibroma, and Koenen tumor (periungual fibroma). Almost all of these entities are easily differentiated from ADF on biopsy. A fibrokeratoma does not exhibit the atypia seen in squamous cell carcinoma. The multiple fibroepithelial projections and nail plate perforations characteristic of onychomatricoma are not observed in ADF. Onychopapilloma shows acanthosis and papillomatosis, similar to ADF; however, onychopapilloma lacks the characteristic vertical orientation of collagen in ADF. Verruca vulgaris classically shows koilocytosis, dilated blood vessels in papillae, and hypergranulosis. A supernumerary digit clinically lacks a collarette of scale and often presents in a bilateral fashion on the lateral fifth digits in children; histopathologically, a supernumerary digit is distinct from an ADF in that nerve bundles are abundant in the dermis, defining a form of amputation neuroma. Neurofibroma exhibits a spindle cell proliferation that assumes a patternless disposition in the dermis, accompanied by mucin, mast cells, and delicate collagen. The defining cell populace has a typical serpiginous nuclear outline that is characteristic of a Schwann cell. Cellular digital fibroma can present similar to ADF; it is considered by some to be a mucin-poor variant of superficial acral fibromyxoma. Its morphology is distinct: a proliferation of bland-appearing spindled cells exhibiting a storiform or fascicular growth pattern and CD34 positivity.

The differential diagnosis to consider when ADF is suspected is a Koenen tumor, which resembles a fibrokeratoma clinically and also is localized to the digits. Koenen tumors can be differentiated from fibrokeratoma by its association with tuberous sclerosis; a multiple, rather than solitary, presentation; a distinctive clove-shaped gross appearance; and an appearance on histopathology of stellate-shaped fibroblasts with occasional giant cells. Despite these important differences, Koenen tumor does exhibit a striking morphologic similarity to ADF, given that the vertical orientation of collagen bundles in Koenen tumor is virtually identical to ADF.

**Management**—There are no known associations between ADF and medication use, including methotrexate and adalimumab, which our patient was taking; additionally, no association with RA or other systemic disorder has been reported. The preferred treatment of ADF is complete excision to the basal attachment of the tumor; recurrence is uncommon. Alternative therapies include destructive methods, such as cryotherapy, CO₂ laser ablation, and electrodesiccation.

**REFERENCES**