Multiple Acquired Periungual Fibrokeratoma

Yoon Seob Kim, Ji Hyun Lee, Young Min Park, Jun Young Lee

Department of Dermatology, Seoul St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Seoul, Korea

Dear Editor:

Acquired periungual fibrokeratoma (APF) is a rare and benign tumor that usually presents as a solitary, fibrous, and hyperkeratotic lesion¹. Multiple APF is rarely reported, and to our knowledge, symmetrical bilateral distribution of multiple APF is not reported. Herein, we describe a rare case of multiple APF, which could be misdiagnosed as Koenen tumors.

A 19-year-old Korean girl presented for evaluation of periungual tumors detected 9 months previously. Physical examination revealed asymptomatic, four, firm, skin-colored tumors associated with longitudinal grooves of subjacent nails tumors were present on both great toes (Fig. 1). She denied any recent trauma history. There was no specific past medical or family history of dermatological, neurological, or other specific systemic conditions. A shave biopsy specimen of the lateral nodule from the left great toe

revealed hyperkeratosis, acanthosis, and hypergranulosis of the epidermis (Fig. 2A). Excisional specimen revealed a thick collagen bundle with capillary proliferation arranged parallel to the axis of the tumor (Fig. 2B, C). Three other lesions were completely excised, and histopathologic findings were the same as those of the left great toe. Based on these clinical and histological findings, she was diagnosed with multiple APF. There was no recurrence at the 6-month follow-up.

APF typically presents as a solitary lesion originating from the proximal nail fold or the nail bed, and more specifically, from the proximal margin of the germinal matrix². The pathogenesis is unclear; however, trauma and factor XIIIa positive dermal dendrocytes have been suggested as possible causal factors¹. Histopathologic findings indicate that hyperkeratosis, acanthosis, and focal hypergranulosis of the epidermis, and the presence of thick bundles of col-

Fig. 1. Physical examination revealed asymptomatic, four, firm, skin-colored tumors with longitudinal grooves of subjacent nails on both great toes.

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Corresponding author: Jun Young Lee, Department of Dermatology, Seoul St. Mary's Hospital, College of Medicine, The Catholic University of Korea, 222 Banpo-daero, Seocho-gu, Seoul 06591, Korea. Tel: 82-2-2258-6222, Fax: 82-2-599-9950, E-mail: jylee@catholic.ac.kr

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Fig. 2. (A) A shave biopsy specimen of the lateral nodule from the left great toe revealed hyperkeratosis, acanthosis, and hypergranulosis of the epidermis. (B, C) Excisional specimen revealed a thick collagen bundle with capillary proliferation arranged parallel to the axis of the tumor (H&E; A: $\times 20$, B: $\times 40$, C: $\times 100$).

lagen with dilated capillaries orientated along the longitudinal axis of the tumor in the dermis¹. The differential diagnoses of APF include Koenen tumors, supernumerary digits, neurofibroma, verruca vulgaris, and cutaneous horn^{3,4}. Of these, Koenen tumors, which typically present with multiple periungual fibromas, should be excluded in multiple APF. Koenen tumors are observed in approximately 50% of tuberous sclerosis complex (TSC) cases, and usually occur during puberty². The presence of conspicuous, atypical, and stellate myofibroblasts within the fibrous core of Koenen tumors is distinct from the histopathologic features of APF². In addition, hematomas or typical signs of TSC, such as adenoma sebaceum, shagreen patches, or ash leaf macules, usually coexist with Koenen tumors^{2,4}. The preferred management of APF is total excision, and germinal matrix should be preserved in order to prevent nail dystrophy¹. Local recurrence is rare, but could occur when partial excision or curettage is per-

formed². The interest of our case is the rare clinical presentation of bilateral and symmetrical distribution of APF.

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