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CASE REPORT

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An unusual presentation of acquired periungual fibrokeratoma: A mixed form of dome-shaped and branching type

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Abstract

Acquired periungual fibrokeratoma is an uncommon benign fibrous tumour occurring in the periungual area, predominantly the proximal nail fold. We describe a case of acquired periungual fibrokeratoma with an unusual presentation of mixed features including dome-shaped and branching type in order to facilitate the recognition of this variant.

KEYWORDS

fibroma, fibrous tissue, keratosis, nails, neoplasms, toes

1 | INTRODUCTION

Acquired periungual fibrokeratoma is an uncommon benign fibrous tumor occurring in the periungual area, predominantly the proximal nail fold. It is considered as the topographical variant of acquired digital fibrokeratoma (ADFK), which was first described by Bart et al in 1968.[1] Classic ADFK usually manifests as an asymptomatic, solitary, flesh-colored papule or a nodule less than 1 cm in diameter with a characteristic hyperkeratotic collarette at its base, but this variant has more diversities in morphological features, such as dome-shaped, rod-shaped/branching, and flat types,[2] and mostly manifests as a single feature of these types.

A diagnosis of acquired periungual fibrokeratoma is made based on clinical features and characteristic histological findings; however, it often presents diagnostic challenges to clinicians because of its rarity and morphological diversities. Herein, we report a case of acquired periungual fibrokeratoma with an unusual presentation of mixed features of dome-shaped and branching types in order to facilitate the recognition and proper management of this variant.

2 | CASE REPORT

A 67-year-old man presented with an asymptomatic, dome-shaped mass with multiple elongated projections on his left second toe. The physical examination revealed a garlic-shaped, pedunculated mass with multiple branches at its base (Figure 1A). It measured 1.3 × 0.9 × 0.7 cm and had a smooth surface. The color of the tumor's surface was variegated from a violaceous hue to dark purple. Laboratory and physical examinations showed no remarkable findings. According to his medical history, it recurred with similar features to those of the tumor that was treated surgically at a local clinic 1 year ago, and not associated with previous trauma. The lesion was managed simply with shave excision. The surgical wound healed well without repair. The intraoperative findings revealed that the tumor originated from the proximal nail fold; the eponychium and proximal nail plate were partially injured (Figure 1B), and the undersurface of the tumor had a very tiny base (Figure 1C). His postoperative courses were uneventful without recurrence until the 6-month follow-up.

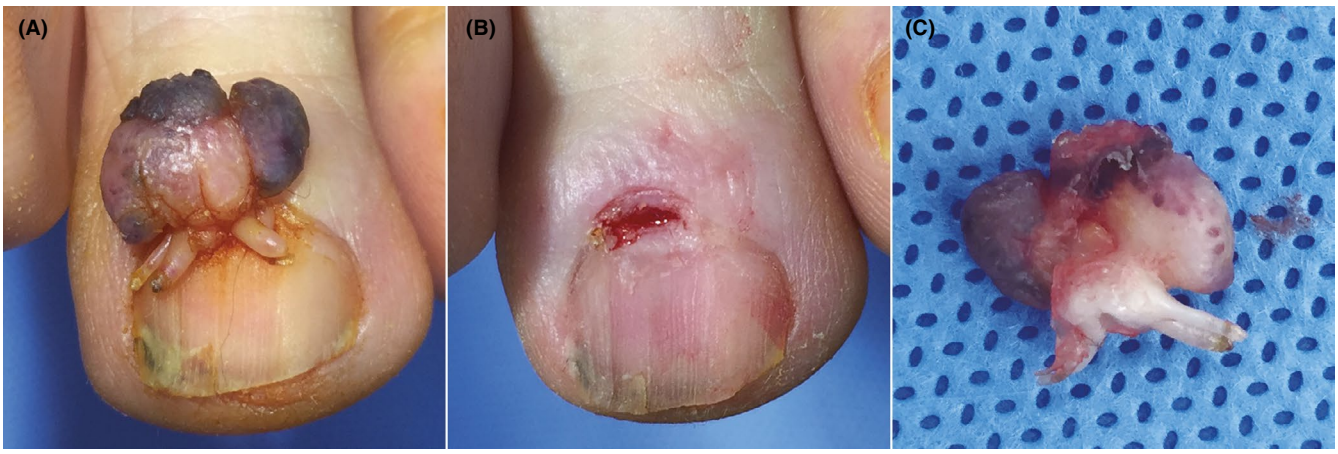

FIGURE 1 A, Preoperative photograph showing the unique features of acquired periungual fibrokeratoma: the garlic-shaped mass and multiple elongated projections at its base. B, Postoperative photograph showing a tiny open wound at the proximal nail fold. C, The undersurface of the resected tumor specimen shows a narrow tumor base (*)

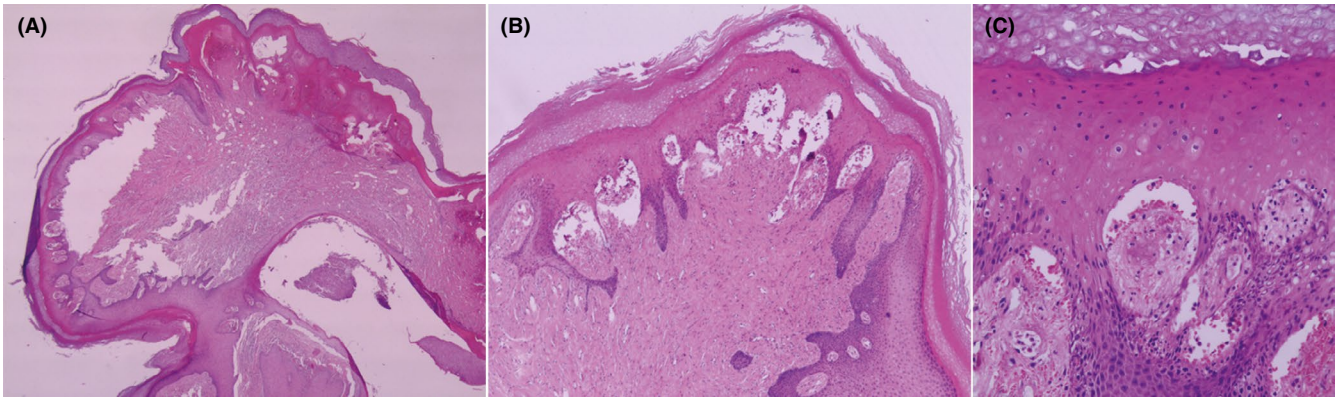

FIGURE 2 Histological examination of the resected tumor. A, A papillary lesion is shown (hematoxylin and eosin [H&E], ×15). B, The dermis comprises vertically oriented collagen bundles and focal dilated blood vessels along with epidermal hyperkeratosis and acanthosis (H&E, ×40). C, Dermal papillary necrosis and hemorrhage are shown (H&E, ×200)

The histopathologic examination revealed a benign papillary lesion (Figure 2A). The epidermis showed hyperkeratosis, parakeratosis, and acanthosis. The dermal lesion comprised thick collagen bundles with arrangement parallel to the axis of the tumor and the proliferation of fibroblasts (Figure 2B). Focal papillary dermal necrosis and hemorrhage, suggesting that the phenomenon was secondary to repeated trauma, were also observed in the upper dermis (Figure 2C). Based on clinical and histopathologic findings, the patient was diagnosed with acquired periungual fibrokeratoma.

3 | DISCUSSION

Classic ADFK is commonly found on the fingers or toes. However, it occurs occasionally on nondigital areas of the hands and feet as well as the elbow, forearm, wrist, and exceptionally on the heel, knee, lower leg, thigh, and buttock.[3] Because of its predominant location on the fingers and toes, Verallo[4] described it as acral fibrokeratoma. However, acquired fibrokeratoma seems to be a more appropriate description rather than ADFK, as several authors already pointed out the unsuitability of the original nomenclature because its occurrence is not limited to the digits.

ADFK found on the digit, especially in the nail apparatus, such as the nail fold or nail bed, was named acquired ungual fibrokeratoma by Cahn[5] in 1977, and this term encompasses two variants of periungual and subungual fibrokeratoma depending on the location of the tumor. In their retrospective study of 20 patients, Hwang et al[2] classified acquired ungual fibrokeratoma as the periungual, intraungual, and subungual types, and they described morphologic types of the tumors: rod-shaped, dome-shaped, flat, and branching. In their study, the periungual type (15/20) occurred most frequently, followed by intraungual (4/20) and subungual (1/20). All of their cases of the periungual type occurred at the proximal nail folds. They reported that tumors occurred more in toes than in fingers with predilection of the greater toe. Of the 15 periungual fibrokeratomas, a dome-shaped lesion (7/15) was most common, followed by branching type (4/15), flat type

(2/15), and rod type (2/15) lesions. Kim et al[6] also classified acquired ungual fibrokeratoma into four clinical types: pine-pit-like, dome-shaped, finger-like, and chicken's foot-like. However, to our knowledge, the unique features such as a mixed form of the dome-shaped and branching types shown in our case have never been reported in the literature.

ADFK usually manifests as a solitary lesion, but may be multiple like Koenen's tumors in patients with tuberous sclerosis or in other rare cases.[7] Koenen's tumors are observed in approximately 50% of patients with tuberous sclerosis, and manifest as pedunculated, flesh-colored lesions in periungual and subungual areas.

The etiology of ADFK is still unknown, although trauma or repeated irritation seems to be a contributing factor. Kint et al[8] suggested that the condition develops from the neoformation of collagen by fibroblasts, whereas Nemeth et al[9] hypothesized that large stellate fibroblasts expressing factor XIIIa play an important role in the formation of fibrovascular tumors, including fibrokeratoma. Suh et al[10] also demonstrated an increased number of factor XIIIa-positive dermal dendritic cells in his case of acquired digital fibrokeratoma, which supports that hypothesis.

The histopathologic examination of the tumor typically reveals a hyperkeratotic epidermis with irregular acanthosis. The core of the lesion comprises thick, interwoven collagen bundles, mostly orienting along the vertical axis of the lesion with dilated capillaries. On the basis of the histologic examination of 50 cases of acquired digital fibrokeratoma, Kint et al[8] disclosed three histologic variants of these lesions. The first variant is a dome-shaped lesion that contains a core of thick, densely packed collagen bundles with fine elastic fibers. The second variant is a mainly tall, hyperkeratotic lesion that contains more fibroblasts in the cutis that are more regularly arranged than those in the first variant with reduced elastic fibers. The third variant is a flat to dome-shaped lesion with poor cellular structure and absent elastic fibers. The epidermis of all variants showed the same features of acanthosis and hyperkeratosis. The first variant was most commonly found in Hwang et al[2] and Kint et al's studies.[8] The histopathologic findings of our patient were also suggestive of those of the first variant.

Although the diagnosis of the periungual variant is based on the clinical and histopathologic features, it should be differentiated from other conditions encountered in the digits such as verruca vulgaris, digital mucous cyst, eccrine poroma, supernumerary digit, superficial acral fibromyxoma, and Koenen's tumor to ensure proper diagnosis and management. The preferred treatment is complete excision because superficial removal usually results in recurrence.[2,3] The tumor in our case recurred once despite previous surgical excision. Therefore, close observation of its recurrence is needed after surgical treatment. Our case report described unique morphological features of acquired periungual fibrokeratoma, which will help facilitate the recognition and proper management of this variant.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

AUTHOR CONTRIBUTIONS

K-YS: conceptualized and designed the study, and revised the manuscript. SKL: contributed to definition of intellectual content. S-YL: collected data and drafted the initial manuscript. All authors: approved the final manuscript.

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