Case Report

Horn on the Nail: Acquired Ungual Fibrokeratoma

Noor Jahan, Padubidri Kombettu Ashwini, Santhebachall Gurumurthy Chethana, Jayadev Betkerur, Veeranna Shastry
Department of Dermatology, Venereology, and Leprology, JSS Medical College, JSS Academy of Higher Education and Research, Mysuru, Karnataka, India

Abstract

Acquired ungual fibrokeratomas are uncommon fibrous tissue tumors that are located in the ungual area. They usually present as asymptomatic, solitary, smooth, dome-shaped, or fingerlike, flesh-colored papules accompanying nail deformities, including a longitudinal groove and trachyonychia. Acquired periungual fibrokeratoma is considered a topographical variant of acquired digital fibrokeratoma; it has a unique “garlic clove” shape. A traumatic origin has been suggested, as acquired ungual fibrokeratomas occur most frequently on the fingers and toes. Herein, we report a case of a 29-year-old man with growth over left little finger, who was treated with surgical excision of the tissue. Histopathological examination confirmed ungual fibrokeratoma. It is a rare variant of digital fibrokeratoma, and so far the first ungual fibrokeratoma to appear de novo on little finger to the best of our knowledge.

Keywords: Garlic clove, horn like, ungual fibrokeratoma

Introduction

Acquired ungual fibrokeratoma is an uncommon benign fibrous tissue tumor located in the nail area. The term was first coined by Cahn in 1977.[1] He postulated that this was the same as the garlic clove fibroma, which was first identified by Steel in 1965.[1,2] Digital ungual fibrokeratoma is seen in all races, between 12 and 70 years of age, with peak in the middle age.[3] Higher incidence was seen in males than that in females. Acquired ungual fibrokeratomas usually present as asymptomatic, solitary, smooth, dome-shaped, or fingerlike, flesh-colored papules, accompanying nail deformities that include a longitudinal groove and trachyonychia.[4] It is a rare variant of digital fibrokeratoma.

Case Report

A 29 year old man presented with an asymptomatic growth over the left little finger since 18 months. There was no history of preceding trauma. It started as a small raised lesion projecting out from the center of proximal nail fold. Over a period of 18 months, it developed into a hornlike structure. There was no history of manipulation of lesion. No past history of similar skin lesions was reported.

On physical examination, solitary longitudinal firm fingerlike growth, emerging from the center of proximal nail fold toward distal part of nail, was observed. The distal tapering end of the growth appeared necrosed. The nail plate of the involved finger showed linear depression beneath the growth. The lesion was not adherent to underlying nail plate. Minimal tenderness was present. No bony deformity was apparent [Figures 1–4].

Following clinical differentials were considered—acquired ungual fibrokeratoma, onychomatricoma, Koenen tumor, and glomus tumor. An excision biopsy was planned. Under aseptic conditions, a proximal digital block was performed using 2% lignocaine without adrenaline. Incision was put at proximal nail fold, roofing the growth. The proximal nail fold was retracted to expose the nail matrix by skin hooks. The whole tumor was excised with its basal attachment. Tissue was sent for histopathological examination. Wound healed with secondary intention [Figure 4]. No recurrence was observed in 5 months.
following excision. Histopathology revealed keratinized stratified squamous epithelium with a prominent granular layer of thick orthokeratosis with superficial region showing fibrocollagenous matrix with increased blood vessels. Features were suggestive of fibrokeratoma.

**Discussion**

Acquired periungual fibrokeratomas are rare, benign, fibrous, and hyperkeratotic tumors,\(^1\,^5\) presenting as lesions that emerge from the proximal nail fold or sometimes from the nail bed.\(^6\,^7\) Although acquired periungual fibrokeratoma is considered a topographical variant of acquired digital fibrokeratoma, it has a unique “garlic clove” shape.\(^2\,^8\)

Including the word “acquired” in the terminology of this unusual tumor leads one to believe that there is a precipitating factor. However, the etiology is still unknown. Hare and Smith\(^9\) proposed that trauma is involved, whereas Nemeth and Penneys\(^10\) reported that factor XIIIa-positive dermal dendritic cells play an important role in the regulation of collagen synthesis.\(^9\,^12\) A traumatic origin has been suggested because acquired ungual fibrokeratomas occur most frequently on the fingers and toes.\(^13\) Acquired ungual fibrokeratomas usually present as asymptomatic, firm, smooth, flesh-colored papules, protruding laterally on the nail plate from beneath the cuticle of the proximal nail fold. Verruca vulgaris, a supernumerary digit, Koenen tumor of tuberous sclerosis, pyogenic granuloma, eccrine poroma, cutaneous horn, epidermoid cyst, acrochordons, and neurofibromas must be included in the differential diagnosis of acquired ungual fibrokeratoma.\(^9\,^13\,^15\)

Histopathological findings include hyperkeratosis, acanthosis, focal hypergranulosis, and irregular rete ridge elongation in the epidermis. The core of the tumor consists of thickened collagen in the dermis, oriented parallel to the long axis of the lesion [Figures 5 and 6]. In 1985, Kint et al.\(^16\) classified acquired digital fibrokeratomas into three histological subtypes. In all three, the epidermis is
acanthotic and hyperkeratotic. In the first variant, tumor cores are composed of thick, dense, closely packed, and irregularly arranged collagen bundles. The second variant shows an increase in grouped fibroblasts and reduced elastic fibers. The third variant is composed of a poorly cellular and edematous structure with completely absent elastic tissue. Our case showed a mixed variety of first and second variants.

Kint et al.\cite{16} divided acquired ungual fibrokeratomas into two types based on appearance, that is, dome shaped and fingerlike. However, more diverse morphologies such as rod shaped, dome shaped, flat, and branching are also seen. Kim et al.\cite{17} classified acquired ungual fibrokeratoma into four clinical types, namely pine pitlike, dome shaped, fingerlike, and chicken's footlike.

Kim et al.\cite{17} found that dome-shaped acquired ungual fibrokeratomas were the most common clinical type, our case was of fingerlike morphology. Acquired ungual fibrokeratomas should be considered in the differential diagnosis of periungual tumors with a longitudinal nail groove. The groove is thought to be due to the fibrokeratoma originating from the underside of the proximal nail fold, compressing the nail matrix.\cite{16}

Acquired ungual fibrokeratomas do not spontaneously regress.\cite{14} They should be excised when symptomatic or when it causes significant cosmetic problems. The preferred treatment is total excision because superficial removal usually results in recurrence.\cite{18} Various surgical techniques can be used to resect these tumors, depending on their size and location. Usually, the tumor is incised around its base and dissected from the bone. If a lesion is beneath the nail plate, partial or total removal of the nail is required first. Lesions above the nail plate, however, can be removed by shaving followed by phenolization, CO₂ laser vaporization, or surgical resection after lifting the proximal nail fold as a banner flap.\cite{19-21} During the procedure, the germinal matrix should be preserved by careful dissection along the under surface of the tumor.\cite{22,23} To minimize the risk of recurrence, it is recommended that careful elevation of lesion and retraction of the proximal nail fold to expose the whole tumor and surrounding nail matrix is performed, avoiding additional injury to the nail matrix.
Acquired ungual fibrokeratoma is a rare variant of digital fibrokeratoma. In this case, we found the lesion to be de novo (without any history of trauma), it appeared on little finger, which has not been reported so far to the best of our knowledge. We conclude that complete excision of tumor from base is the treatment of choice for the condition. This case has been reported for its rarity.

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Conflicts of interest
There are no conflicts of interest.

References
1. Cahn RL. Acquired periungual fibrokeratoma. A rare benign tumor previously described as the garlic-clove fibroma. Arch Dermatol 1977;113:1564-8.
2. Steel HH. Garlic-clove fibroma. JAMA 1965;191:1082-3.
3. Ali M, Mbah CA, Alwadiya A, Nur MM, Sunderamoorthy D. Giant fibrokeratoma, a rare soft tissue tumor presenting like an accessory digit, a case report and review of literature. Int J Surg Case Rep 2015;10:187-90.
4. Hwang S, Kim M, Cho BK, Park HJ. Clinical characteristics of acquired ungual fibrokeratoma. Indian J Dermatol Venereol Leprol 2017;83:337-43.
5. Verallo VV. Acquired digital fibrokeratomas. Br J Dermatol 1968;80:730-6.
6. Saito S, Ishikawa K. Acquired periungual fibrokeratoma with accessory germinal matrix. J Hand Surg Br 2002;27: 549-55.
7. Hashiro M, Fujiy Y, Tanaka M, Yamatodani Y. Giant acquired fibrokeratoma of the nail bed. Dermatology 1995;190: 169-71.
8. Yasuki Y. Acquired periungual fibrokeratoma—a proposal for classification of periungual fibrous lesions. J Dermatol 1985;12:349-56.
9. Hare PJ, Smith PA. Acquired (digital) fibrokeratoma. Br J Dermatol 1969;81:667-70.
10. Nemeth AJ, Penneys NS. Factor XIIIa is expressed by fibroblasts in fibrovascular tumors. J Cutan Pathol 1989;16:266-71.
11. Cerio R, Griffiths CE, Cooper KD, Nickoloff BJ, Headington JT. Characterization of factor XIIIa positive dermal dendritic cells in normal and inflamed skin. Br J Dermatol 1989;121:421-31.
12. Cerio R, Spaull J, Oliver GF, Jones WE. A study of factor XIIIa and MAC 387 immunolabeling in normal and pathological skin. Am J Dermatopathol 1990;12:221-33.
13. Altman DA, Griner JM, Faria DT. Acquired digital fibrokeratoma. Cutis 1994;54:93-4.
14. Berger RS, Spielvogel RL. Dermal papule on a distal digit. Acquired digital fibrokeratoma. Arch Dermatol 1988;124:1559-60, 1562-3.
15. Vinson RP, Angeloni VL. Acquired digital fibrokeratoma. Am Fam Physician 1995;52:1365-7.
16. Kint A, Baran R, De Keyser H. Acquired (digital) fibrokeratoma. J Am Acad Dermatol 1985;12:816-21.
17. Kim JM, Cho HH, Kim WJ, Mun JH, Song M, Kim HS, et al. Clinical characteristics of acral fibrokeratoma. Korean J Dermatol 2014;52:535-9.
18. Shelley WB, Phillips E. Recurring accessory “fingernail”: periungual fibrokeratoma. Cutis 1985;35:451-4.
19. Mazeira M, del Pozo Losada J, Fernández-Jorge B, Fernández-Torres R, Martínez W, Fonseca E. Shave and phenolization of periungual fibromas, Koenen's tumors, in a patient with tuberous sclerosis. Dermatol Surg 2008;34:111-3.
20. Berlin AL, Billick RC. Use of CO2 laser in the treatment of periungual fibromas associated with tuberous sclerosis. Dermatol Surg 2002;28:434-6.
21. Vazquez-Doval FJ. Surgery of the nail region. In: Atlas of nail diseases. Barcelona, Spain: ESMON Pharma; 2008. pp. 112-3.
22. Lee CY, Lee KY, Kim KH, Kim YH. Total excision of acquired periungual fibrokeratoma using bilateral proximal nail fold oblique incision for preserving nail matrix. Dermatol Surg 2010;36:139-41.
23. Yelamos O, Alegre M, García JR, Puig L. Periungual acral fibrokeratoma: Surgical excision using a banner flap.Acta Dermosifiliogr 2013;104:830-2.