

Clinical characteristics of acquired unguis fibrokeratoma

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Abstract

Background: Acquired unguis fibrokeratomas are uncommon fibrous tissue tumors located in the unguis area. Though there are many reports of this entity, only some reports have reviewed the clinical features of the tumor.

Aims: The aim of this study was to clarify the clinical characteristics of acquired unguis fibrokeratomas.

Methods: We reviewed twenty patients who were treated surgically at our clinic from 2003 to 2014 for acquired unguis fibrokeratomas confirmed on histopathological examination. Our study was conducted by retrospective analysis of charts, clinical pictures and patient records. Cases of tuberous sclerosis were not included.

Results: Acquired unguis fibrokeratomas occurred on toenails in 16 (80%) patients and on fingernails in 4 (20%) patients. Periunguis lesions were noted in 15 (75%) patients followed by intraunguis lesions in 4 (20%) patients and subunguis lesions in 1 (5%) patient. A longitudinal groove was observed in 80% of patients. Surgical resection was performed in all cases for both medical and cosmetic reasons. After excision, recurrence occurred in three cases.

Limitations: This was a retrospective study of a limited number of patients.

Conclusions: Acquired unguis fibrokeratomas occurred more commonly on toenails than on fingernails and were located in the periunguis area in most patients. A longitudinal groove in the nail plate was a frequent finding. Surgical resection led to medical and cosmetic improvement with a recurrence in 3 (15%) patients.

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Key words: Acquired unguis fibrokeratoma, benign fibrous tumor, nail

Introduction

Acquired unguis 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} Acquired unguis fibrokeratomas usually present as asymptomatic, solitary, smooth, dome-shaped or finger-like, flesh-colored papules. Accompanying nail deformities include a longitudinal groove and trachyonychia. There are many reports of acquired acral fibrokeratomas but only a few review their clinical features. This study looked at the clinical features, accompanying nail changes, response to treatment and follow-up in twenty patients with acquired unguis fibrokeratomas.

Methods

Twenty patients who were treated surgically at our clinic for acquired unguis fibrokeratomas from 2003 to 2014 were included. We

analysed patient charts, clinical pictures and patient records (which included sex, age, duration of illness, trauma history and history of previous treatment). Cases with tuberous sclerosis were not included. Acquired unguis fibrokeratomas were classified into four morphological types: Rod-shaped, dome-shaped, flat and branching [Figure 1]. They were also classified based on location as periunguis, subunguis and intraunguis (“dissecting unguis”) [Figure 2]. All patients were treated by complete surgical excision and the diagnosis confirmed by histopathologic examination of all excised lesions.

Results

The main clinical data are shown in Table 1 and Figure 3. Patient details are listed in Table 2.

The patients ranged in age from 15 to 79 years with a mean of 41.2 years. The male to female ratio was 10:10. Almost all patients

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Figure 1a: Rod-shape AUFG(Acquired unguial fibrokeratoma)



Figure 1b: Dome-shaped AUFG



Figure 1c: Flat AUFG



Figure 1d: Branching AUFG

(17/20) remembered when the lesion first appeared. The average delay for seeking medical advice was 28 months. Symptom duration was 40.4 months on average. A high proportion of patients were in their 40s (5/20). Three patients had lesion onset in childhood. Associated diseases observed included hypertension (2/20), diabetes mellitus (1/20) and gastric cancer (1/20). Four patients (20%) had a history of trauma with the lesion developing 2.5 years later on average.

All patients had a single lesion. Toenails (16/20, 80%) were involved more commonly than fingernails (4/20, 20%), most commonly the right great toenail. Tumors presented as asymptomatic, skin-colored, hyperkeratotic papules and the base of each lesion was surrounded by an epidermal collarette. In order of decreasing frequency, lesions were dome-shaped (8/20, 40%), branching (5/20, 25%), flat (4/20, 20%) and rod-shaped (3/20, 15%) [Figure 4a]. Distribution was periungual (15/20, 75%), intraungual (4/20, 20%) or subungual (1/20, 5%) [Figure 4b]. A longitudinal groove was the most commonly associated nail deformity (16/20, 80%). Also trachyonychia (8/20, 40%), chromonychia (4/20, 20%), onycholysis(4/20, 20%), subungual hyperkeratosis (3/20, 15%), and dissected nail plates (2/20, 10%) were observed.

The histopathological findings in our cases are summarized in Table 3.

All patients were treated by surgical excision [Figure 5a-d]. The mean post-excision follow-up period was 8.6 months. After excision, recurrence occurred in three cases, all of whom gave a history of trauma preceding the appearance of the tumors. During follow-up, only one patient had residual scarring around the proximal nail fold and almost all patients were satisfied with the postoperative result as they regained normal nail contours.

Discussion

Acquired periungual fibrokeratomas are rare, benign, fibrous, hyperkeratotic tumors.^{1,3} They usually present as lesions that emerge from the proximal nail fold or sometimes from the nail bed.^{4,5} Cahn proposed the term “acquired periungual fibrokeratoma” because these lesions were thought to be histologically identical to acquired digital fibrokeratomas.¹ Although acquired periungual fibrokeratoma is considered a topographical variant of acquired digital fibrokeratoma, it has a unique “garlic clove” shape.^{2,6}

Including the word “acquired” in the terminology of this unusual tumor leads one to believe that there is a precipitating factor. However, the etiology of acquired unguial fibrokeratoma is still unknown. Hare and Smith proposed that trauma is involved, while Nemeth and Penneys reported that factor XIIIa-positive dermal dendritic cells play an important role in the regulation of collagen

synthesis.⁷⁻¹⁰ A traumatic origin has been suggested because acquired unguial fibrokeratomas occur most frequently on the fingers and toes.¹¹ However, a history of injury is usually present in only a few cases and in only four of our patients, although other patients might not have noticed minor trauma. In our study too, the middle and ring fingernails as well as the great and fourth toenails were more commonly affected and these are sites likely to be exposed to trauma or infection, supporting the above hypothesis.^{7,8} Trauma may be a trigger due to damage to the nail matrix but further research is needed.

Acquired unguial 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

Table 1: Location of acquired unguial fibrokeratomas

Site	Number of cases (%)
Fingers:	
Thumb	0
Index finger	2 (10)
Middle finger	2 (10)
Ring finger	0
Little finger	0
Toes:	
Great toe	9 (45)
2 nd toe	2 (10)
3 rd toe	1 (5)
4 th toe	3 (15)
5 th toe	1 (5)



Figure 2a: Periungual AUFK



Figure 2c: Intraungual (dissecting unguial) AUFK



Figure 2b: Subungual AUFK

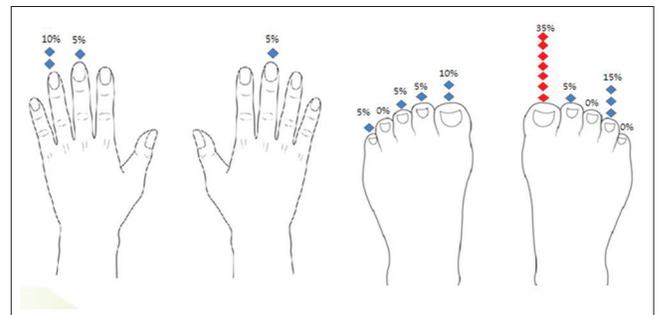


Figure 3: Distribution of acquired unguial fibrokeratoma of nail unit of fingers and toenails

Table 2: Patient characteristics

Patient	Sex/age	Nail involvement	Location	Clinical type	Duration (months)	PHx.	Follow-up (month)	Treatment	Recurrence
1	Male/62	O	Left 4 th finger Intraungual	Flat	3		1	Excision	X
2	Male/41	X	Right 4 th toe Periungual	Dome-shaped	12		2	Excision	X
3	Female/67	O	Left 2 nd toe Subungual	Dome-shaped	36		6	Excision	X
4	Male/47	X	Left 5 th toe Periungual	Branching	12		4	Excision	X
5	Female/55	O	Right great toe Intraungual	Branching	1	Trauma History	8	Excision	O
6	Male/55	X	Right 2 nd toe Periungual	Dome-shaped	48	DM	1	Biopsy	X
7	Male/29	O	Right 3 rd finger Intraungual	Flat	24		1	Excision	X
8	Male/30	O	Right great toe Periungual	Flat	48		1	Excision	X
9	Male/53	X	Left 3 rd toe Periungual	Dome-shaped	180	Trauma History	15	Excision	X
10	Female/40	O	Left 4 th finger Periungual	Branching	12		1	Excision	X
11	Male/61	X	Right 4 th toe Periungual	Dome-shaped	36	Gastric cancer	36	Excision	X
12	Female/16	O	Right great toe Periungual	Dome-shaped	12		24	Biopsy	X
13	Female/45	O	Right great toe Intraungual	Rod-shaped	12	HTN	1	Excision	X
14	Female/29	O	Right great toe Periungual	Rod-shaped	60	Trauma History	12	Excision	O
15	Male/15	O	Right great toe Periungual	Flat	36	Trauma History	5	Excision	X
16	Female/79	X	Right 4 th toe Periungual	Dome-shaped	96		1	Excision	X
17	Male/46	O	Left great toe Periungual	Branching	5	HTN	36	Excision and EC	O
18	Female/38	X	Left 3 rd finger Periungual	Branching	7		1	Excision and EC	X
19	Female/19	O	Right great toe Periungual	Dome-shaped	12		5	Excision and EC	X
20	Female/32	O	Left great toe Periungual	Rod-shaped	24		4	Excision and EC	X

EC: Electrocauterization, HTN: Hypertension, DM: Diabetes mellitus

vulgaris, a supernumerary digit, Koenen's tumor of tuberous sclerosis, pyogenic granuloma, eccrine poroma, cutaneous horn, epidermoid cyst, acrochordons and neurofibromas must be included in the differential diagnosis of acquired unguis fibrokeratoma.^{8,11-13}

The histopathologic appearance of an acquired unguis fibrokeratoma is distinct. Histopathologic 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 [Figure 6].¹ In 1985, Kint *et al.* classified acquired digital fibrokeratomas into three histologic subtypes.¹⁴ 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. When this classification was applied to our patients, the first variant was found to be the most common (70%) followed by the second (25%) and third (5%) variants [Table 3], consistent with the above report by Kint *et al.*

Kint *et al.* divided acquired unguis fibrokeratomas into two types based on appearance, dome-shaped and finger-like.¹⁴ However, we observed more diverse morphologies and classified them into four subtypes: Rod-shaped, dome-shaped, flat and branching. Rod-shaped acquired unguis fibrokeratomas are smooth and trigonal; dome-shaped lesions curving and round; flat ones have a horizontal surface without a slope and branching acquired unguis fibrokeratomas appear to be divided into two or more parts. Kim *et al.* classified acquired unguis fibrokeratoma into four clinical types viz., pine-pit-like, dome-shaped, finger-like and chicken's foot-like.¹⁵

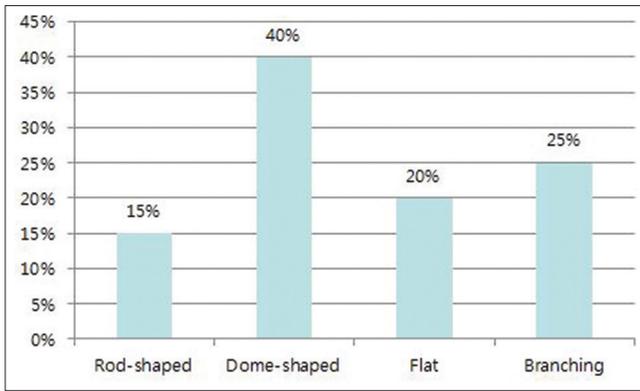


Figure 4a: Clinical manifestation of AUFK

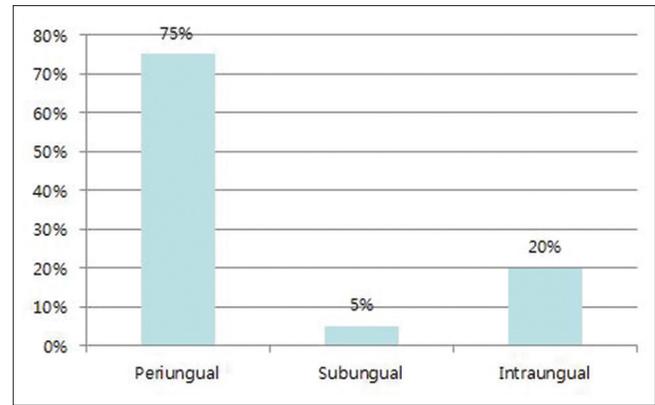


Figure 4b: Distribution of AUFK of nail unit



Figure 5a: Oblique incisions were made at corners of the perionychium



Figure 5b: Oblique incisions were made at corners of the perionychium



Figure 5c: The proximal nail fold was retracted to expose the nail matrix by skin hooks



Figure 5d: The whole tumor was exposed and excised with its basal attachment

Although there are differences in classification, Kim *et al.* found that dome-shaped acquired unguis fibrokeratomas were the most common clinical type, as in our study. In this study accompanying nail abnormalities include longitudinal grooving, trachyonychia, chromonychia, subungual hyperkeratosis, onycholysis and dissected nail plates. Acquired unguis 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.¹⁴

Acquired unguis fibrokeratomas do not spontaneously regress.¹² They should be excised when they cause symptoms or significant cosmetic problems. The preferred treatment is total excision

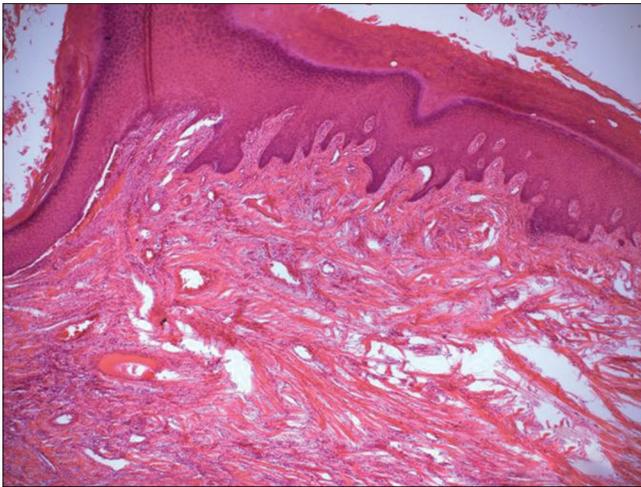


Figure 6a: A microscopic image shows hyperkeratosis, acanthosis, and elongated rete ridge in the epidermis and core of the tumor, which consists of dense, closely packed collagen bundles (H and E, x40)

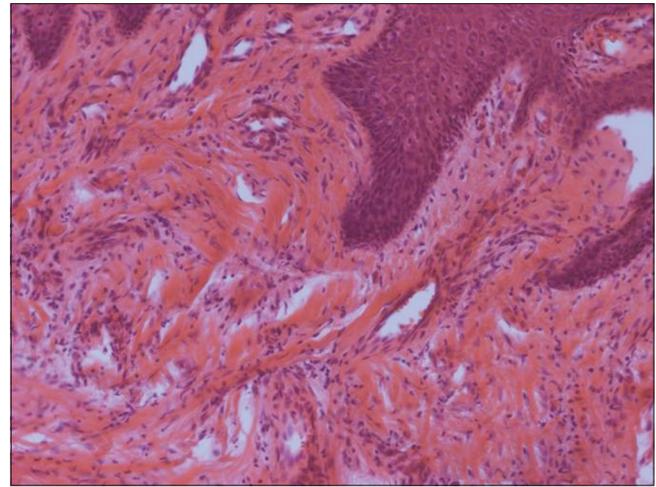


Figure 6b: Irregularly arranged collagen fibers and numerous capillaries and fibroblasts were observed (H and E, x200)

Table 3: Histopathological features of acquired unguis fibrokeratoma

Feature	Number of cases (%)
Epidermal changes:	
Hyperkeratosis	20/20 (100)
Acanthosis	20/20 (100)
Parakeratosis	1/20 (5)
Focal hypergranulosis	11/20 (55)
Irregular rete ridges	10/20 (50)
Core of the tumor:	
Thick, dense and closely packed collagen bundles (Type 1)	14/20 (70)
Increased number of fibroblasts in dermis (Type 2)	5/20 (25)
Edematous and poorly cellular structure (Type 3)	1/20 (5)

because superficial removal usually results in recurrence.¹⁶ 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, carbon-dioxide laser vaporization or surgical resection after lifting the proximal nail fold as a banner flap.¹⁷⁻¹⁹ During the procedure, the germinal matrix should be preserved by careful dissection along the undersurface of the tumor and the tumor should be completely excised with its basal attachment.^{20,21} To minimize the risk of recurrence, we recommend careful elevation of the nail plate and retraction of the proximal nail fold to expose the whole tumor and surrounding nail matrix, avoiding additional injury to the nail matrix.

Our study had some limitations. Apart from being a retrospective study with inevitable biases, we only had a limited number of patients. Further studies are needed to look into the etiology of acquired unguis fibrokeratomas.

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Conflicts of interest

There are no conflicts of interest.

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