Case Report

Acral Fibrokeratoma- A Report of Two Cases

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Abstract

Observation: Acral fibrokeratoma (AF) is an uncommon benign tumor of fibrous tissue and usually presents with skin to flesh colored exophytic papulonodules with a collarette of slightly raised skin at the base. The commonly affected sites are fingers and toes and sometimes, palm or sole. We here report two cases of AF- one classical and the other atypical.

Introduction

Acral fibrokeratoma (AF) is an uncommon benign fibroepithelial tumor of middle age. The core of the lesion is formed by thick collagen [1]. It usually presents with solitary, skin-colored to pink, slightly keratotic exophytic papulonodules on acral parts, most commonly digits [2]. It usually grows up to a size of 1.5 cm and is largely asymptomatic [3]. It needs to be differentiated from other common conditions including periungual fibroma, supernumerary digit, verruca, cutaneous horn and dermatofibroma [2, 3]. We here report two cases of AF- one classical and the other atypical.

Case Report 1

A 29 year old male presented with an asymptomatic growth over right thumb for last 6 months. The lesion started as a small growth following trauma and had been growing in size since then. Rest of the history was unremarkable. On examination a firm pedunculated lesion of 1 cm size was found on right thumb (**Figure 1**).

The lesion was of skin color, firm, non tender and was having collarette of raised skin at the base.

There was mild hyperkeratosis over top most part of lesion. Rest of the cutaneous examination was unremarkable. Clinical diagnosis of AF was made and excision biopsy was performed. Histopathological findings were consistent with diagnosis.

Case Report 2

A 36 year old female presented with two firm growths on right great toe and adjacent web space for 2 years and one firm growth over right sole for last 6 months (**Figure 2**).

The lesions started first in first web space and had been growing in size since then. Soon after the development of 1st lesion she noticed small growth on right great toe, destroying nail. All the lesions were asymptomatic and hence she did not bother much. Some 6 months back she noticed flat firm growth over right sole. Initially it was smaller in size but grew in size slowly. On further enquiry she revealed she had similar firm lesion in 1st web space some 5 years back. She had consulted a surgeon for this and was operated on. She could not tell the diagnosis made then and could not produce any relevant document. Rest of the history was unremarkable. On examination three lesions were found. 1st lesion was found in right periungual area. It was well demarcated but irre-



Figure 1. Firm pedunculated growth on right thumb. Note collarette of skin at the base.

gular in shape (maximum diameter being 3 cm), slightly pink with collarette of skin at base and was destroying almost lateral half of great toe nail. Second lesion was seen in the first web space and was totally separate from the periungual lesion. Its appearance was similar to 1st lesion but was much bigger in size, extending over proximal and lateral part of great toe and plantar surface (**Figure 3**).

Third lesion was found on center of right sole (**Figure 4**). It was flat, firm, skin colored and irregular in shape. All the lesions were well demarcated, firm, non tender and were having no surface changes. Rest of the cutaneous examination was unre-



Figure 3. Lesion on web space extending to plantar surface



Figure 2. Firm irregular shaped growths on periungual area and web space

markable. Differential diagnoses included AF, fibroma, dermatofibroma and superficial acral fibromyxoma. Punch biopsy was performed from the periungual lesion. Histopathological examination showed acanthosis with thickened rete ridges. There was sparse mononuclear cell infiltrate in the dermis (**Figure 5**). Verhoeff- Van Gieson staining was done and it showed thick collegen bundles, arranged vertically (**Figure 6**). Histopathological examination from the web space lesion showed similar changes (**Figure 7**). Based on history, clinical findings and histopathological findings, diagnosis of AF was made and patient was referred to Department of General surgery for excision.



Figure 4. Firm irregular flat growth over sole

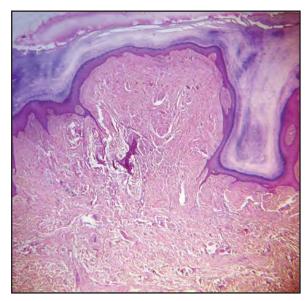


Figure 5. Histopathological photograph showing acanthosis with thickened rete ridges. Sparse mononuclear cell infiltrate can be appreciated. Note hyperkeratosis (H&E stain X 100).

Discussion

Acquired digital fibrokeratoma (ADF) was described first by Bart et al in 1968. Later on Pinkus reported 28 more cases and suggested the name "Acral fibrokeratoma (AF)" [3] as lesions were not restricted to digits only. It is an uncommon benign tumor characterized clinically by skin or flesh colored pedunculated or dome shaped firm growth on acral parts, often resembling rudimentary supernumerary digit and histologically by thick col-



Figure 7. Histopathological photograph showing thickened collagen. Vertical arrangement can be well appreciated. (VVG stain X100)

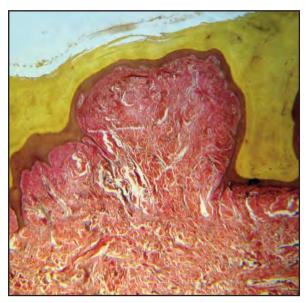


Figure 6. Histopathological photograph showing thickened collagen arranged vertically (VVG stain X100).

lagen bundles arranged vertically. Trauma has been postulated as predisposing factor but pathogenesis is largely unknown [1, 2]. One case reports co-occurrence of AF and pyogenic granuloma [4]. It further supports the role of trauma in pathogenesis.

AF has been reported in all races and no sex predominance has been established so far. It is most commonly seen in middle aged patients (range 12-70 years) [3]. Infantile digital fibromatosis too is known [1]. But it usually presents at birth or during infancy. Single or multiple nodules are seen on digits, excluding thumb or great toe. It grows up to a size of 2 cm and involutes spontaneously. However recurrences may be seen in childhood. Clinically it resembles AF but histopathology is quite different [1].

AF usually presents as solitary, skin-colored to pink, slightly keratotic exophytic papulonodule with a collarette of elevated skin [2]. It is most commonly found on digits and less frequently on palm and sole. It can be seen in periungual area too where it may be confused with Koenen tumor [3, 5]. It usually grows up to 1.5 cm but lesion as large as 11 x 70 x 50 mm has been described by Spitalny and Lavery [6]. AF is largely asymptomatic but giant lesions can cause discomfort [3].

Common differential diagnoses include rudimentary supernumerary digit, periungual fib-

roma, verruca, eccrine poroma, pyogenic granuloma or fibroma, cutaneous horn and dermatofibroma. Supernumerary digits are often bilateral, present on 5th fingers and are present since birth. Histopathological examination can differentiate AF from other conditions. It shows hyperkeratotic and acanthotic epidermis with thickened, often branching, rete ridges. The core of the lesion is formed by thick, interwoven bundles of collagen, predominantly vertically oriented. Elastic fibers are usually present but are sparse [1, 2, 3]. Treatment is done by surgical excision. Recurrence is rare but known [3].

Our case 1 is a classical AF resembling rudimentary supernumerary digit. But case 2 is atypical. Firstly, there was more than one lesion. AF is known to be solitary only. However one case of multiple acral fibrokeratoma had been reported long back [7]. Almost all the lesions were quite larger than 1.5 cm.

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