

Acquired Digital Fibrokeratoma

Clinical and Histopathological Study

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Introduction:

Acquired digital fibrokeratoma was described 1986⁽¹⁾ as a benign acquired projection occurring on the digits. The lesion is skin colored, asymptomatic, elongated or pedunculated with a flat top or hyperkeratotic Verruciform surface⁽²⁾. The projection arises out of a collarette of slightly raised skin⁽³⁾.

Most of the reported cases were on the fingers and were relatively small and did not involve the nail⁽⁴⁾. Fibrokeratoma of the nail bed was described as a red, hard hook shaped hyperkeratotic tumor arising from nail bed of the third toe of right foot⁽⁴⁾. Two cases of subungual fibrokeratoma were described in the literature^(5,6). The subungual fibrokeratomas are considered of rare origin and rarely could reach a big size that may measure 20 mm x 25 mm x 10 mm while the known average size of a digital fibrokeratoma is 3 to 5 mm in all three dimensions⁽⁴⁾.

A clinical variant of acquired digital fibrokeratoma a giant one that was reported to affect the heel and reached a size of 25mm x 23mm x 12mm⁽⁷⁾. Acquired digital fibrokeratoma are almost always solitary and are seen in adults. Sites other than the fingers reported to be affected are proximal hand, sole⁽⁷⁾, prepatellar region and that is why it has been suggested by Verallo et al⁽⁸⁾ that the entity might be called acral fibrokeratoma. Fibrokeratoma of the heel and foot may reach a big size. A 3cm sessile fibrokeratoma of the heel was described⁽⁹⁾. Big fibrokeratoma of the foot that measured 40 mm x 30 mm x 9 mm was reported⁽¹⁰⁾. Another big fibrokeratoma of heel with the size of 11mm x 7mm x 50 mm was described⁽¹¹⁾.

Recently a giant acquired digital fibrokeratoma was described⁽¹²⁾ as a solitary red pedunculated nodule measuring 32 x 38 mm in diameter and 15 mm in thickness on the nail fold of the right big toe in a 33 years old female. The nodule was hard with scaly papillomatous surface, which is possibly due to repeated minor trauma to the affected toe during exercise⁽¹²⁾.

Fibrokeratoma usually show a smooth hyperkeratotic skin colored surface⁽¹³⁾. The acquired digital

fibrokeratoma could be confused clinically with other lesions including rudimentary supernumerary digits, fibromas, neuromas, neurofibromas⁽¹⁴⁾, verruca vulgaris, Koenen's tumor, achrochordon, cutaneous horn⁽¹⁵⁾ and granuloma pyogenicum⁽¹⁶⁾.

Acquired periungual fibrokeratoma is rare and was previously described as garlic-clove fibroma^(17,1). All the lesions with which acquired digital fibrokeratoma are confused may be differentiated clinically. An important differentiating clinical characteristic of acquired digital fibrokeratoma is the presence of a collarette of slightly raised skin encircling the lesion^(13,18). The supernumerary digits are usually located on the proximal portion of the fifth digit and are usually present since birth and are usually sessile and do not have the collarette described with digital fibrokeratoma. Koenen's tumors are clinically similar to isolated fibrokeratoma and patients with Koenen's tumor show other diagnostic stigmata of tuberous sclerosis as angiofibromas of the face, shagreen patches and ash leaf hypopigmented macules.

High magnetic resonance imaging is a new tool to investigate masses related to the nail apparatus and can exactly locate the site and size of nail tumor mass, which are usually excised for histopathological diagnosis⁽¹⁹⁾.

Solitary periungual lesion has to be differentiated clinically from angiofibroma, seborrhoeic keratosis, fibroma and digital fibrokeratoma⁽²⁰⁾. The neurofibromas are skin colored and are usually multiple, pedunculated and associated with other clinical characteristics of neurofibromatosis. Cutaneous horns are more hyperkeratotic.

Neuromas are usually seen after excision of supernumerary digit. The histopathology of acquired digital fibrokeratoma is the most helpful in making a correct diagnosis. A histological examination of 50 cases of acquired digital fibrokeratoma discloses three histological variants of these lesions⁽¹³⁾.

1. A tumor composed of thick dense and closely packed collagen bundles.
2. A variant with an increased number of fibroblasts in the cutis.
3. A type with an edematous and poorly cellular structure.

The acquired digital fibrokeratoma is considered to be resulting from a new formation of collagen by fibroblasts. The acanthosis of the epidermis seen in probably secondary to the dermal alteration⁽¹³⁾.

Histopathologic findings in acquired digital

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fibrokeratoma includes acanthotic hyperkeratotic epidermis which envelope a core of connective tissue⁽²¹⁾. Histologically it is also described as a cylinder of connective tissue proliferation surrounded by acanthotic papillomatous and hyperkeratotic epidermis⁽¹⁶⁾.

Two new histological variants of acquired fibrokeratoma of the nail apparatus are described and showed three histologic characteristics⁽²²⁾.

- a) A deep epithelial invagination, which develops from the proximal nail fold and is oriented in the same direction as the nail matrix but more proximal to it.
- b) The postero inferior aspect of the Cul-de-sac produced by invagination acts as an accessory matrix-giving rise to a pseudo nail plate. The granular layer is reduced or absent.
- c) This accessory nail apparatus lies on a conical dermal tumor sharply demarcated from the surrounding dermis with a wide base narrowing toward the tip⁽²²⁾.

15-periungual fibromas were studied histologically⁽²³⁾. The lesions were elongated and capillaries were found in the distal part surrounded by thin collagen whereas the proximal part was made up of dense closely packed fibers.

The epidermis covering the tumor appeared to be connected with the nail fold. The dense collagen of the lesion faded into the normal structure of the cutis of the proximal nail fold⁽²³⁾.

Periungual fibromas can be subdivided into:

- a) Fibrokeratoma originating from the dermal connective tissue and
- b) Fibrokeratomas originating from the proximal nail fold⁽²³⁾.

Patient and method

Four cases were studied and diagnosed clinically as acquired digital fibrokeratoma in which only three of them proved histopathologically to correlate with the clinical diagnosis and the remaining one was an accessory digit. We searched the database of our histopathology laboratory among the previous ten years and these three cases were the only available ones of acquired digital fibrokeratoma. History, clinical picture and histopathology of the cases were as follows:

Case One:

A 48-year-old Egyptian male who is a known case of hypertension and hyper-cholesterolemia on treatment presented with a warty asymptomatic lesion of the right little finger of one-year duration. Clinically there was an epidermal collarette of scale around the base of the lesion (Fig.1).

Diagnosed as wart, cutaneous horn or acquired digital fibrokeratoma. Surgical excision of the lesion done and diagnosed histopathologically as acquired digital fibrokeratoma (Fig. 2, 3, 4, 5, 6, 7).



Fig. 1 Acquired digital Fibrokeratoma.



Fig.2 : Low power (4x) of the tip of filiform lesion showing hyperkeratosis, acanthosis (elongated rete ridges) & central fibrovascular stalk showing collagen bundles lying perpendicular to the apex with increased vasculature. (H & E stain; original manifestation x 24).

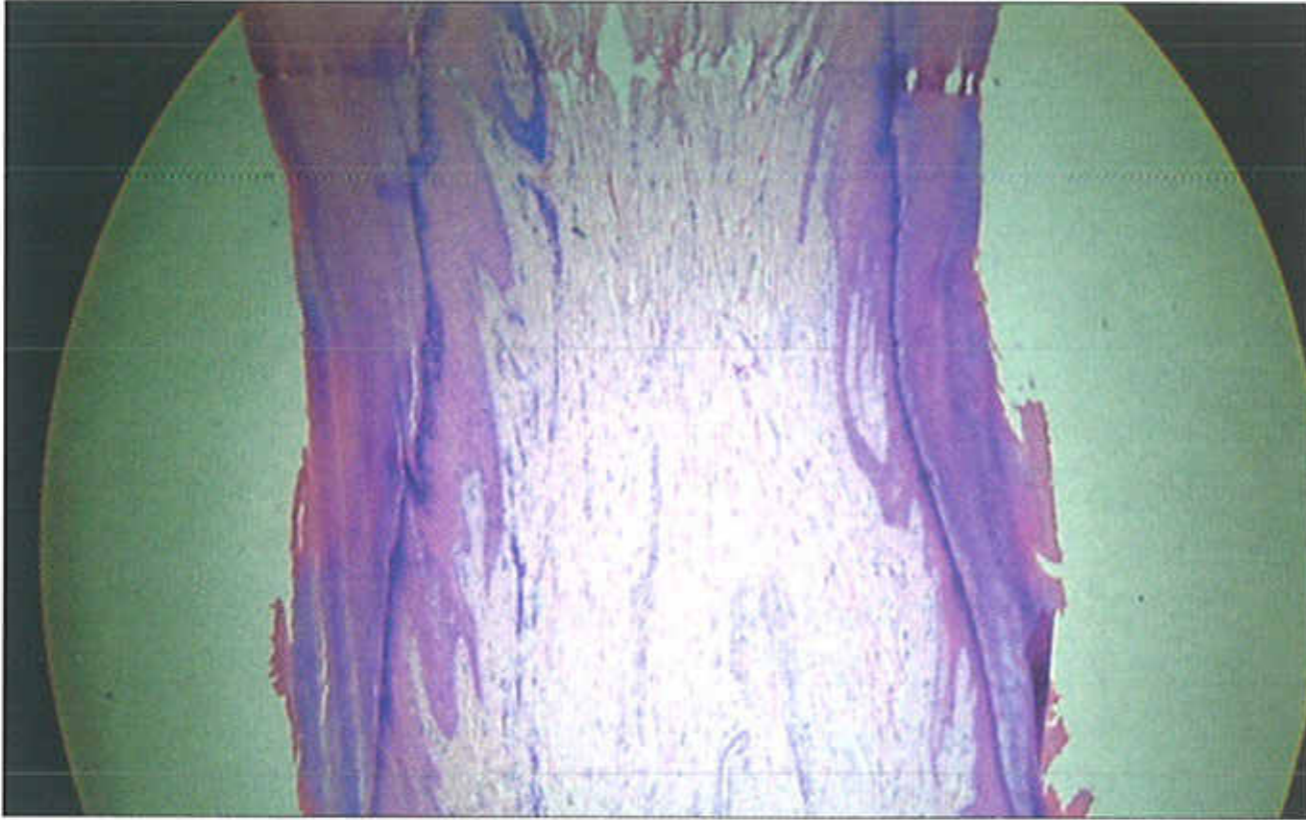


Fig.3 : Low power (4x) of the base of the same lesion as in one showing epidermis in both sides with hyperkeratosis & acanthosis together with a central fibrovascular stalk showing collagen bundles oriented perpendicular to the apex with increase vasculature.

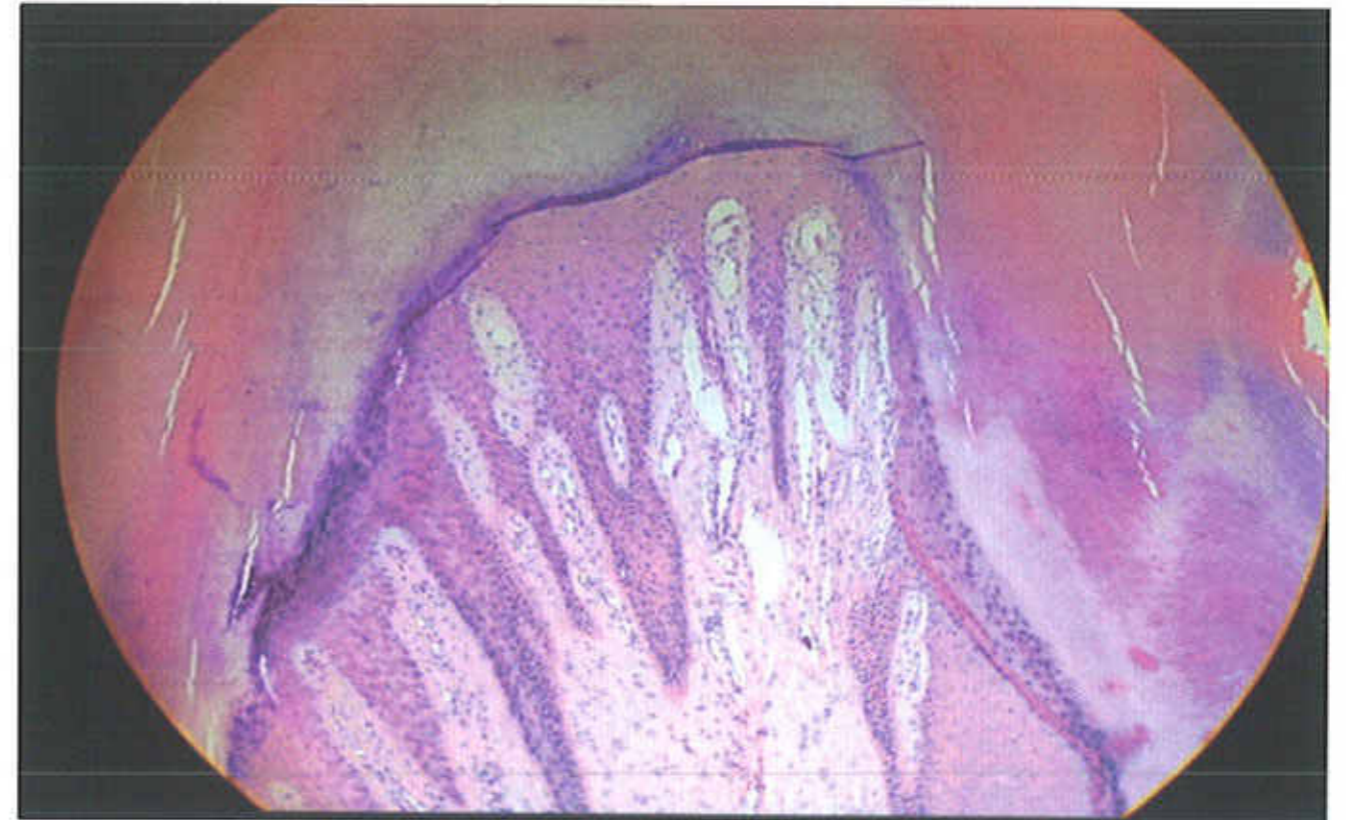


Fig.4 : Medium power view (10x) of the apex of the lesion showing surface hyperkeratosis, acanthosis and central fibrovascular stalk. (H & E stain; original manifestation x 60).

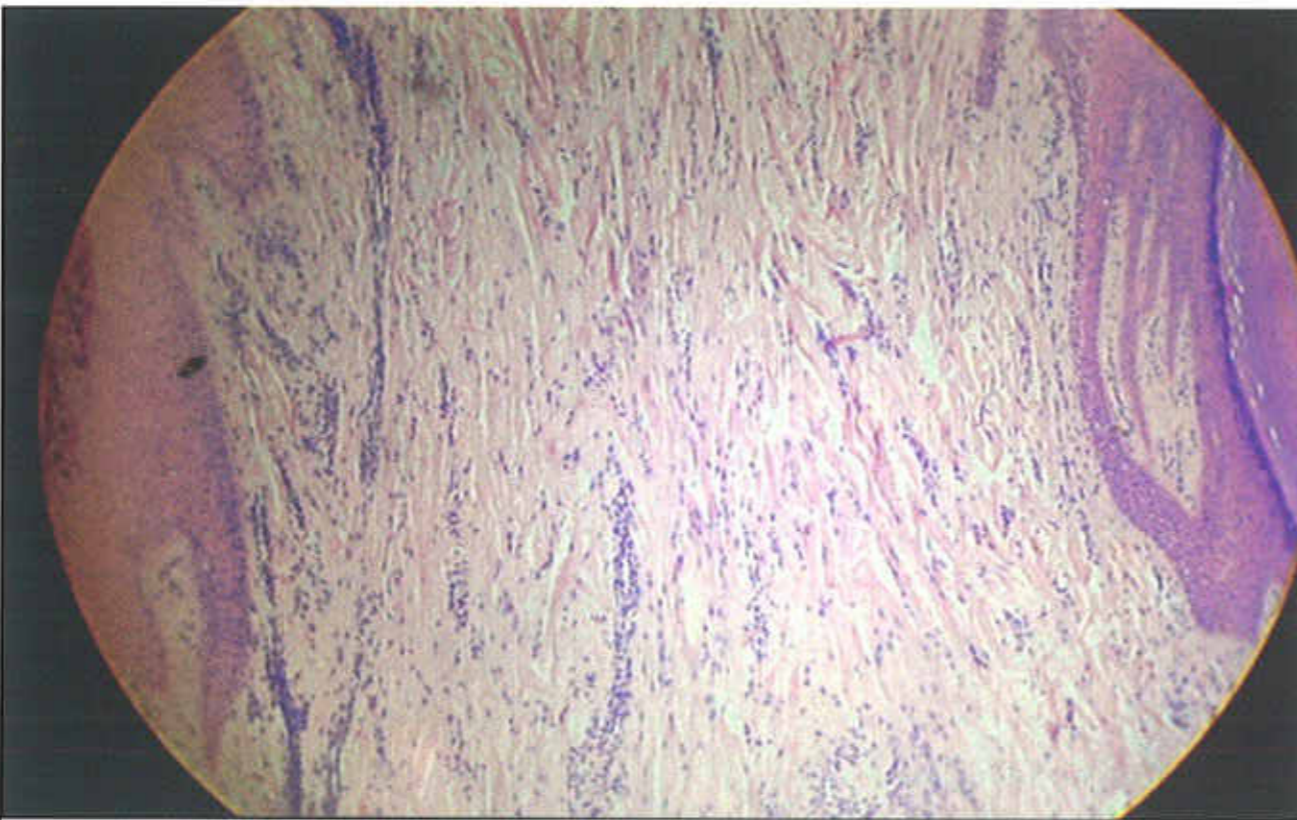


Fig.5 : Medium power view (10x) of the central stalk showing increase vasculature with parallel collagen bundles oriented perpendicular to the apex. (H & E stain; original manifestation x 60).

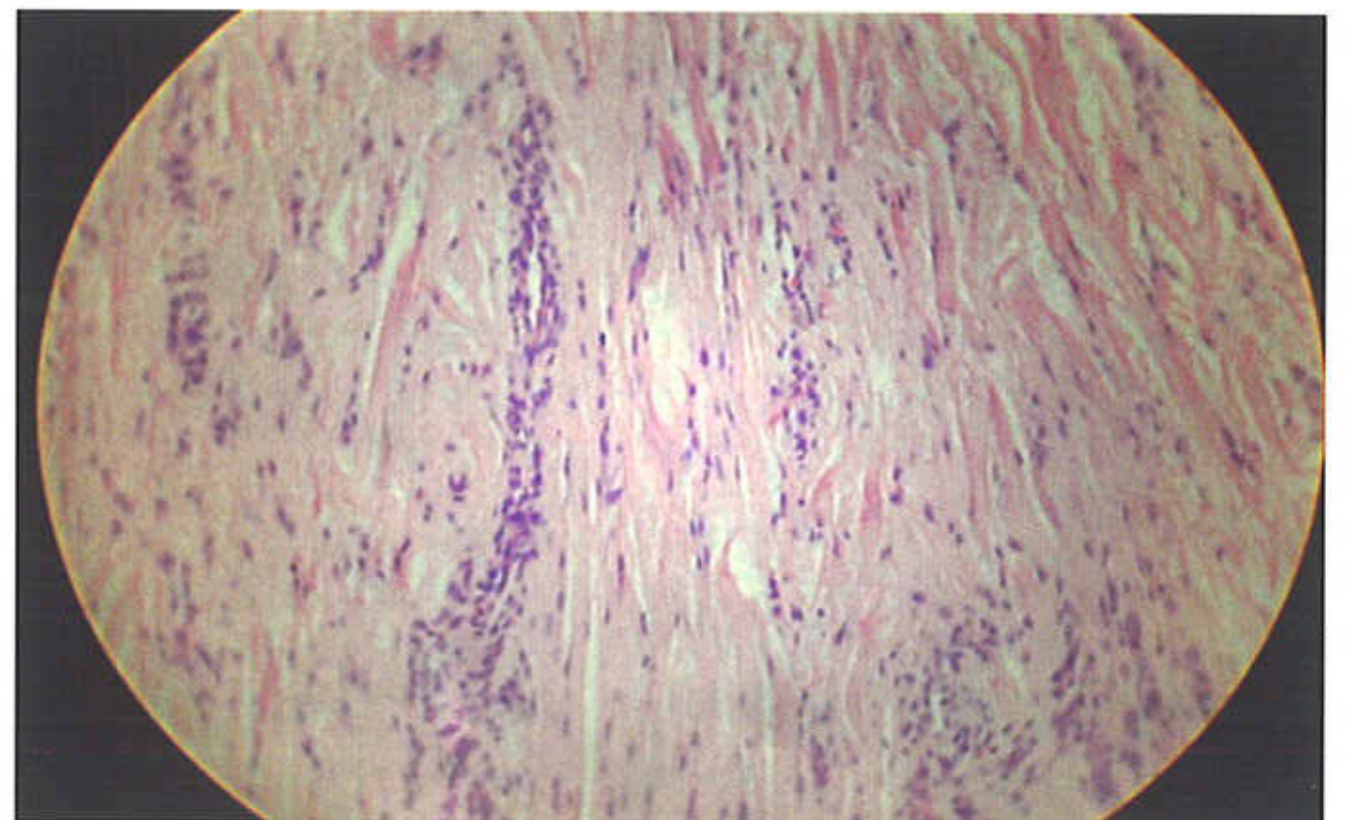


Fig.6 : Higher power view (20x) of collagen bundles running parallel to each other and oriented perpendicular to the apex of the lesion with multiple scattered blood vasculature. (H & E stain; original manifestation x 120).

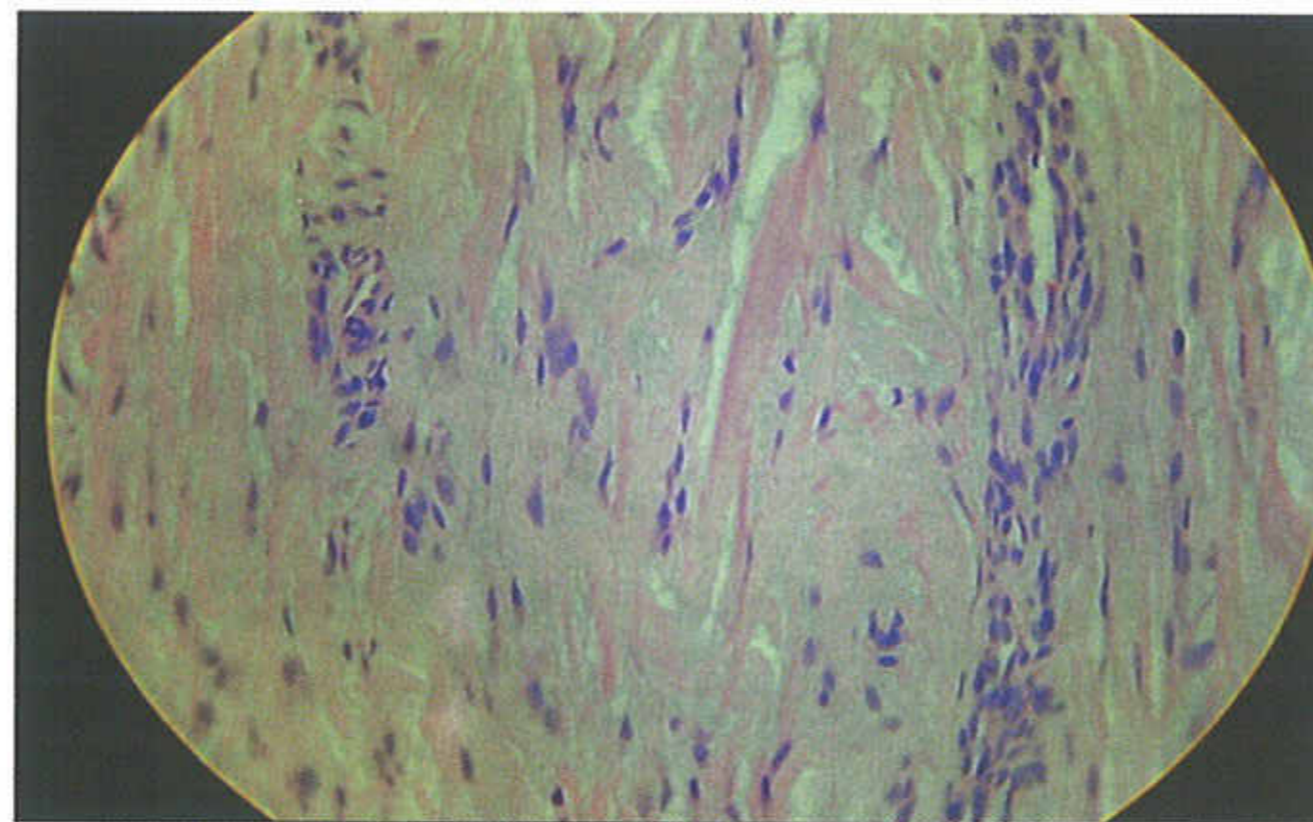
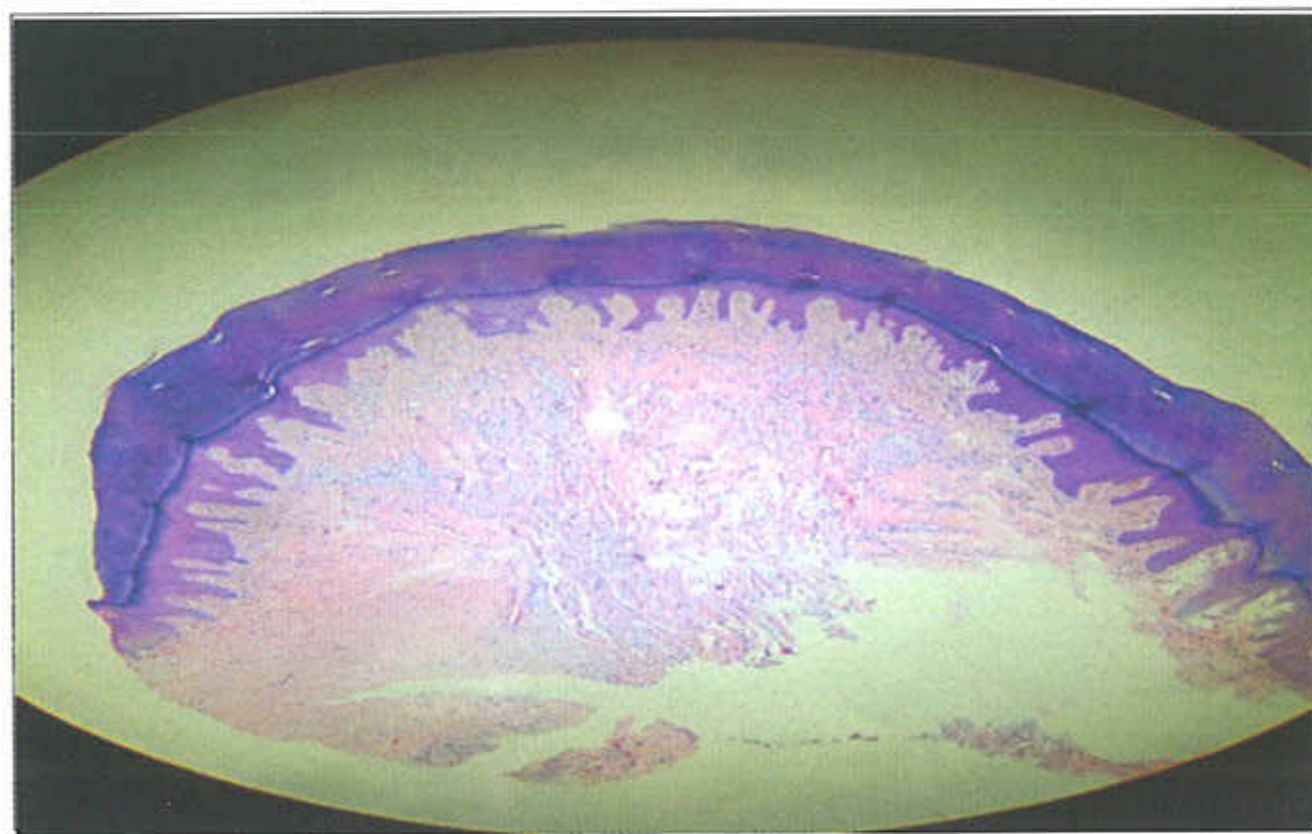


Fig.7 : High power view (40x) showing the same finding of the previous slide. (H & E stain; original manifestation x 240).

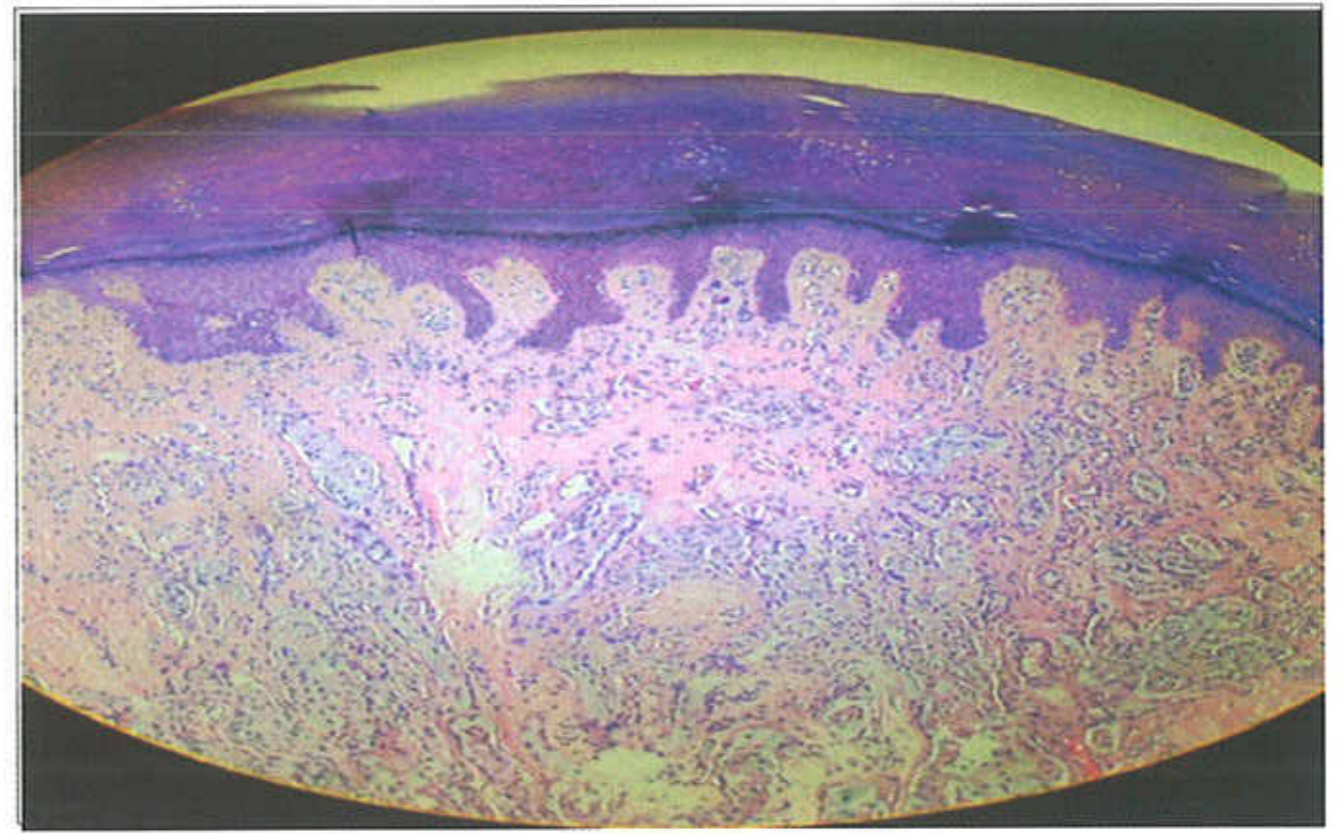
Case Two:

A 13-year old Qatari boy who is a known case of atopic dermatitis, bronchial asthma on treatment presented with dome shaped papule on the lateral aspect of the little finger. The lesion was of years duration and asymptomatic, clinically diagnosed as a wart or acquired

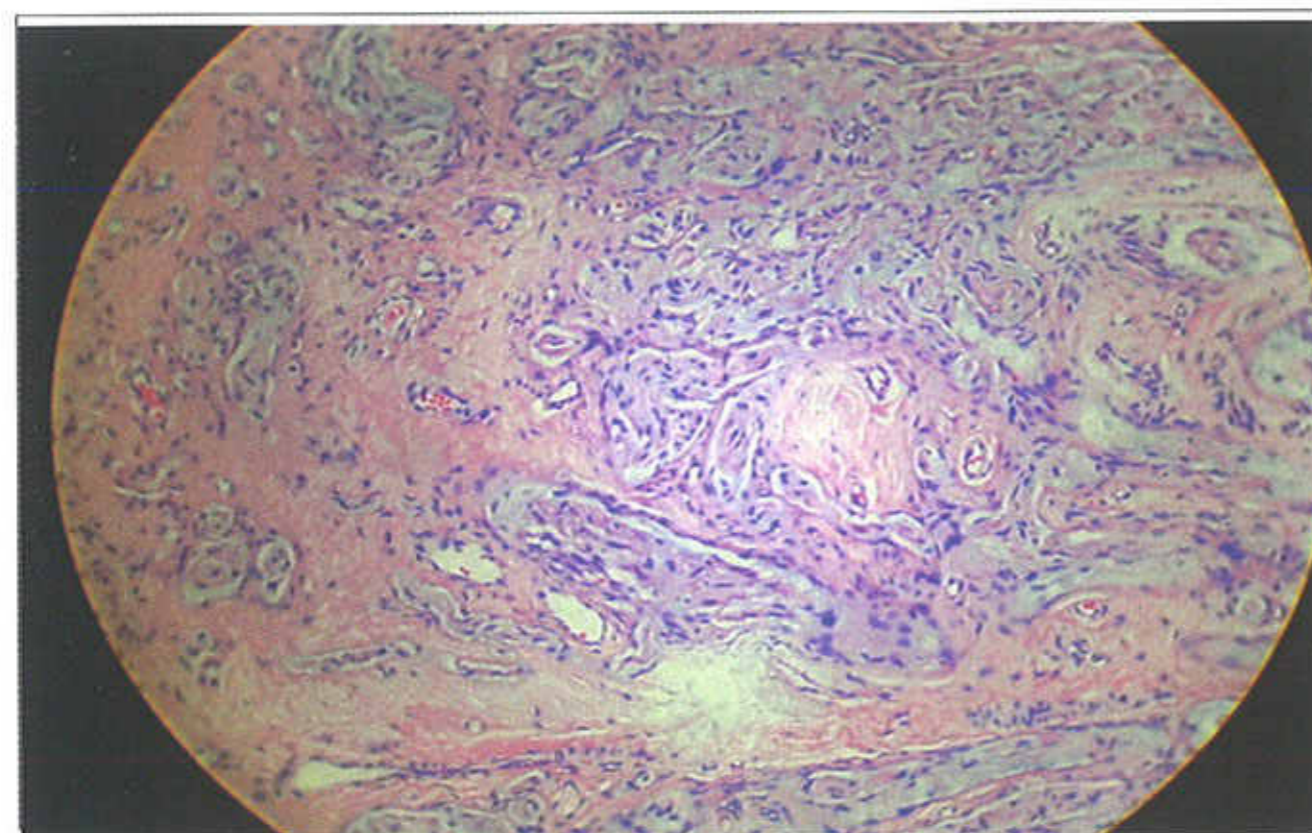
digital fibrokeratoma. The lesion excised surgically under local anesthesia and sent for histopathological study. It was diagnosed as a case of accessory digit due to the presence of many nerve bundles in the fibrovascular stroma as shown in (Fig. 8, 9, 10, 11).



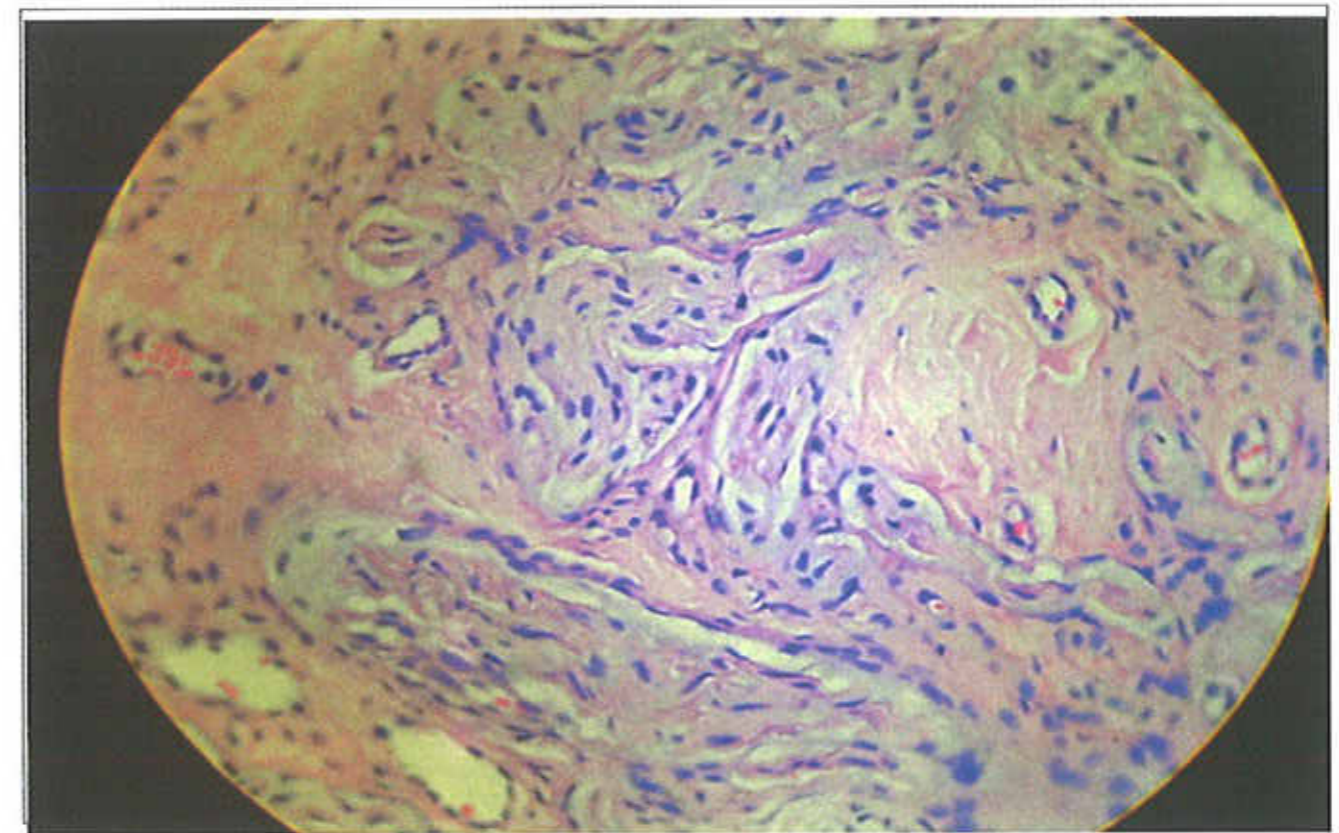
*Fig. 8 : Low power view (4x) of a polypoid lesion covered by Hyperkeratotic stratum corneum, acanthotic stratum spinosum with a central fibrovascular stroma containing many nerve bundles.
(H & E stain; original manifestation x 24).*



*Fig. 9 : Medium power view (40x) of the superficial part of the lesion showing hyperkeratosis, acanthosis and many nerve bundles in the underlying fibrovascular stalk.
(H & E stain; original manifestation x 60).*



*Fig. 10 : Higher power view (20x) of the fibrovascular stalk showing formation of haphazardly proliferation of nerve bundles.
(H & E stain; original manifestation x 120).*



*Fig. 11 : High power view (40x) of nerve bundles seen in the stalk of the lesion.
(H & E stain; original manifestation x 60).*

Case Three:

A 21-year-old Tunisian male patient who is not known to have any chronic illnesses presented to the dermatology clinic with a one and a half year history of a growth on the index finger of the right hand. There was no history of trauma, it was not painful, nor did it itch or bleed.

The patient came to the clinic to have the lesion removed since it was becoming a cosmetic nuisance. Physical examination demonstrated a dome shaped papule with a hyperkeratotic surface on the lateral aspect of the right index finger (Fig. 12, 13).

Histopathological findings were compatible with acquired digital fibrokeratoma. (Fig. 14, 15, 16, 17, 18, 19).

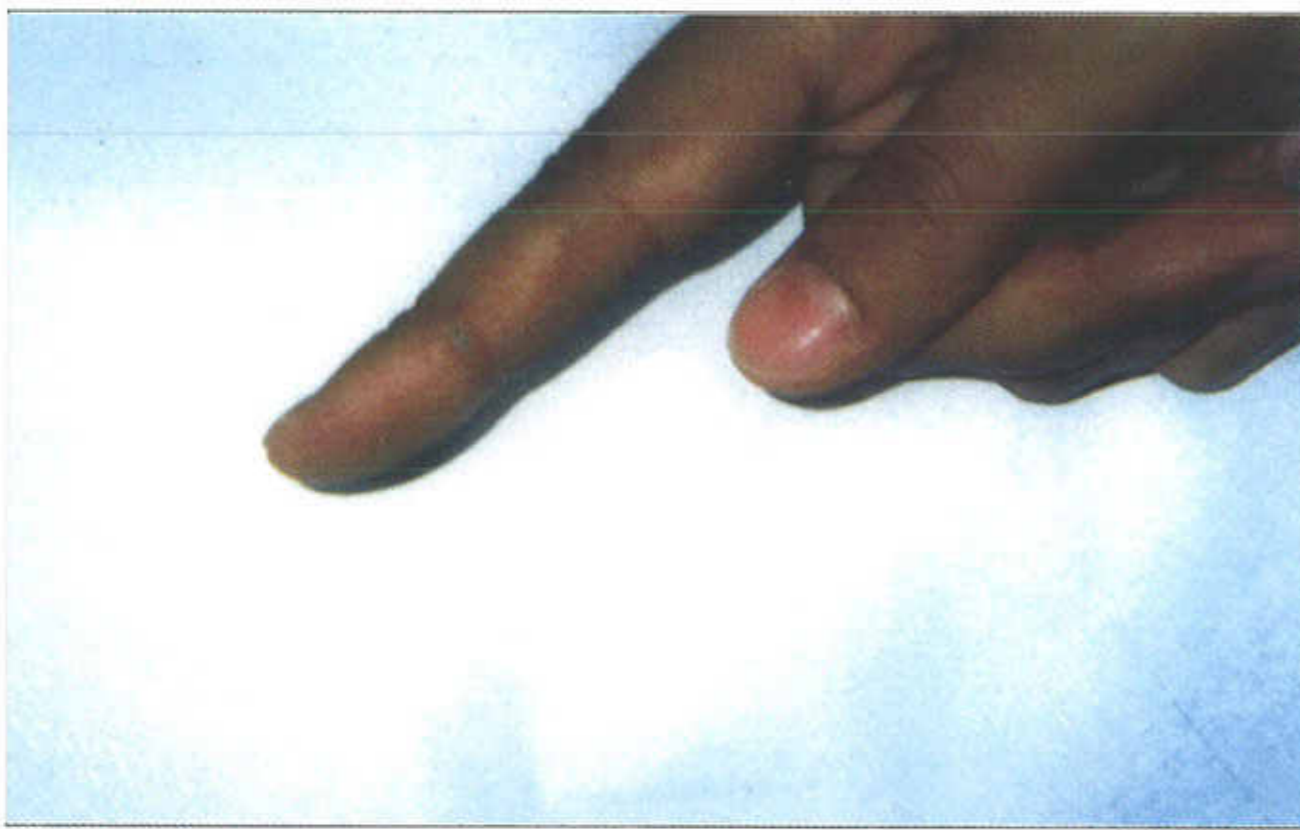


Fig. 12 Dome shaped papule

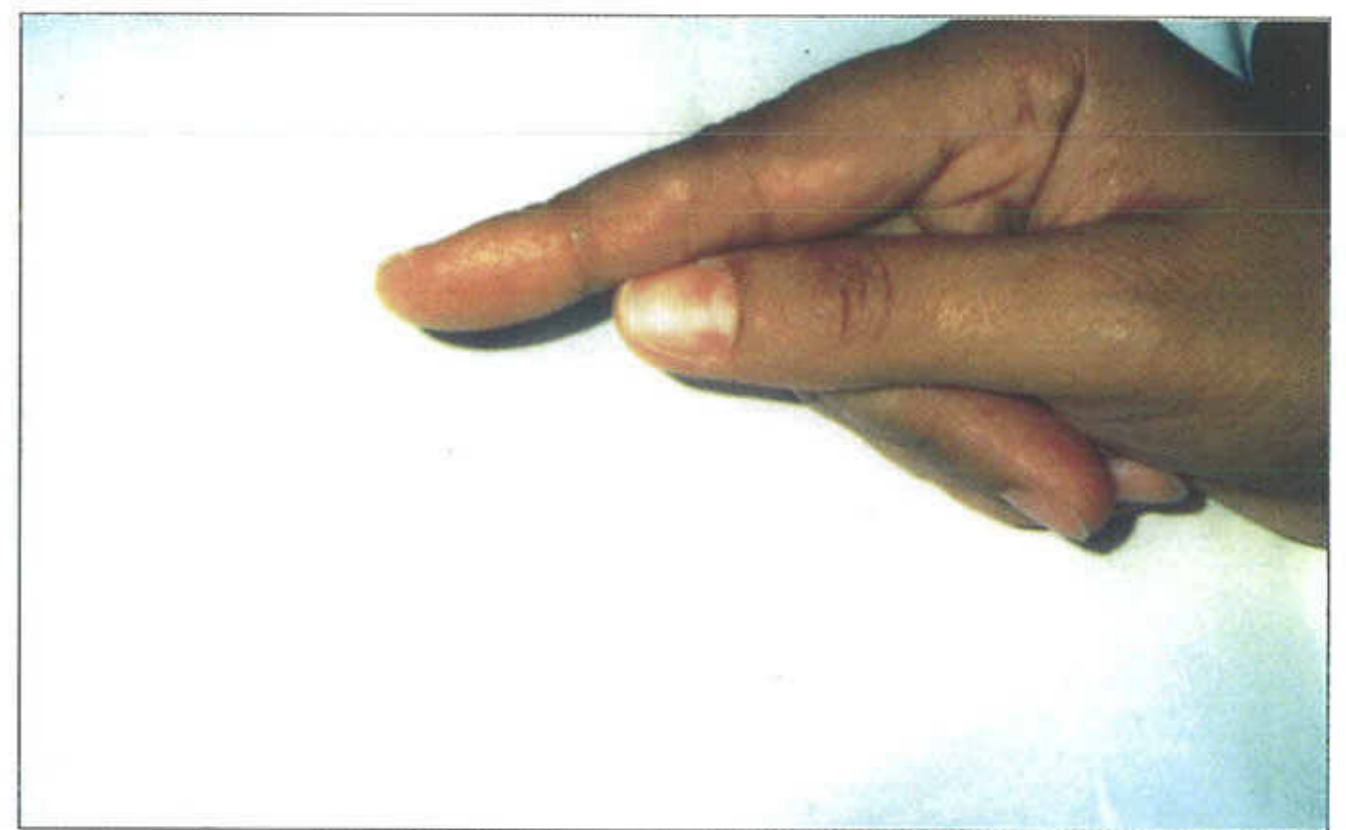


Fig. 13 Dome shaped papule

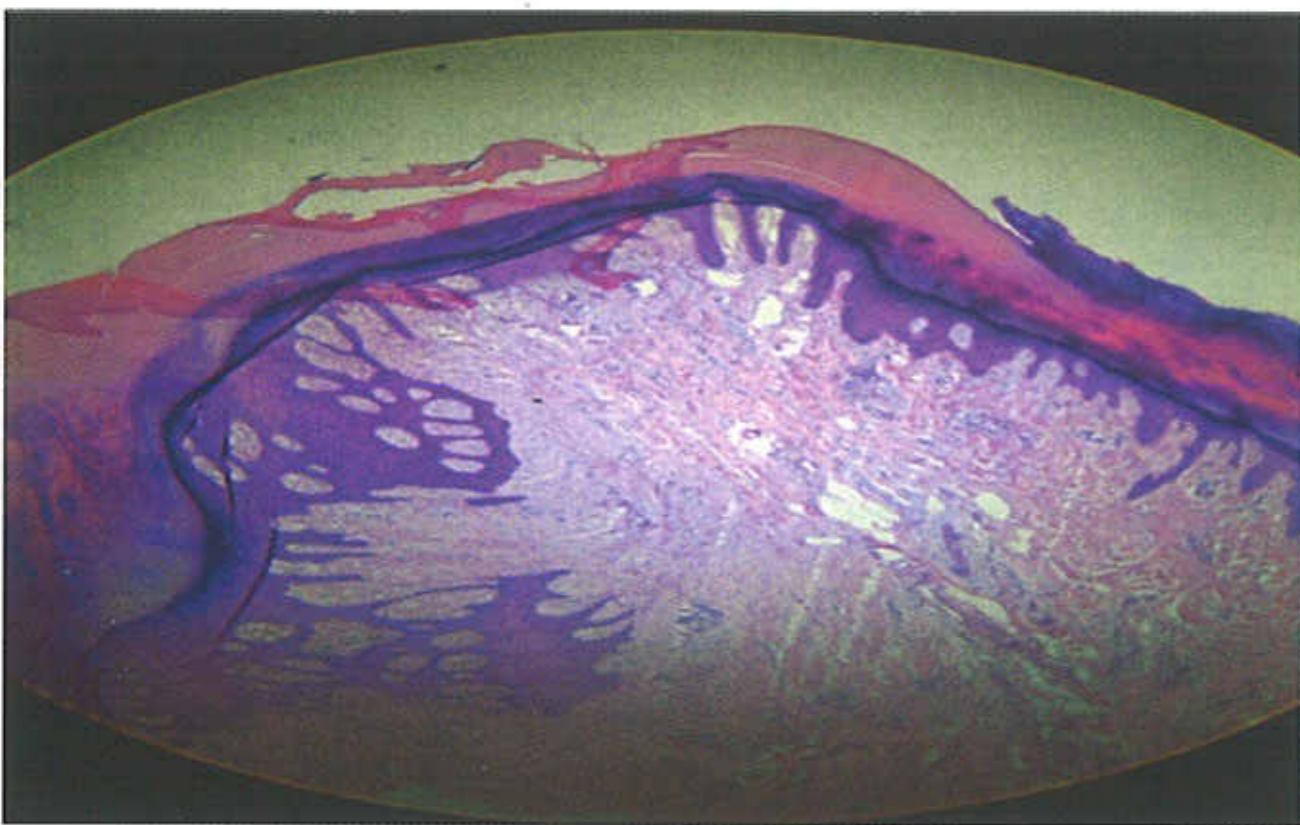


Fig. 14: A low power view 4x (panoramic view) of a dome shaped lesion with surface hyperkeratosis, acanthosis (elongated branching and anastomosing rete-ridges) with an underlying fibro-vascular stalk showing helter-sceltered collagen bundles, frequent scattered adipocytes & variable size blood vessels.
(H & E stain; original manifestation x 24).

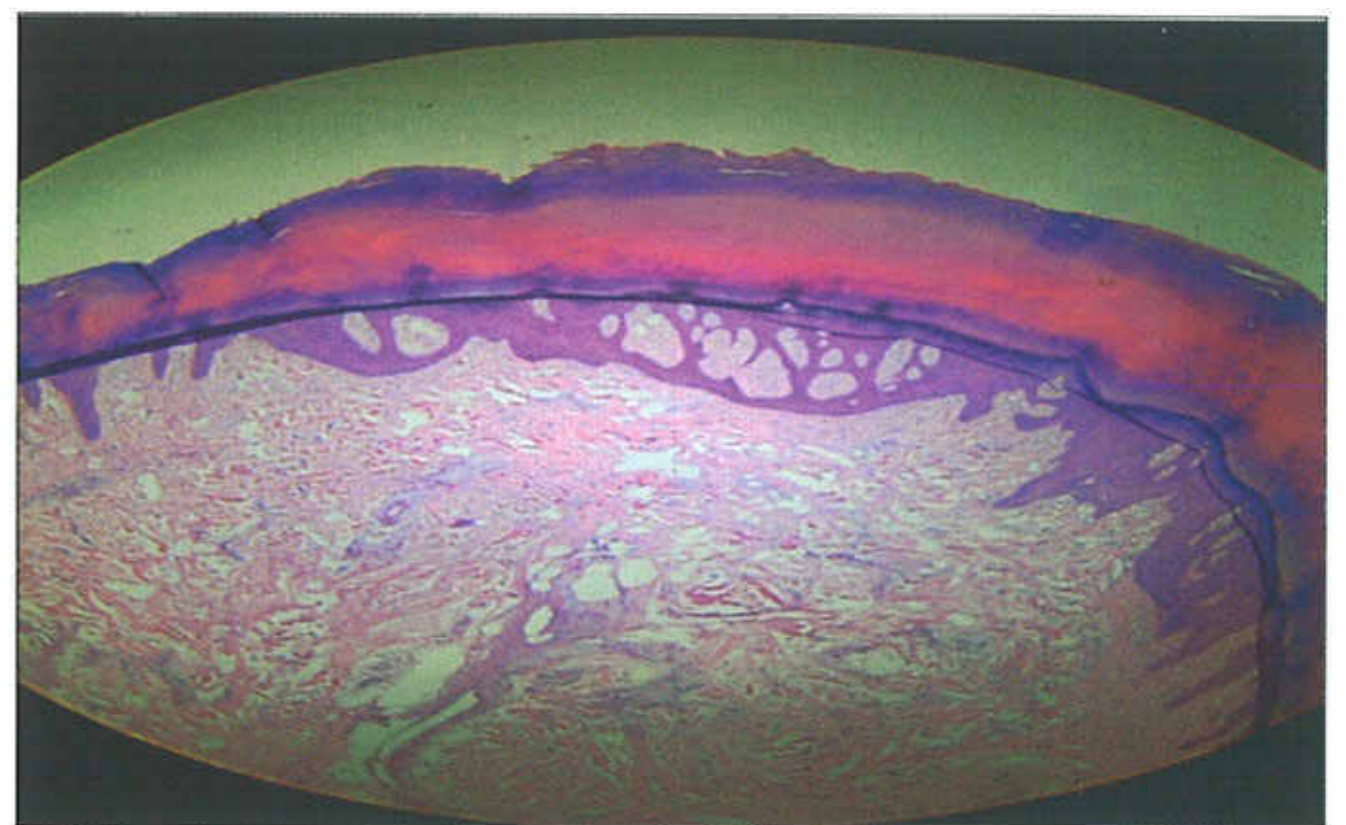
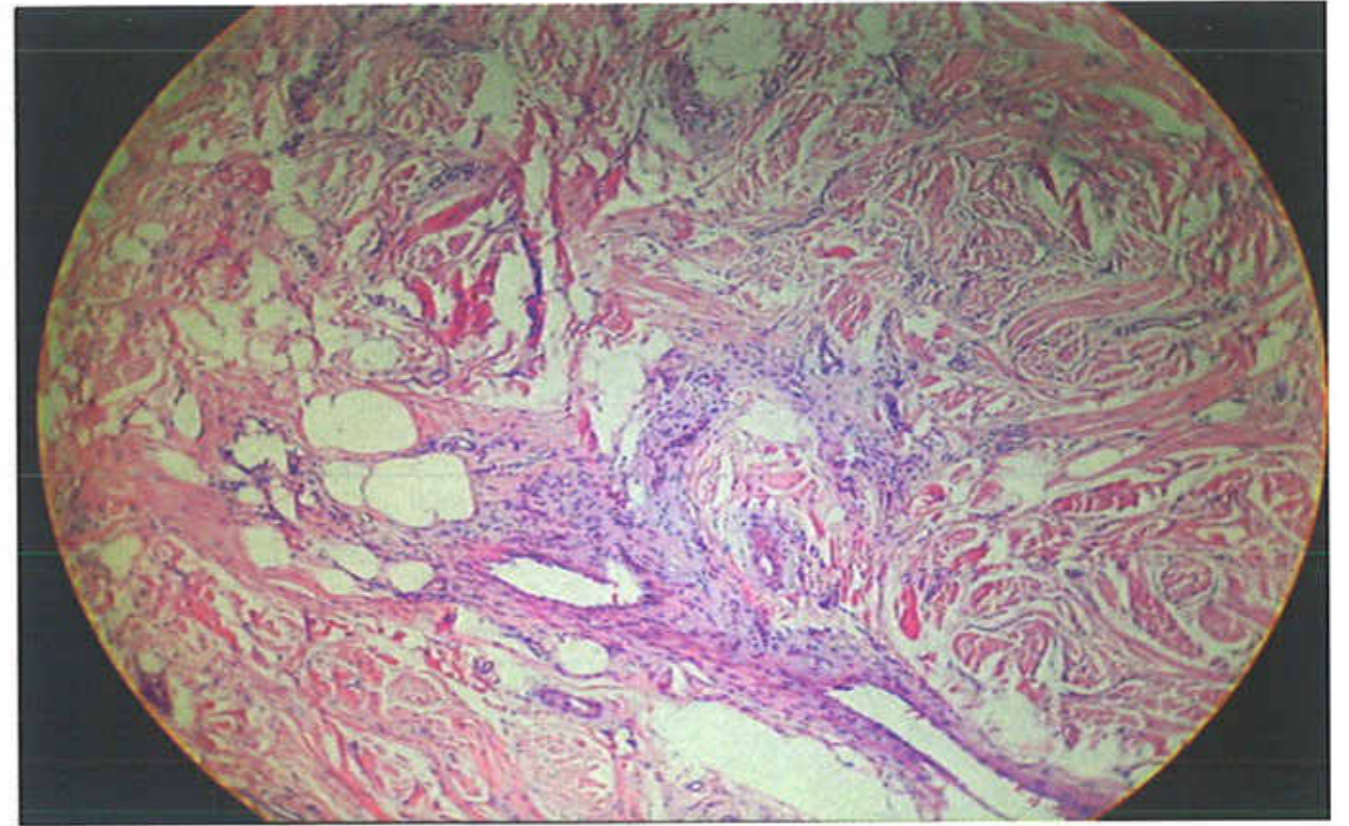


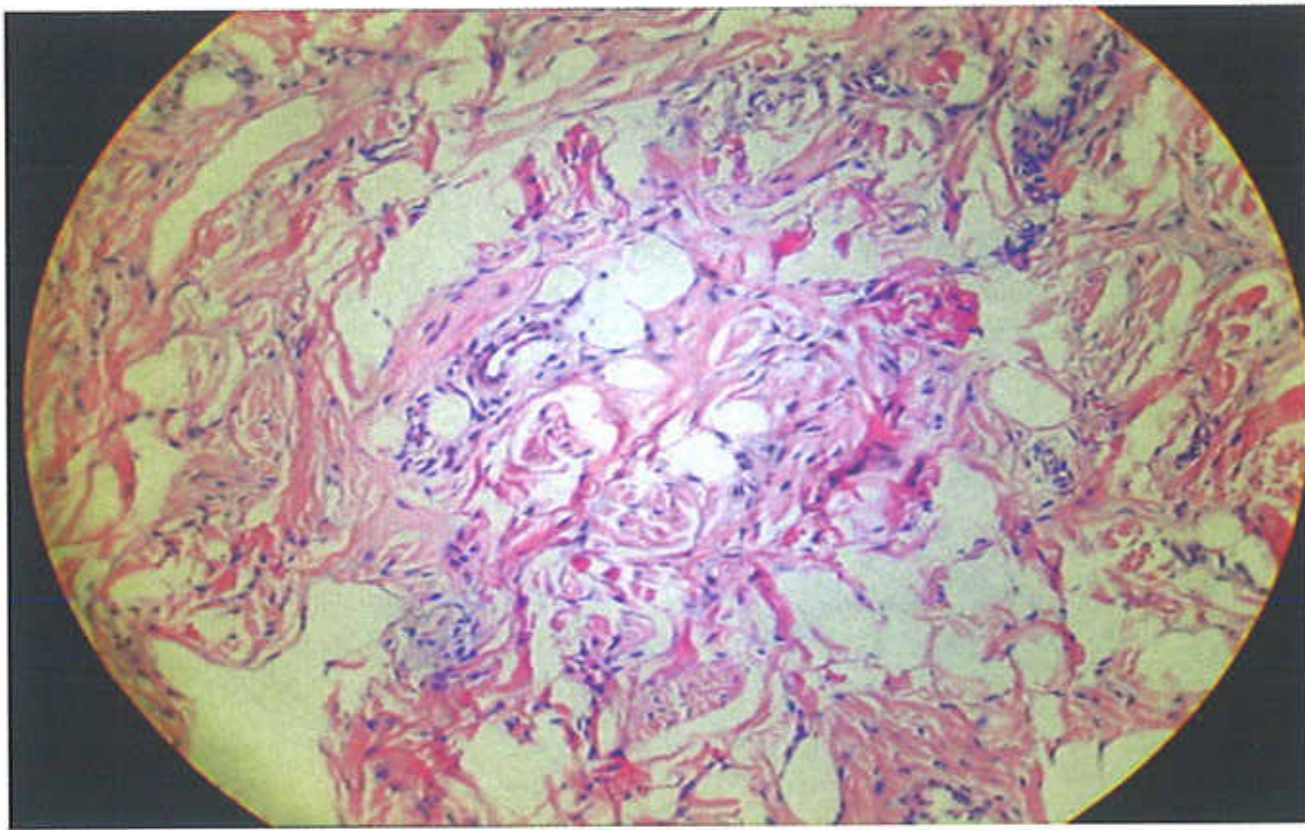
Fig. 15 : Low power view showing the same findings of the previous slide.
(H & E stain; original manifestation x 24).



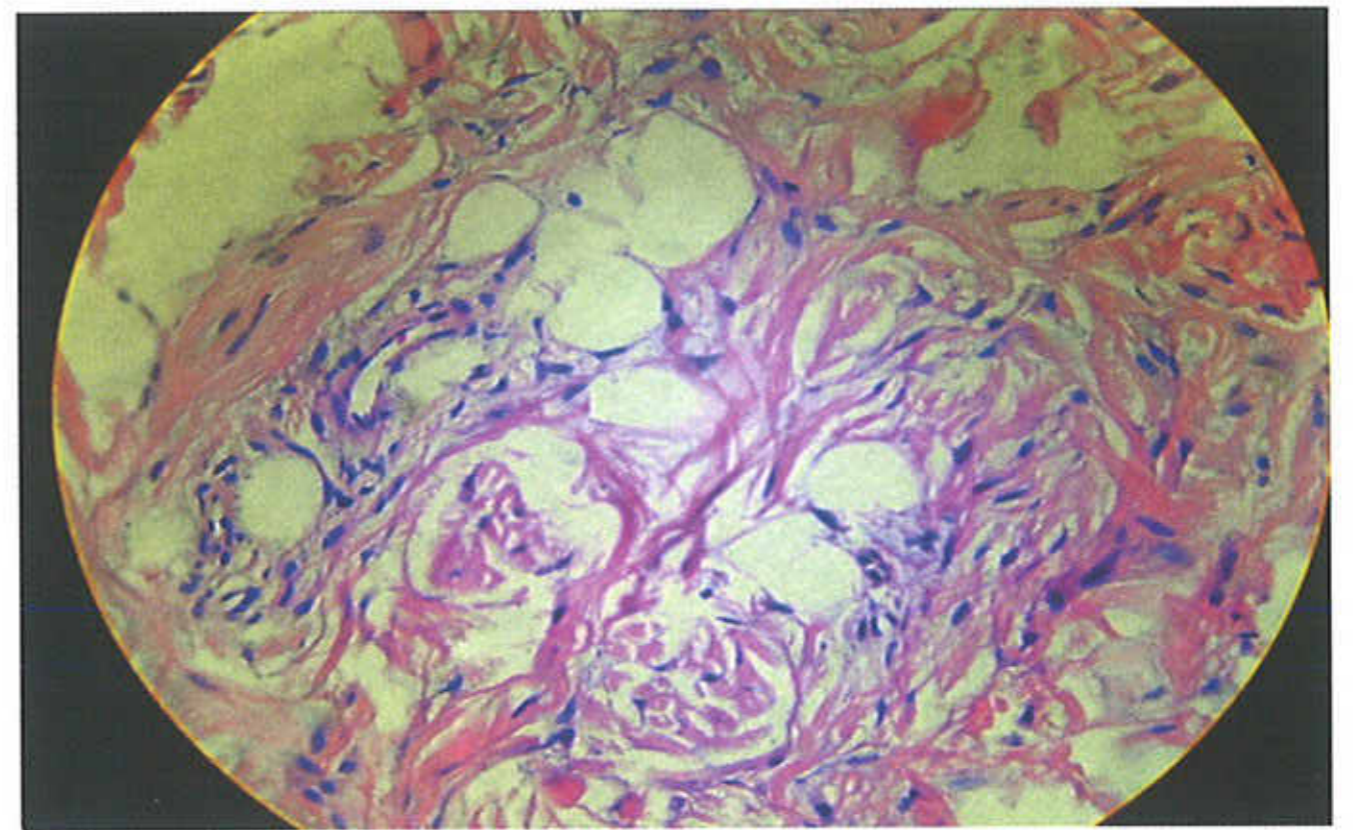
*Fig. 16 : Medium power view 10x of the surface hyperkeratosis, acanthosis in the form of elongated, branching and anastomosing rete-ridges.
(H & E stain; original manifestation x 60).*



*Fig. 17 : Medium power view 10x of the central stalk showing helered sclattered collagen bundles, blood vessels and scattered adipocytes.
(H & E stain; original manifestation x 60).*



*Fig. 18 : Higher power view 20x of scattered adipocytes within collagen bundles.
(H & E stain; original manifestation x 120).*



*Fig. 19: High power view 40x of adipocytes within collagen bundles.
(H & E stain; original manifestation x 240).*

Case Four:

A 41-year-old Iraqi female patient not known to have any chronic illnesses before presented to the dermatology clinic with asymptomatic warty lesion of the second web space (between the ring and middle finger) of the right hand (Fig. 20, 21).

The lesion was of years duration, excised surgically and sent for histopathological studies. The findings as in Diagnosis as acquired digital fibrokeratoma (Fig. 22, 23, 24, 25, 26).



Fig. 20 Warty lesion



Fig. 21 Warty lesion



Fig. 22: Low power view 4x of the polypoid lesion covered by hyperkeratotic stratum corneum, with a central fibrovascular stalk. (H & E stain; original manifestation x 24).

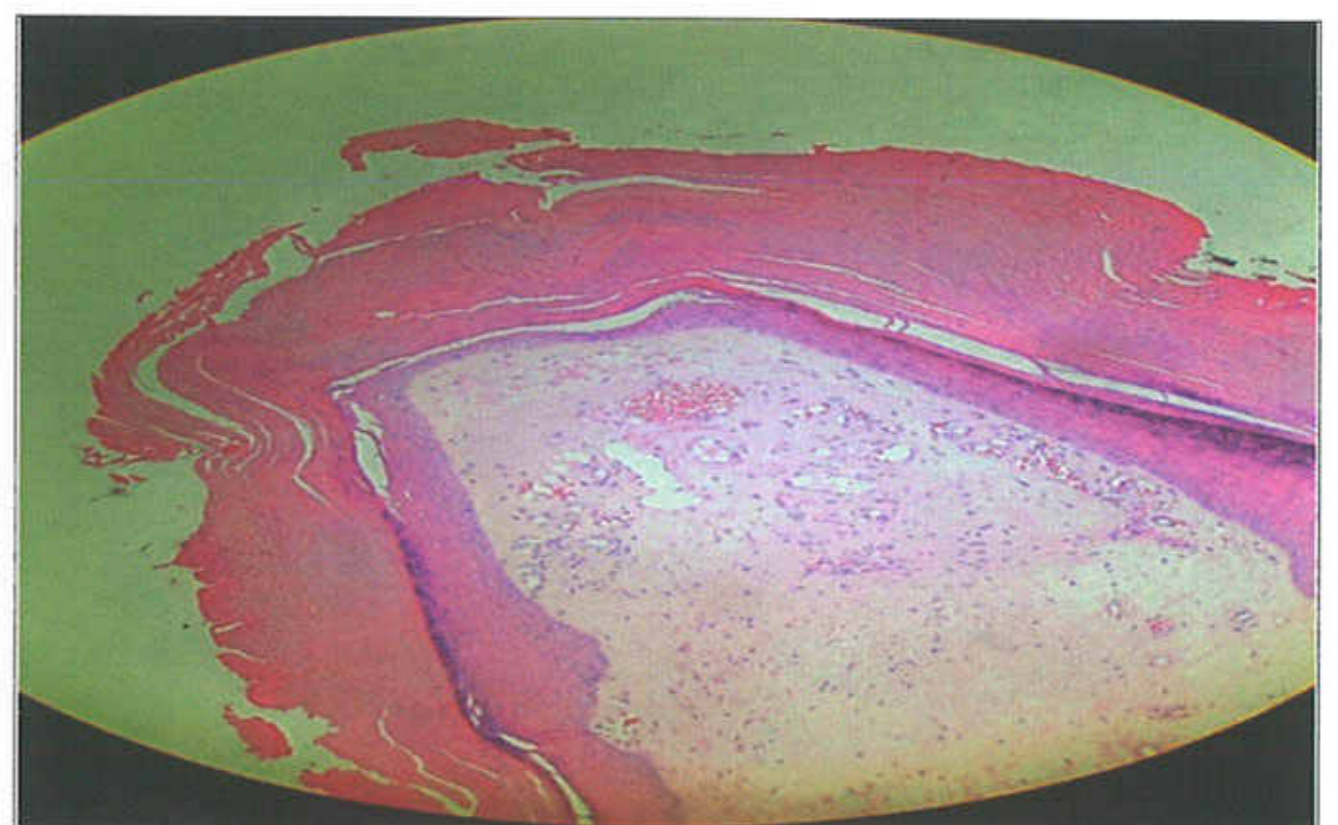


Fig. 23: Medium power view 10x of the apex showing thick hyperkeratotic stratum corneum, slightly acanthotic stratum spinosum with a central fibro-vascular stalk. (H & E stain; original manifestation x 60).

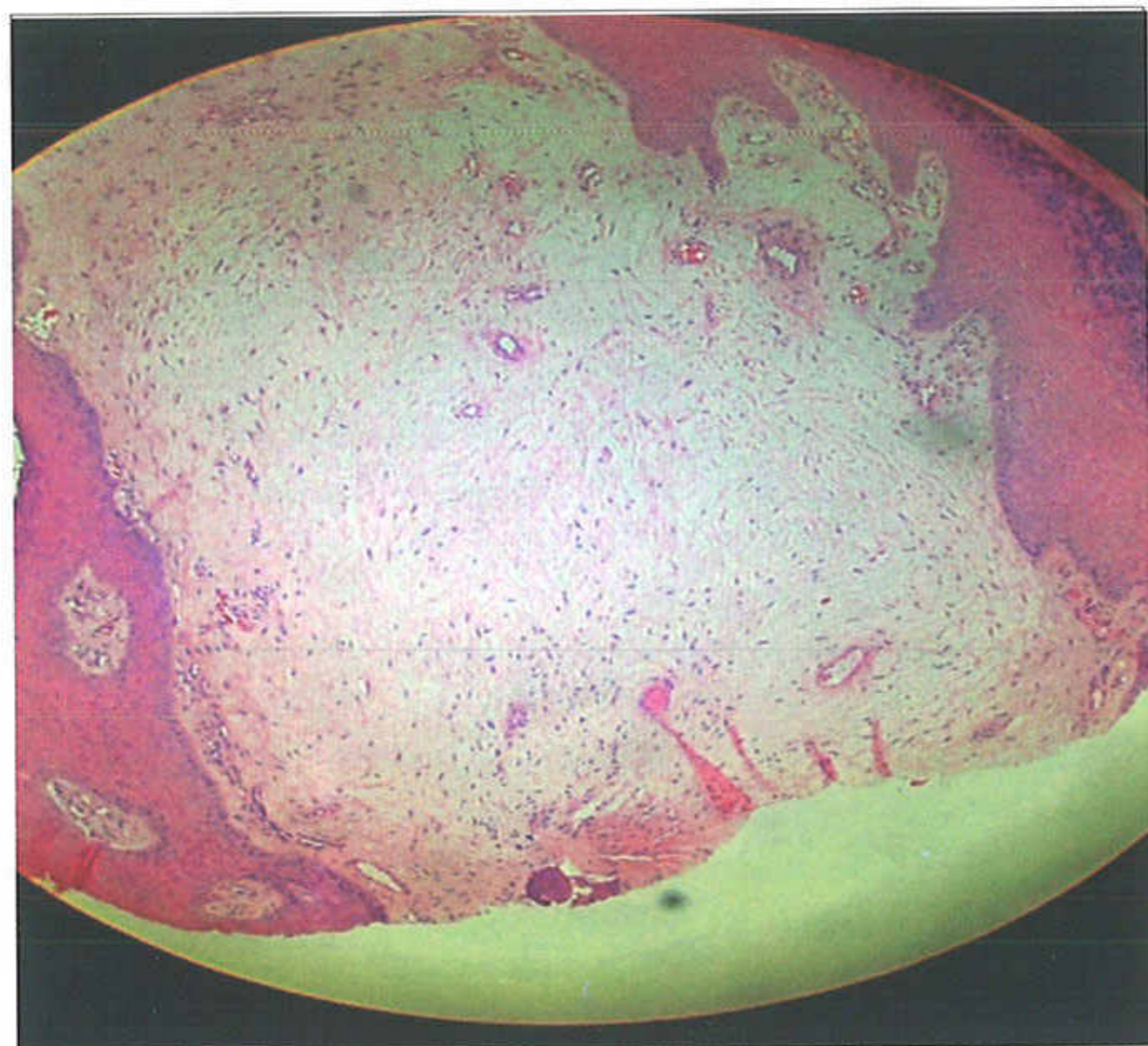


Fig. 24: Medium power view 10x of the basal portion of the lesion showing central fibro-vascular stalk note the absence of neutral structure.
(H & E stain; original manifestation x 60).

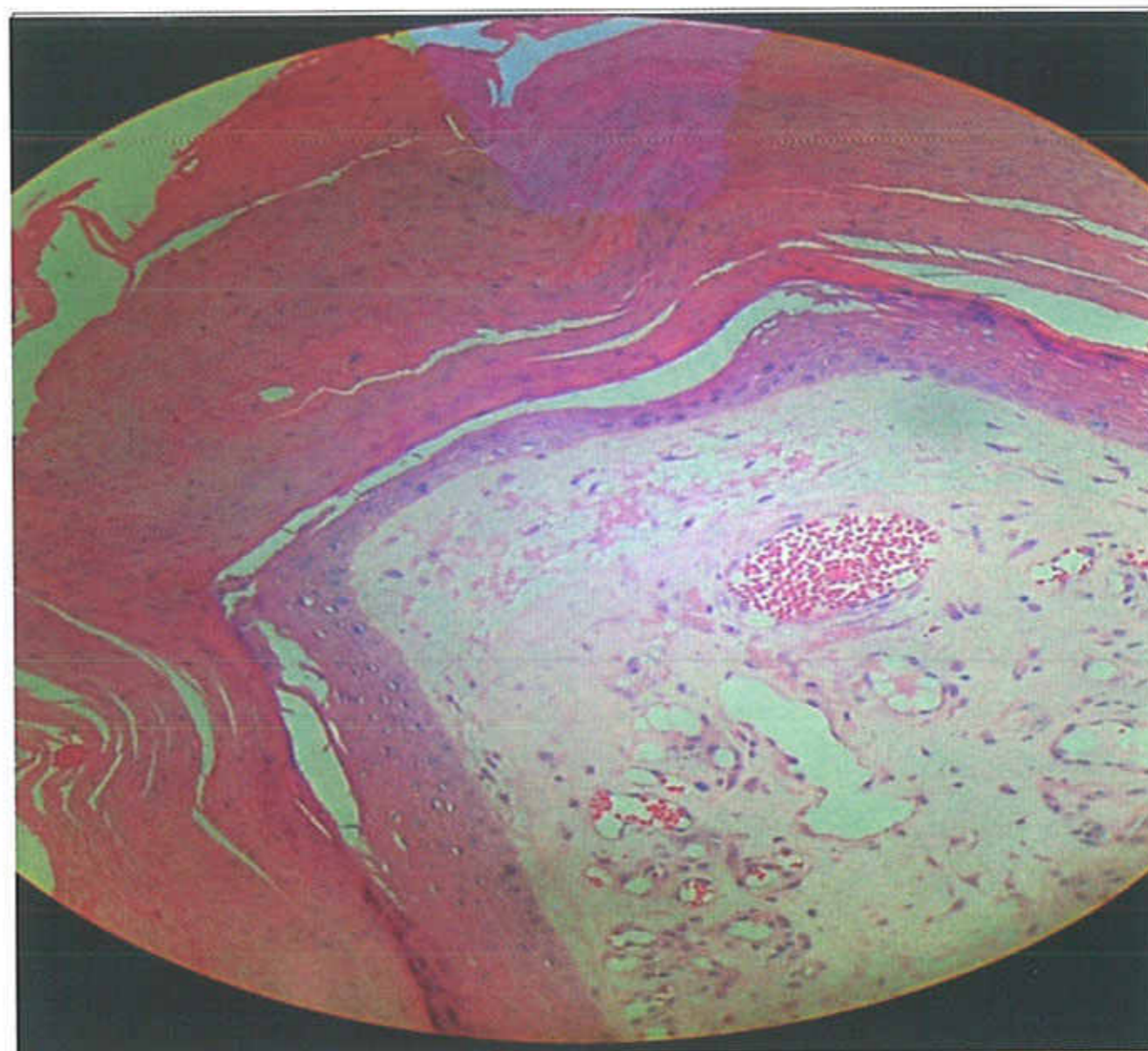


Fig. 25: Higher power view 20x of the apical part of the lesion showing markedly thick hyperkeratotic stratum corneum with an underlying focally atrophic stratum spinosum & a central fibrovascular stalk with telangiectatic thin wall vascular channels.
(H & E stain; original manifestation x 120).

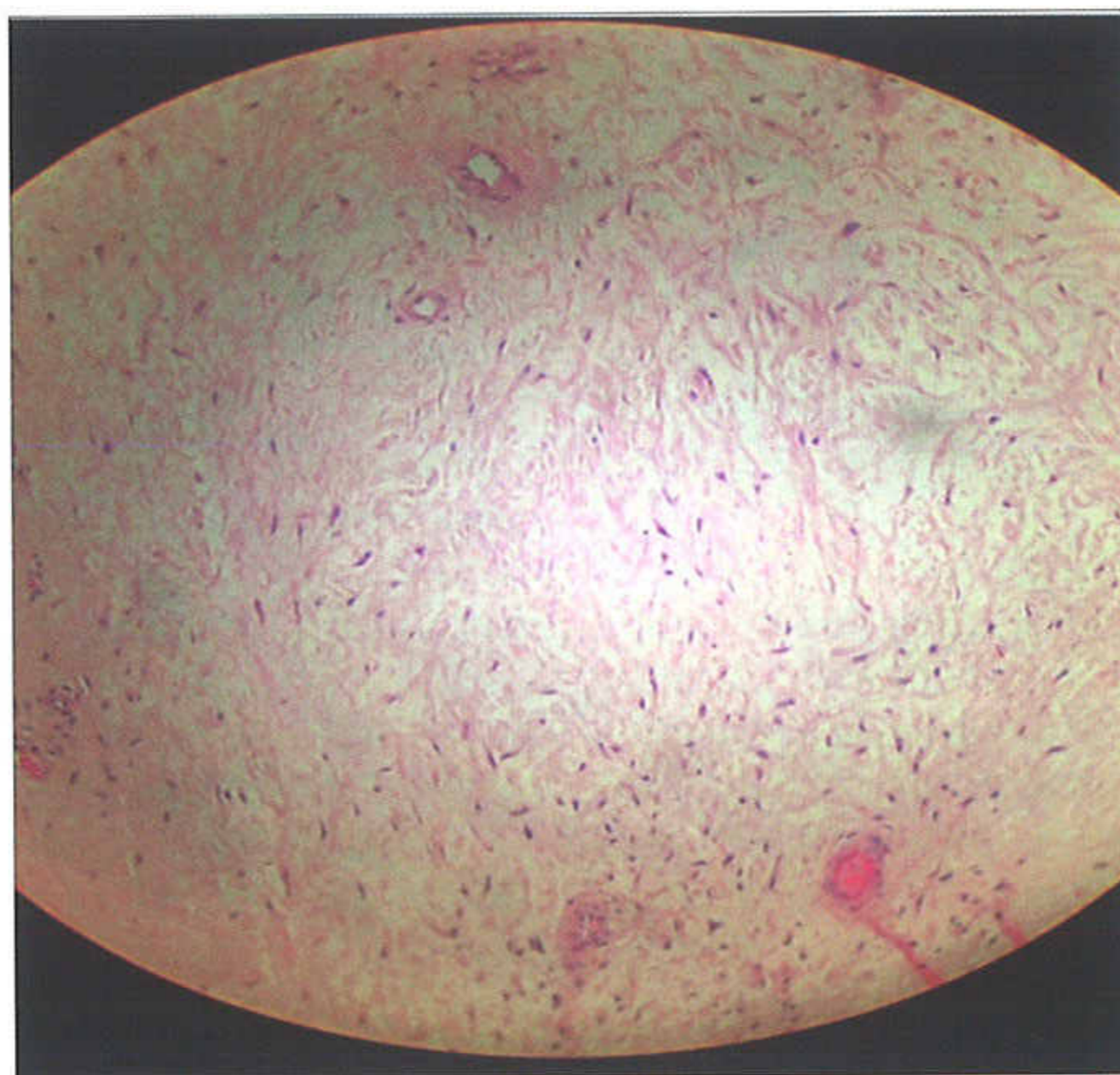


Fig. 26: High power view 20x showing slightly cellular collagenous stroma with vague parallel arrangement of the collagen bundles.
(H & E stain; original manifestation x 120).

Discussion

The clinical features of the cases of acquired digital fibrokeratoma described and reported in this text are shown in Figures (1, 12, 13, 20, 21). Each patient presented with a small filiform solitary mass involving a finger. One of them showed the typical collarette of slightly raised skin (Fig. 1), which is a unique characteristic and diagnostic sign.

Despite that all the cases were acquired digital fibrokeratoma yet one of them was diagnosed histopathologically to be an accessory digit showing the characteristic feature of an increase in the number of nerve bundles (Fig. 8, 9, 10, 11).

None of our cases dated their lesions to birth or infancy, a fact that characterizes rudimentary digit from acquired digital fibrokeratoma and other similar lesions. We suspect that in the case diagnosed histopathologically as an accessory finger the lesion was amputated traumatically in early childhood and such event was unnoticed. The remaining stump appeared clinically as an acquired digital fibrokeratoma and the histopathology was the clue for the correct diagnosis.

The supernumerary digit is often transmitted as an autosomal dominant trait and it is essentially present since birth as a small skin colored pedunculated outgrowth arising usually from the surface of the little finger. It may contain cartilage or have a rudimentary nail. The commonest site is the ulnar border of the hand or near or at the base of the digit and may be bilateral. The digit may be soft and in some cases have a warty surface. The treatment of supernumerary digit is excision for cosmetic reason. Histopathological examination is needed to differentiate it from acquired digital fibrokeratoma since both nearly have the same clinical appearance.

The acquired digital fibrokeratoma may require to be differentiated from multiple vascular fibromas and myxoid fibromas⁽²⁴⁾. It has also to be identified from familial multiple acral mucinous fibrokeratoma⁽²⁵⁾ a condition which has an autosomal dominant inheritance trait and is characterized by multiple verrucous lesions that appear on hands and fingers and was reported to affect three generations in a family (a mother, her son and grand daughter) and they presented since early childhood⁽²⁵⁾. Such lesions highly resembled fibrokeratomas^(8,13). Clinically they were skin colored and were approximately 1 mm in diameter and 4 mm high and the diameter may reach 7 mm⁽²⁵⁾.

Histologically the mucinous fibrokeratoma showed

orthokeratotic hyperkeratosis, irregular acanthosis and papillomatosis with elongated rete ridges and no evidence of human papilloma virus infections. The dermis was highly vascular with mucinous deposit. The number of such lesions on hands and fingers may reach up to 150 or 300 lesions. Such lesions are differentiated clinically and histopathologically from acrokeratosis verruciformis and papular acrokeratoderma.

Acquired digital fibrokeratoma are acquired benign tumors of the fingers and toes. Trauma has been cited as a promoting factor in several cases⁽¹⁷⁾. One lesion was reported to appear after the finger was caught in a car door⁽¹⁸⁾. The lesions are usually not painful or itchy. Acquired digital fibrokeratomas have to be differentiated also from warts, pyogenic granuloma, cutaneous horns, Koenen's tumors and superficial acral fibromyxoma⁽²⁶⁾ which is clinically similar to acquired digital fibrokeratoma but histologically show moderate cellular proliferation of spindle fibroblast like cells embedded in a predominantly myxoid, myxocollagenous or predominantly collagen matrix.

Such tumor may extend to subcutis with occasional presence of pleomorphic cells thus supporting the view that acral fibromyxoma is a neoplastic process hence the necessity of histopathologic differentiation.

Three histological variations of acquired digital fibrokeratomas were described⁽¹³⁾. The first and most common histopathologic finding usually includes a hyperkeratotic corneal layer and thin spinous layer, which cover a well-defined thick, interwined dermal collagen bundles oriented along the vertical axis of the lesion⁽²⁷⁾. Elastic fibers are usually present but are thin⁽²⁷⁾. There is also an increase in the vascularity compared to normal tissue, there is increased number of fibroblasts and vessels between collagen bundles. The second type is described to have an increase of fibroblasts along the vertical axis of the tumor with marked decrease in the elastic fibers and the third type is rare and shows edematous connective tissue and separation of collagen bundles with few elastic fibers.

Our histological findings as shown (Fig. 2, 3, 4, 5, 6, 7, 14, 15, 16, 17, 18, 19, 22, 23, 24, 25, 26) are compatible with the first histopathologic pattern⁽¹³⁾. Cryotherapy or topical keratolytic agents are unsuccessful⁽¹⁵⁾ but the lesion may recur after surgical excision⁽²⁸⁾.

Conclusion:

Four cases clinically diagnosed as acquired digital fibrokeratoma were described. The histopathology

showed three to be of typical pattern and one was an accessory finger. This point that histopathology is essential to differentiate acquired digital fibrokeratomas from many clinically similar conditions. Acquired digital fibrokeratoma is a benign tumor like lesion mostly arising from fingers out of a slightly raised collarette.

Fibrokeratomas apart from fingers may arise from subungual area^(5,6) and hence necessitate differentiation

from Koenen's tumor of tuberous sclerosis. Fibrokeratomas may be Giant and affect heel and sole^(7,9,10,11). Acquired digital fibrokeratomas may be exceptionally big and pedunculated⁽¹²⁾. The clinician should be aware of the clinical variants of acquired digital fibrokeratomas and its three main histological types⁽¹³⁾.

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