

## The Myxoid Extremities- Superficial Acral Fibromyxoma Segregated

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### Preface

Superficial acral fibromyxoma was initially scripted by Fetsch., *et al.* in 2001 and was described as an exceptional, benign neoplasm with a bland, fibromyxoid histology. The neoplasm is also designated as digital fibromyxoma and demonstrates a preferential incrimination of subungual, periungual or acral regions of hands and feet. Additionally, adjunctive areas such as legs, thighs, heel, palm and ankles are incriminated [1]. On account of heterogeneous clinical presentation and histological characteristics, superficial acral fibromyxoma mandates a demarcation from diverse acral lesions of hand and feet. The lesion preponderantly arises from cutaneous and subcutaneous tissue planes and abuts the phalanges, digits, joint capsule and collateral ligament [1].

### Disease characteristics

Superficial acral fibromyxoma can exclusively arise upon hands and feet, especially the toes. An estimated 45.8% subjects demonstrate lesions upon the feet or toes and are termed as pododactyls whereas neoplasms situated upon the fingers are discerned in around 39.1% candidates, referred to as chirodactyls. The neoplasm shares characteristics with several benign pathologies which can delay and challenge a definitive diagnosis. Diagnosis can be delayed on account of indolent nature of the lesion and minimal morbidity encountered [1,2].

Duration betwixt commencement of distinctive symptomatology and specific diagnosis can exceed > 10 years. Neoplasms generally appear within the fifth or sixth decade and mean age of tumour diagnosis is 47 years 7 months although neoplastic incrimination can occur betwixt 4 years to 91 years. The neoplasm demonstrates a male predominance with a male to female proportion of 2:1 and arises in an estimated 61% of males and 39% of females. Approximately 41% of lesions are accompanied by pain [2].

Irrespective of encountered cellular atypia, the tumour is devoid of aggressive biological behaviour or a malignant metamorphosis although the proportionate reoccurrence varies betwixt 10% to 24% [2].

### Clinical elucidation

The neoplasm commonly manifests as an asymptomatic, gradually evolving, painless nodule preponderantly situated upon the acral region of extremities although the heel, ankle, calves and thigh region can be incriminated. Superficial acral fibromyxoma manifests as a gradually enhancing, solitary, painless, soft to firm to cystic tumefaction with a focally gelatinous consistency and a varying magnitude [3].

The neoplasm can also represent as a miniature, painful, tender, non-fluctuant nodule preponderantly situated upon the fingers, particularly the middle phalanx or interphalangeal region. The nodule delineates a well-defined perimeter with a variegated, irregular extra-neous surface. The lesion can appear in the absence of local trauma and is devoid of concurrent neurovascular deficit [2,3].

### Histological elucidation

Macroscopically, a firm, nodular or lobulated soft tissue neoplasm of varying magnitude with a cutaneous covering is enunciated. The nodule can be dome shaped, polypoid or verrucous. Cut surface is greyish/white with a firm, solid or gelatinous consistency and demonstrates multiple cystic zones impacted with glistening, whitish substance [3,4].

On microscopic examination, superficial acral fibromyxoma denominates a well circumscribed, non-encapsulated dermal or subcutaneous neoplasm comprised of stellate or spindle shaped fibroblastic cells demonstrating a random, loosely configured fascicular arrangement, a variable intermingling of myxoid or collagen matrix, varying pleomorphism and an encompassing myxoid, myxo-collagenous or collagen stroma. Vascular configurations predominate within myxoid zones and mast cells are intermingled throughout the lesion [3,4].

Lesions of significant duration demonstrate a predominance of collagen, in contrast to a myxoid matrix. The neoplasm is associated with significantly enhanced micro-vascularization and ectatic blood vessels are augmented. Mast cells are prominent although mitotic figures are infrequent and nuclear atypia is mild to moderate [3,4].

A hyperkeratotic, stratified squamous epithelial layer is exemplified, superimposed upon a lesion composed of spindle -shaped and stellate cells configured in sheets, fascicles and a vaguely defined, whorling pattern. An intermingled, abundant, myxomatous stroma, intervening fibrous tissue septa and occasional multinucleated stromal giant cells are enunciated [4,5].

An accompanying inflammatory infiltrate is typically absent. The preponderantly dermal neoplasm can extend into the subcutaneous tissue, underlying fascia, periosteal layer or bones. Incrimination of periosteum ensures erosion of the underlying bone [2,4].

### Immune histochemical elucidation

The neoplasm demonstrates an immune reactivity to CD34, CD99 and vimentin along with a focal immune reactivity to epithelial membrane antigen (EMA) and CD10.

Immune reactivity to CD34 is observed in an estimated 91.3% instances, epithelial membrane antigen (EMA) in around 72% subjects and CD99 in approximately 84.6% tumours. Immune reactivity to CD34 and CD99 is diffuse and strong whereas EMA demonstrates a variable immune reaction. Although smooth muscle actin, desmin, S100 protein, glial fibrillary acidic protein (GFAP), apolipoprotein D, mucin 4(MUC4), claudin, non-phosphorylated (NFP) immune stain, human melanoma black 45 (HMB-45) and various cytokeratin molecules are immune non-reactive, focal reactivity for smooth muscle actin(12%) and desmin (6%) can be discerned. Mucinous material can be highlighted with stains such as alcian blue [2,4].

### Differential diagnosis

Superficial acral fibromyxoma necessitates a distinction from a gradually progressive mass or nodule situated within the periungual or subungual region of fingers and toes. Segregation from various fibromyxoid or myxoid cellular proliferations is mandatory.

1. Myxoid neurofibroma is a solitary lesion appearing upon the hand and feet and usually delineates a neural appearance in the absence of enhanced vascularization. Tumour cells are characteristically immune reactive to S100 protein and are intermingled with fibroblastic cells immune reactive to CD34 whereas superficial acral fibromyxoma is immune non-reactive to S100 protein and depicts an intense, concentrated microvasculature [6,7].
2. Fibroma of the tendon sheath is a solitary, gradually progressive, subcutaneous tumefaction delineating a predilection for fingers, hands and wrists. The tumefaction is greyish white, well circumscribed and adheres to a tendon sheath. In contrast to a cellular proliferation composed of stellate or spindle- shaped, fibroblastic cells, aforesaid tumour demonstrates star- shaped cells embedded in a fibro-collagenous matrix with accompanying dilated or slit- like vascular channels [7].

3. Glomus tumour engenders as a solitary, extremely painful subcutaneous nodule generally incriminating fingers, toes and the subungual region. Tumour cells are immune reactive to intracytoplasmic vimentin and smooth muscle actin.
4. Acquired digital fibrokeratoma is a solitary, pauci-cellular tumour of fingers and toes which may or may not be immune reactive for CD34, although is immune non-reactive to epithelial membrane antigen (EMA).
5. Superficial angiomyxoma is localized upon the head, neck and trunk and lesions are immune reactive to CD34 and S100 protein [7,8].
6. Sclerosing perineurioma is a neoplasm wherein the cellular component configures distinct cords and trabeculae along with an onion-skin pattern. It is a benign, acral fibrous tumour which is immune reactive to epithelial membrane antigen (EMA), claudin and immune non-reactive to S100 protein and CD34 [6,8].
7. Cutaneous myxoma or superficial angiomyxoma is a poorly demarcated neoplasm demonstrating a lobular pattern and may demonstrate an epithelial component. An association with Carney's complex can be observed [8].
8. Fibrous histiocytoma as a neoplasm is poorly circumscribed and immune non-reactive to CD34. Typically, lesions are absent upon the fingers, palms and soles.
9. Giant cell tumour of the tendon sheath is a gradually progressive, benign tumefaction arising in young adults and delineating a predilection for fingers and interphalangeal joints. Tumours are lobulated, greyish brown and usually attached to a tendon sheath. Histological examination enunciates spindle-shaped cells with an admixture of collagenous stroma and characteristic multinucleated giant cells. Haemosiderin pigment, xanthoma cells and chronic inflammatory cells are commonly exemplified. Mononuclear cells are immune reactive to CD68 although epithelial membrane antigen (EMA), CD34 or smooth muscle actin are immune non-reactive [6,7].
10. Dermatofibrosarcoma protuberans is an exceptional lesion to arise upon hands and feet. The neoplasm is infiltrative and depicts a tightly woven cellular component with a focal storiform pattern. Dermatofibrosarcoma protuberans can display extensive foci of myxoid areas simulating superficial acral fibromyxoma. Combined immune reactivity of aforesaid lesions to CD34 and epithelial membrane antigen (EMA) can ensure a challenging differential diagnosis [5,6].
11. Myxoid variant of dermatofibrosarcoma protuberans is an exceptional, aggressive mesenchymal tumour which is preferentially situated upon the centroidal zones. The lesion is immune reactive to CD34 and immune non-reactive to S100 protein. xii) Low grade fibromyxoid sarcoma is exceptionally discerned upon the acral regions [7].
12. Inflammatory myxohyaline tumour characteristically depicts virocytes or Reed-Sternberg cells. Adjunctive myxoid sarcomas such as myxofibrosarcoma requires a distinction from superficial acral fibromyxoma. Myxoid sarcomas display enhanced cytological atypia and augmented mitotic figures with atypical mitotic forms [5,6]. xvi) Myxoinflammatory fibroblastic sarcoma is enunciated as an inflammatory, myxohyaline tumour of distal extremities and is comprised of virocyte-like and lipoblast-like bizarre cells and can be associated with significant inflammation. The neoplasm is immune non-reactive to epithelial membrane antigen (EMA) [8].

Differentiation is also necessitated from periungual and subungual fibroma or dermal mucinosis [7,8]. Investigative Assay Apart from a cogent clinical and histological enunciation, auxiliary diagnostic modalities can be employed to categorize superficial acral fibromyxoma such as plain radiography, ultrasonography and nuclear magnetic resonance (NMR) imaging.

- Plain X-ray of implicated site can determine bony incrimination and an estimated 36% instances demonstrate erosive or lytic bone lesions. Nevertheless, calcification within soft tissue nodules may not be discernible with singular, plain radiographs. However, as the neoplasm engenders erosion of underlying cortical bone, adoption of a simple radiograph is essential to assess bone involvement, estimated extent of lesion and for appropriate therapeutic planning [7,8]. Plain radiograph depicts a homogenous, soft tissue mass within the incriminated phalanx with an absence of bony involvement.

- Ultrasonography exemplifies magnitude, location, outline and content of the tumour nodule. Additionally, monitoring of tumefaction and relapse of lesions treated with surgical eradication can be achieved.
- Colour Doppler can assess tumour vasculature with evaluation of specific, demonstrable vascular pattern which can aid therapeutic decisions [7,8].
- Magnetic resonance imaging (MRI) of superficial acral fibromyxoma typically displays a homogenous, hyper-intense signal within T2 weighted imaging and an enhanced contrast within normal soft tissue upon post-contrast MRI.

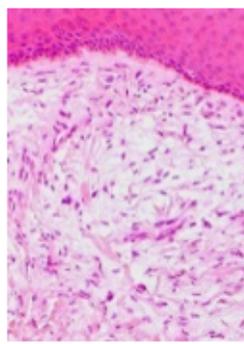
Magnetic resonance imaging (MRI) reveals a complex lesion with multiple locules and a hypo-intense T1 weighted imaging with concurrent hyper-intense T2 weighted imaging along with an intensely illuminated image upon inversion recovery (IR) sequence [7,8].

Thin, hypo-intense septa admixed with attenuated, hypo-intense foci possibly represent calcification. Delayed, intense image enhancement is enunciated upon contrast administration [9,10].

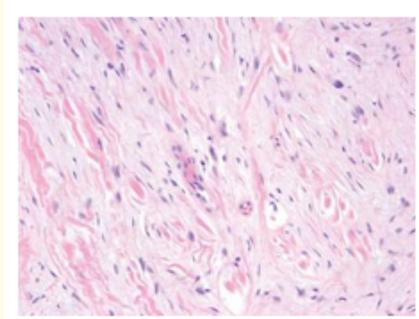
Apart from a competent histological categorization and pertinent immune histochemical assessment, employment of magnetic resonance imaging (MRI), especially T1 weighted and T2 weighted imaging is considered optimal in diagnosing and demarcating superficial acral fibromyxoma from adjunctive neoplasia [9,10].

### Therapeutic options

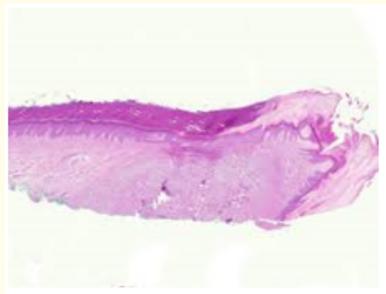
Preferential mode of therapy is a comprehensive surgical extermination of the neoplasm in order to circumvent disease relapse or malignant conversion. Surgical eradication of the neoplasm along with a significant margin of normal, uninvolved tissue is recommended. Surgical defect engendered upon the superimposed cutis and soft tissue can be repaired by a full thickness cutaneous graft. Although the neoplasm is devoid of aggressive biological behaviour, infrequent emergence of cytological atypia can be indicative of a dubious potential for malignant transformation, even though malignant conversion remains undetermined [10]. Adequate post-operative monitoring of the neoplasm is recommended in order to reduce proportion of tumour relapse from an estimated 24% which is exemplified following incomplete surgical excision. Local reoccurrence is almost comprehensively associated with inadequate surgical excision and tumour entrapped within the surgical perimeter [11].



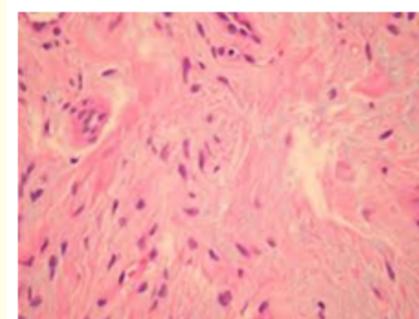
**Figure 1:** Superficial acral fibromyxoma with a dual component of stellate and spindle shaped cells, intermingled mast cells and a superimposed stratified epidermis [12].



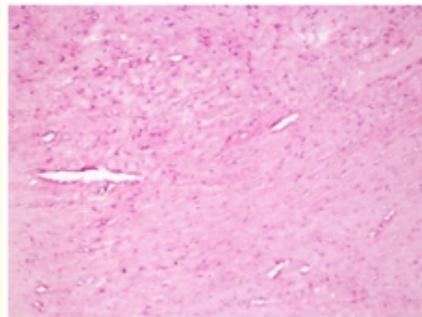
**Figure 2:** Superficial acral fibromyxoma with commingled stellate and spindle shaped fibroblastic cells, dispersed mast cells and ectatic blood vessels [13].



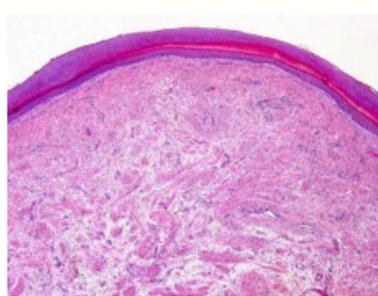
**Figure 3:** Superficial acral fibromyxoma with a nodule demonstrating a superimposed squamous epithelial lining and intermixed star-shaped and spindle fibroblastic cells, few mast cells and a lack of inflammatory cells [14].



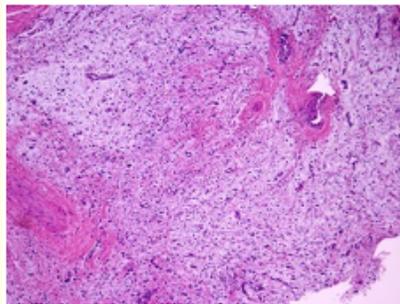
**Figure 4:** Superficial acral fibromyxoma with admixture of spindle and stellate cells, mast cells and a loose, fascicular arrangement [14].



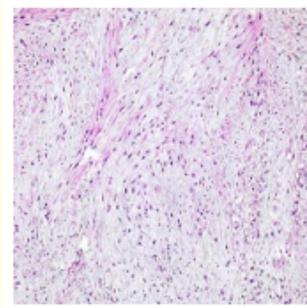
**Figure 5:** Superficial acral fibromyxoma with intermixed spindle and star-shaped cells, numerous mast cells and ectatic vasculature [15].



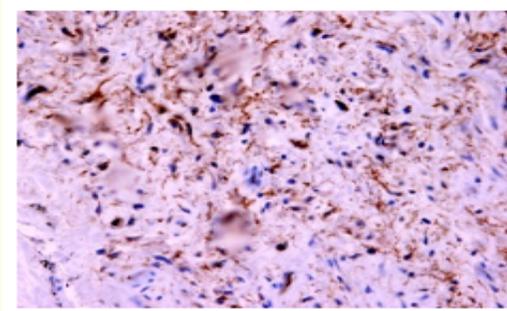
**Figure 6:** Superficial acral fibromyxoma with admixed star and spindle shaped cells, mast cells, dilated vasculature and a superimposed stratified squamous epithelial lining [16].



**Figure 7:** Superficial acral fibromyxoma with a fascicular arrangement of spindle and stellate cells, foci of myxoid stroma and intermingled mast cells [17].



**Figure 8:** Superficial acral fibromyxoma with commixture of spindle and stellate cells, mast cells and a myxo-collagenous stroma [18].



**Figure 9:** Superficial acral fibromyxoma immune reactive to CD34+ [19].

Neoplasms CD34+	Neoplasms CD34-	Associated Lesions
Myxoid neurofibroma	Giant cell tumour of the tendon sheath	Fibroma of the tendon sheath
Superficial angiomyxoma	Glomus tumour	Onicocryptosis
Dermatofibrosarcoma protuberans	Sclerosing perineurioma	Cutaneous myxoma
Sclerotic fibroma	Benign fibrous histiocytoma	
Acral myxoinflammatory	Acral fibrokeratoma	
Spindle cell lipoma		

**Table:** Differential diagnosis of superficial acral fibromyxoma [2].

**Conclusion**

Superficial acral fibromyxoma is described as an exceptional, benign neoplasm with a bland, fibromyxoid histology. An estimated 45.8% lesions appear upon feet or toes, termed as pododactyls and around 39.1% neoplasms are situated upon fingers, referred to as

chirodactyls. Superficial acral fibromyxoma manifests as a gradually progressive, solitary, painless, soft to firm to cystic, focally gelatinous tumefaction of variable magnitude. Morphological evaluation depicts a well circumscribed, non-encapsulated dermal or subcutaneous neoplasm constituted of stellate or spindle- shaped, fibroblastic cells in a random, loosely configured fascicular arrangement and a variable intermingling of myxoid or collagen matrix with an encompassing myxoid, myxo- collagenous or collagen stroma. Superficial acral fibromyxoma is immune reactive to CD34, CD99 and vimentin with a focal immune reactivity to epithelial membrane antigen (EMA) and CD10. Superficial acral fibromyxoma necessitates a distinction from fibroma of the tendon sheath, acquired digital fibrokeratoma, periungual and subungual fibroma, superficial angiomyxoma, dermal mucinosis, myxoid neurofibroma, sclerosing perineurioma, myxoid dermatofibrosarcoma protuberans, low grade fibromyxoid sarcoma, glomus tumour, giant cell tumour, fibrous histiocytoma, cutaneous myxoma, inflammatory myxohyaline tumour of distal extremities or myxoinflammatory fibroblastic sarcoma. Cogent clinical or histological enunciation, plain radiography, ultrasonography and magnetic resonance imaging (MRI) can be adopted to adequately categorize the neoplasm. Comprehensive surgical extermination of the neoplasm is a preferred therapy.

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12. Image 1 Courtesy: Wordpress.com.
13. Image 2 Courtesy: MDEdge.com.

14. Image 3 and 4 Courtesy: Dermnet.NZ.com.
15. Image 5 Courtesy: Science direct.
16. Image 6 Courtesy: Pathpedia.com.
17. Image 7 Courtesy: escholarship.com.
18. Image 8 Courtesy: Basic medical key.
19. Image 9 Courtesy: Pathology outlines.

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