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Case Report

Surgery Section

An Unusual Acral Swelling: Histopathological Confirmation of Superficial Acral Fibromyxoma

CHAITANYA MAHAJAN[1], VARUN SHETTY[2], IQBAL M ALI[3]

ABSTRACT

Superficial Acral Fibromyxoma (SAF) is an uncommon, benign mesenchymal neoplasm that predominantly involves the subungual and periungual soft tissues of the digits. Its indolent growth, painless nature, and non specific clinical features often lead to misdiagnosis, resulting in delayed treatment or inappropriate management. The present case described a 36-year-old male who presented with a decade-long history of a gradually enlarging, painless mass on the distal phalanx of the right ring finger, followed by a recent phase of accelerated growth over two months. Magnetic Resonance Imaging (MRI) demonstrated a well-circumscribed, T2-hyperintense lesion without underlying osseous involvement. Complete surgical excision was performed. Histopathological evaluation revealed a proliferation of bland spindle and stellate fibroblastic cells embedded in a fibromyxoid stroma, without evidence of cytologic atypia or mitotic activity. Immunohistochemical analysis showed diffuse positivity for CD34 and vimentin, with negative staining for S-100 and Epithelial Membrane Antigen (EMA), supporting the diagnosis of SAF. The postoperative course was uneventful, and there has been no evidence of recurrence to date. Given the potential for local recurrence, especially in incompletely excised lesions, accurate diagnosis through histopathology and Immunohistochemistry (IHC) is essential. Awareness of SAF's distinguishing features among clinicians, radiologists, and pathologists can aid in timely diagnosis, reduce the risk of misclassification, and facilitate appropriate surgical management, ultimately improving patient outcomes.

Keywords: Benign, Immunohistochemistry, Soft-tissue neoplasms, Subungual

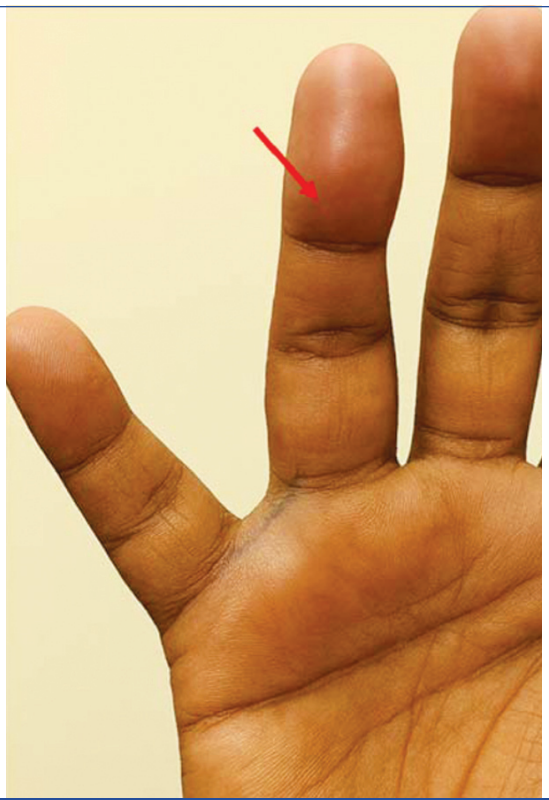
CASE REPORT

A 36-year-old male presented to the outpatient surgical department with complaint of a swelling over the distal aspect of his right ring finger [Table/Fig-1]. The lesion had been insidiously progressive over a span of 10 years, with no history of antecedent trauma, pain, discharge or functional impairment. He reported a rapid increase in the size of the swelling over a span of two months, which brought him to visit Dr. D. Y. Patil Hospital and Research Centre. There were no systemic complaints, and his personal and family medical history was non contributory.

On local examination, a well-circumscribed, globular swelling measuring approximately 2×3 cm was noted over the distal phalanx of the right ring finger. The swelling was firm in consistency, non tender and exhibited a smooth surface with thinning and broadening of the overlying nail plate. The skin over the lesion appeared unremarkable, and range of motion in the involved digit was preserved. Based on the clinical features and anatomical location, a provisional diagnosis of acral lipoma was considered, and further evaluation was undertaken accordingly.

Routine laboratory investigations, including complete blood count, liver and renal function tests, coagulation profile, and serum electrolytes, were within normal reference ranges. The MRI of the right hand revealed a hyperintense soft-tissue lesion on T2-weighted sequences, located in the volar and periungual region of the distal phalanx of the right ring finger, with no evidence of underlying cortical erosion or bony involvement [Table/Fig-2]. In view of the benign nature and absence of alarming features, the decision was made to proceed with surgical excision under regional anaesthesia.

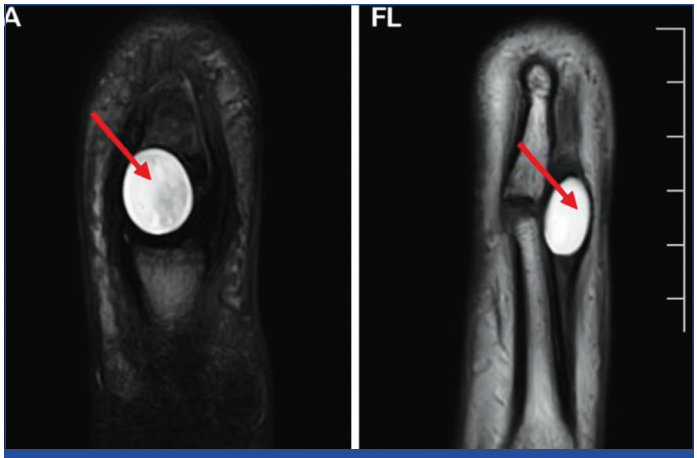
Intraoperatively, the lesion was excised in toto and sent for histopathological evaluation. Microscopic examination revealed a well-delineated, hypocellular tumour composed of spindle- and stellate-shaped fibroblast-like cells embedded in a loose fibromyxoid stroma. There was no evidence of nuclear atypia or increased mitotic activity. Immunohistochemical analysis demonstrated strong positivity for vimentin and CD34, while markers such as S-100 and Epithelial Membrane Antigen (EMA) were negative, confirming the diagnosis of SAF [Table/Fig-3]. The postoperative period was uneventful, and the surgical wound healed well with no signs of local recurrence at follow-up after one year.



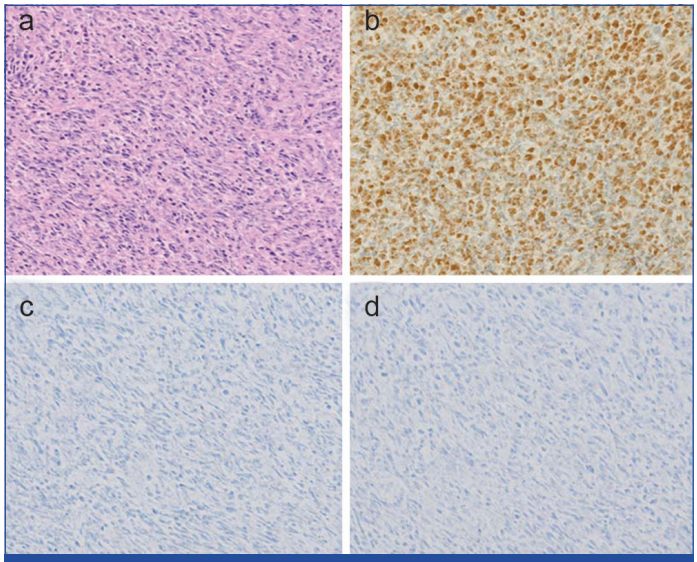
[Table/Fig-1]: Clinical image showing the swelling arising against the distal phalynx of the right ring finger (red arrow).

DISCUSSION

The SAF is an uncommon, benign fibroblastic neoplasm predominantly affecting acral sites, particularly the periungual and subungual regions of the fingers and toes. Initially delineated by



[Table/Fig-2]: T2-weighted MRI images of the affected finger in axial (A) and coronal (FL) planes; A) Axial T2-weighted MRI image showing a well-circumscribed, ovoid hyperintense lesion (red arrow) located in the volar soft-tissue adjacent to the distal phalanx. The lesion demonstrates homogeneous internal signal intensity and is surrounded by hypointense soft-tissue structures; FL) Coronal T2-weighted MRI image showing the same lesion (red arrow) in continuity with the flexor tendon sheath, appearing hyperintense relative to surrounding musculature and bone, without evidence of bony involvement.



[Table/Fig-3]: Histological and immunohistochemical analysis of the excised lesion: a) Showing spindle and stellate-shaped fibroblast-like cells embedded in a loose myxoid matrix (H&E,200X); b) IHC stain for CD34 demonstrates diffuse strong cytoplasmic positivity; c) S-100 stain is negative, excluding neural origin; d) EMA stain is negative, helping differentiate from perineurioma and epithelial lesions.

Fetsch JF et al., in 2001, SAF has since been recognised as a distinct pathological entity characterised by a lobulated architecture composed of bland spindle- and stellate-shaped fibroblastic cells embedded within a fibromyxoid stroma [1]. SAF typically manifests as a slow-growing, painless, and often asymptomatic nodule with variable pigmentation and occasional nail plate distortion. Its deceptively indolent presentation, combined with non specific clinical and radiologic features, frequently results in misdiagnosis with common differentials including digital myxoid cysts, epidermoid inclusion cysts, Giant Cell Tumour of the Tendon Sheath (GCTTS), and, less frequently, Low-Grade Fibromyxoid Sarcoma (LGFMS). These entities overlap in clinical appearance and superficial location, thereby increasing the risk of local recurrence due to inadequate resection [Table/Fig-4] [2-5].

According to the 2024 World Health Organisation (WHO) Classification of Soft Tissue and Bone Tumours, SAF is now recognised as a distinct benign fibroblastic neoplasm with a clear predilection for acral sites, particularly the periungual and subungual regions of the fingers and toes. This updated classification underscores both its site-specific nature and its characteristic immunohistochemical profile, including diffuse CD34 and EMA positivity, with consistent negativity for S-100, desmin, and SMA [6].

The SAF typically presents in adults between the fourth and seventh decades of life and exhibits a slight male predominance. It is most commonly localised to the fingers and toes, but has also been sporadically reported in atypical sites such as the heel, dorsum of the foot, leg, and ankle [7]. The lesion usually manifests as a slow-growing, painless, and well-circumscribed nodule, occasionally associated with nail plate deformity when arising in periungual locations. Variants in clinical morphology, including polypoid, dome-shaped, and verrucous forms, have also been described [8].

Histologically, SAF is composed of bland spindle and stellate cells embedded in a fibromyxoid matrix, often accompanied by a network of thin-walled vasculature and sparse inflammatory infiltrates. Mitotic figures are rare, and nuclear atypia is typically absent. Immunohistochemistry serves as an indispensable tool for diagnostic confirmation. The tumour exhibits strong and diffuse positivity for CD34 and vimentin, while showing negativity for S-100 protein, EMA, and desmin. These immunophenotypic features help differentiate SAF from morphologically similar myxoid neoplasms such as neurofibroma (S-100+), low-grade myxofibrosarcoma (CD34+ but mitotically active), and sclerosing perineurioma (EMA+, Glut-1+) is consistent with the immunohistochemical differentiation

Parameters	Superficial Acral Fibromyxoma (SAFM)	Digital Myxoid (Mucous) Cyst	Giant Cell Tumour of Tendon Sheath (GCTTS)	Epidermoid Inclusion Cyst	Low-Grade Fibromyxoid Sarcoma (LGFMS)
Definition	A rare, benign, soft-tissue tumour of fibroblastic origin affecting the acral (fingertip/toe) regions	A benign cystic lesion arising from the Distal Interphalangeal (DIP) joint or tendon sheath with mucin content	A benign, slow-growing lesion arising from the tendon sheath, commonly on the fingers	A benign cyst filled with keratinous material, resulting from epidermal implantation after trauma	A rare, deep-seated, low-grade malignant soft tissue sarcoma with fibromyxoid histology
Typical location	Periungual or subungual region of fingers or toes, particularly the great toe or middle/ring finger	Dorsal side of distal fingers near the DIP joint, often periungual	Flexor surface of fingers, near interphalangeal joints	Palmar aspect or subungual region of digits	Deep soft tissues of extremities or trunk; rarely acral
Clinical features	Painless, slow-growing, flesh-coloured or erythematous nodule with nail plate deformity in some cases	Translucent, dome-shaped, compressible lesion; may fluctuate in size; may cause nail grooving	Firm, rubbery, non mobile nodule; typically painless and well-defined	Firm, round subcutaneous mass, usually painless unless inflamed	Deep-seated, painless mass; often mistaken for benign lesion due to slow growth
Imaging findings	Well-circumscribed subungual or periungual soft tissue lesion; hyperintense on T2-weighted MRI, non invasive to bone	Fluid-filled cystic lesion on ultrasound/MRI, connected to DIP joint or tendon sheath	Well-defined mass near tendon sheath with low to intermediate signal intensity on MRI	Cystic lesion with central keratinous content; may have a capsule and signal void on MRI	Heterogeneous soft tissue mass with variable signal intensity and occasional calcification on imaging
Histopathology	Spindle-shaped fibroblastic cells in a myxoid to fibrous stroma, positive for CD34 and EMA; negative for S100	Pseudocyst without epithelial lining, filled with mucin and fibroblasts	Multinucleated giant cells, histiocytes, and haemosiderin deposits within a fibrous stroma	Cyst lined by stratified squamous epithelium with laminated keratin debris	Spindle cell proliferation in alternating fibrous and myxoid zones; positive for MUC4 on IHC

Immunohistochemistry (IHC)	CD34 positive, EMA positive, CD99 variably positive; negative for S100, desmin, cytokeratin, and SMA.	Vimentin positive; lacks definitive epithelial or neural markers. No cytokeratin or EMA expression.	CD68 and CD163 strongly positive in histiocytes and giant cells; S100 and desmin negative.	Strongly positive for pancytokeratin (AE1/AE3) and EMA; S100, CD34 negative.	MUC4 strongly positive; also expresses vimentin; negative for S100, desmin, CD34, cytokeratin.
Treatment	Complete surgical excision with clear margins to prevent local recurrence.	Aspiration, steroid injection, or surgical excision; high recurrence if underlying joint connection persists.	Surgical excision; low recurrence with complete resection.	Surgical excision; recurrence rare if excised completely.	Wide surgical excision with oncologic margins; long-term surveillance recommended.
Prognosis	Excellent; benign with potential for local recurrence if incompletely excised.	Benign; recurrence common if not addressed at joint connection.	Benign; recurrence rate up to 10-20%.	Benign; rarely recurs after complete excision.	Indolent but malignant; capable of local recurrence and distant metastasis over time.

[Table/Fig-4]: Comprehensive comparison of Superficial Acral Fibromyxoma (SAF) and its common differential diagnoses [3-5].

Authors and years	Age/ Gender	Duration of swelling	Clinical features	Histopathological examination	Immunohistochemistry	Radiological profile	Earlier misdiagnosis and treatment	Recurrence/ follow-up/ outcome
Riebesell M et al., 2024 [13]	45/M	12 months	Painless periungual swelling of right index finger; clinically resembling glomus tumour.	Myxoid stroma with bland spindle-shaped cells and low mitotic activity.	CD34+, EMA+, CD99+; S100-, desmin-.	MRI: T2-hyperintense, well-circumscribed lesion without bone involvement.	Initially diagnosed as glomus tumour; excised without biopsy.	No recurrence at 6-month follow-up.
Debordes O et al., 2023 [14]	52/F	6 months	Firm palmar nodule misinterpreted as epidermoid cyst.	Spindle cells in fibromyxoid matrix with well-circumscribed borders.	CD34+, CD10+; EMA-, desmin-.	Ultrasound: non specific cystic lesion; no MRI performed.	Treated as epidermoid cyst and excised without IHC.	Recurrence reported at 27 months.
Yang Z et al., 2025 [15]	36/M	4 months	Erythematous rash-like lesion on fingertip initially treated as dermatitis.	Spindle and stellate cells in a myxoid stroma; no atypia or necrosis.	CD34+, CD99-, EMA-, desmin-.	MRI: small subcutaneous nodule without bone invasion.	Initially managed with topical steroids; later excised after growth.	No recurrence at 4-month follow-up.
Gupta A et al., 2023 [16]	60/M	15 years	Firm mass on great toe initially diagnosed as fibroma.	Fibroblastic cells in myxoid to collagenous matrix.	CD34+; CD99 variable, S100-, SMA-.	X-ray: soft tissue mass; no MRI done.	Assumed to be fibroma; delayed diagnosis led to complete excision with DIP joint, disarticulation.	No recurrence at 28 months.
Nagata Y et al., 2025 [17]	29/F	8 months	Painful fingertip swelling with nail deformity, presumed glomus tumour.	Well-circumscribed myxoid neoplasm with bland spindle cells.	CD34+, EMA+, CD68 partial+; S100-, desmin-.	MRI: T2-bright, non invasive lesion near distal phalanx.	Initially diagnosed as glomus tumour; excised without biopsy.	Full recovery; no recurrence reported.
Present case	36/M	10 years	The swelling was firm in consistency, non-tender, and exhibited a smooth surface with thinning and broadening of the overlying nail plate.	A well-circumscribed, globular swelling measuring approximately 2×3 cm	CD34+, S-100-, EMA-	MRI: T2-weighted sequences, located in the volar and periungual region of the distal phalanx	Initially diagnosed of acral lipoma	Surgical excision under regional anaesthesia

[Table/Fig-5]: Detailed comparison of recent case reports of Superficial Acral Fibromyxoma (SAF) of the finger initially misdiagnosed as other benign lesions [13-17].

principles described in the literature, including in the paper by Awasthi R et al., [9].

Radiological imaging plays a supportive role in preoperative evaluation. On MRI, SAF typically appears as a well-defined, homogeneously hyperintense lesion on T2-weighted sequences, confined to the soft tissue without cortical erosion. However, rare instances of bone involvement have been documented in longstanding or neglected lesions is consistent with the findings reported in the study by Tavares C et al., [10].

The principal therapeutic approach for SAF is complete surgical excision with clear histological margins. The National Comprehensive Cancer Network (NCCN) recommends wide local excision for benign soft-tissue tumours to minimise recurrence risk. Mohs micrographic surgery may be considered in anatomically sensitive areas such as the nail bed or volar finger pad to preserve function and achieve margin control [11].

The overall prognosis for SAF is excellent following complete excision is consistent with the findings reported in García-Arpa M et al., [12]. Regular clinical follow-up is advised, particularly in cases with prior incomplete resection. Surveillance aims to detect local recurrence early and ensure timely reintervention. Multiple studies have documented the frequent misdiagnosis of SAF due to its overlapping clinical and histological features with other benign and low-grade acral lesions [Table/Fig-5] [13-17].

CONCLUSION(S)

The SAF, though benign, remains an under-recognised tumour of the distal extremities, often overshadowed by more commonly encountered digital lesions. Its subtle clinical presentation and non specific radiological features necessitate a high-index of suspicion, particularly in long-standing, painless swellings of the fingers or toes. Histopathological confirmation, supported by immunohistochemical profiling, is essential for accurate diagnosis and appropriate management. The present case highlights the importance of including SAF in the differential diagnosis of acral soft-tissue masses, especially in resource-constrained settings where it may be mistaken for more prevalent benign conditions. With timely surgical excision and vigilant follow-up, patients can achieve excellent outcomes with minimal recurrence risk. Enhancing clinical awareness and diagnostic precision is key to improving recognition of this deceptively rare yet clinically significant entity.

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PARTICULARS OF CONTRIBUTORS:
1. Resident, Department of General Surgery, Dr. D. Y. Patil Hospital and Research Centre, Dr. D. Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra, India.
2. Assistant Professor, Department of General Surgery, Dr. D. Y. Patil Hospital and Research Centre, Dr. D. Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra, India.
3. Professor, Department of General Surgery, Dr. D. Y. Patil Hospital and Research Centre, Pimpri, Pune. Dr. D. Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Varun Shetty,
Swarganga Society, B1/704, 7th Floor, Vallabh Nagar, Pimpri, Pune,
Maharashtra, India.
E-mail: shettyvarun03@gmail.com

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