

## HAND &amp; WRIST

# Superficial acral fibromyxoma: insights from case management and comprehensive literature review

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- Superficial acral fibromyxoma, also known as digital fibromyxoma, is a slow-growing, benign, solitary soft tissue tumor. First described in 2001 by Fetsch *et al.*, it is a condition that often occurs in middle-aged individuals. However, it has also been reported across a wide range of ages, ranging from 4 to 86 years, with males more commonly reported. The condition often presents as solitary soft tissue swelling over the periungual or subungual.
- We present the management experience of the rare presentation of this rare tumor and a detailed review of the past literature on this condition. Detailed management of the condition has been described, along with the outcome after 2 years of follow-up and treatment experience.
- Our detailed analysis shows that 2 years is the shortest duration of follow-up to rule out recurrence. Hence, most of the cases reported earlier had given the false sense of the recurrence rate of the tumor, which could lead to undertreatment of the condition.
- The purpose of this article is to allow the readers to understand better the tumor's characteristics with bone involvement and the tumor's diagnostic strategies and treatment options.

Keywords: superficial acral fibromyxoma; digital fibromyxoma; periungual tumor; benign tumor; isolated finger clubbing; bone scalloping

## Introduction

Superficial acral fibromyxoma, also known as digital fibromyxoma, is a slow-growing, benign, solitary soft tissue tumor (1). It was first described in 2001 by Fetsch *et al.* (2). Currently, 326 cases have been reported worldwide since its description. After its initial description, this particular tumor was not classified until 12 years later. It is categorized as a 'Tumor of uncertain differentiation', per the WHO classification of soft tissue

tumors in the 2013 (fourth) edition (3). Although it is more commonly found in middle-aged individuals, it has been observed across a wide range of ages, from 4 to 86 years old. Notably, males are more frequently affected, according to Gupta *et al.* (4). The condition often presents as solitary soft tissue swelling over the periungual or subungual region (2). Here, we describe a case of a rare presentation of acral fibromyxoma with

bone involvement, diagnostic strategies, treatment of the condition, and the literature review. Our case is also the first reported case in Malaysia.

## Case review

A 34-year-old Malay gentleman presented with isolated right ring finger clubbing for 6 years and initially started with swelling over the right ring fingertip with disturbance of normal finger contour, progressively worsening and painless, ignored by the patient as not disturbing daily routine. However, due to the worsening of the finger deformity, the patient seeks medical attention. The patient is otherwise well and can perform his daily routine without difficulties – no other associated constitutional symptoms.

Clinical examination shows an isolated bulbous enlargement with shiny change in the right ring fingernail. Both longitudinal and transverse curvatures of the right ring fingernail plate are disturbed, obliteration of the Lovibond's angle ( $>180^\circ$ ), positive in Curth's modified profile sign, and obliteration of the Schamroth's diamond window. The findings are consistent with grade 5 clubbing (Figs 1, 2 and 3). Otherwise, the sensation and the perfusion of the right ring finger pulp remained intact.

Radiographs of the right ring finger show an increased dorsal tissue shadow with scalloping of the mid and distal part of the distal phalanx without calcification or adjacent periosteal reaction (Figs 4 and 5). MRI shows a localized, solitary homogeneous soft tissue swelling (low T1 and high T2 signals) over the dorsum of the distal phalanx within the subcutaneous tissue level with scalloping of the distal phalanx (Figs 6, 7 and 8).

Excision biopsy was performed with a surgical tourniquet and adequate magnification with the insertion of autologous bone graft. The sterile matrix was preserved for functional and cosmetic purposes. The tumor was gently dissected from the sterile matrix. The tumor could be traced to the insertion site of the terminal tendon sheath and was removed in an *en bloc* manner. Subsequently, the underlying bone was curetted. As for the bony defect, a corticocancellous bone graft was taken from the distal radius and temporarily fixed to the donor region with a k-wire. As the fingernail was severely deformed, a temporary sterile artificial fingernail was reinserted to splint the eponychium fold. Macroscopically, the nodule (2 cm × 1 cm × 0.8 cm) appeared well circumscribed but unencapsulated, soft to firm (Fig. 9), and gelatinous white-grayish cut surface without necrosis or hemorrhage. Microscopically, the tumor comprises spindled and stellate-shaped fibroblasts in loose fascicular and broad storiform growth patterns embedded in a collagenous to a myxoid matrix (Fig. 10). There are mildly accentuated vasculature and many mast cells in the stroma. Mitotic figures are sparse ( $<1$  mitosis per 10 high-power fields)



**Figure 1**

Dorsal profile of the right hand, with isolated clubbing of the right ring finger.

– no multinucleate cells seen. Immunohistochemical studies showed the tumor has moderate-to-strong positivity to CD34 and is negative for S100 (Figs 9, 10 and 11).

Postoperatively, the patient recovered well and returned to work without any disability or recurrence on follow-up by 1 year. However, there was a recurrence at the same site during the 2-year follow-up. The patient was counseled for amputation, considering the bony structure of the distal phalanx was minimal and to eliminate the risk of recurrence, which the patient refused due to the debilitating effect and cultural factors.

## Literature review chart

The summary of the literature is listed in Supplementary Table 1 (see the section on [supplementary materials](#) given at the end of this article).



**Figure 2**  
Medial profile of the right ring finger.

## Discussion

Finger clubbing is a clinical condition whereby there is uniform bulbous swelling of the soft tissue of the terminal phalanx of a digit/toe – also known as drumstick fingers or Hippocratic fingers. A well-known clinical description could be traced back to 400 BC by Hippocrates in an empyema patient. Many causes and etiology of such had been described, primarily systemic causes (ranging from pulmonary, infective, heart, hepatobiliary, gastrointestinal, and endocrine to even congenital) and some rare local causes (intraosseous epidermoid cyst, enchondroma, glomus tumor). The



**Figure 3**  
Lateral profile of the right ring finger.



**Figure 4**  
Posteroanterior radiograph (X-ray, PA view) of lateral three digits of the right hand, showing signs of scalloping of the distal phalanx of the right ring finger.

local single-clubbed finger is a rare condition due to the local proliferation of tissues/bone leading to clubbed features (1, 5).

Superficial acral fibromyxoma or digital fibromyxoma is still a relatively uncommon entity: it was first described in 2001 by Fetsch *et al.* (2), enlisted by WHO under 'Tumors of uncertain differentiation' in 2013 (3). Our literature search shows 326 cases reported worldwide since 1970 (41 retrospective cases reported by the UK from 1970 to 2006). The etiology of superficial acral fibromyxoma or an established predisposing factor is not well established. Though 20% of the reported cases were preceded by trauma, there is no clear association between the two. It is the possibility that the trauma drew the patient's attention to the swelling (6). Clinically, the condition presents as a solitary nodular mass with firm consistency with a higher frequency of occurrence at the finger (125 cases) and toe region (150 cases), mainly occurring in the periungual/subungual locations. However, a small number of cases were reported in the

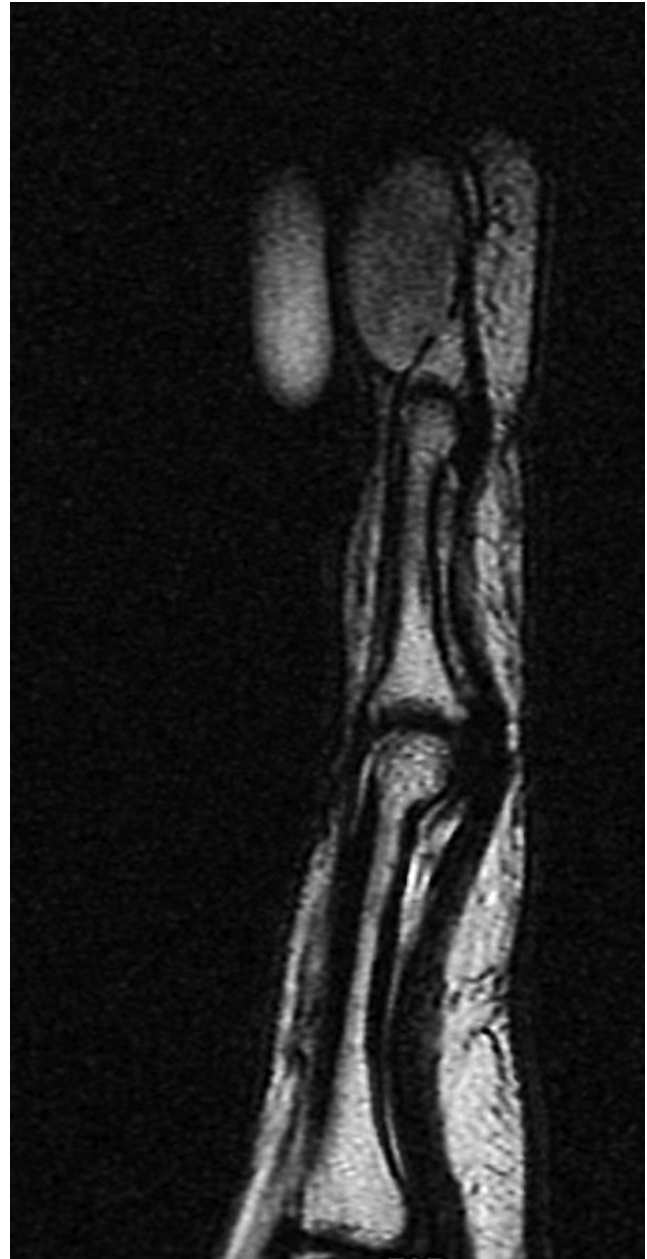


**Figure 5**

Lateral view of the right ring finger radiograph (X-ray), showing increased soft tissue shadow over the dorsum with scalloping of the distal phalanx of the right ring finger.

other areas of the extremities, such as the hand (11 cases), foot (10 cases), heel (5 cases), leg (2 cases), and even thigh (1 case). There is a higher predisposition in middle-aged male patients with an estimated ratio of 2:1 (male:female), which is consistent with our analysis that revealed between the ages of 40 and 49 years had the highest incidence rate and slightly higher male predilection with a ratio of 1.5:1 (male:female). The unique characteristic of superficial acral fibromyxoma is the slow-growing tumor, which appears harmless. Still, it has been previously reported to cause scalloping of the underlying bone and block the small joint's range of motion due to the pressure effect (7).

Demographically, most cases were reported from the Western Hemisphere and Western Europe (UK, USA, Spain, and France). In contrast, only a few isolated cases were reported from Eastern countries (China, Japan, Korea, and India), as shown in Fig. 12. The lower reported incidence from the Eastern countries could be due to the lower incidence in the region

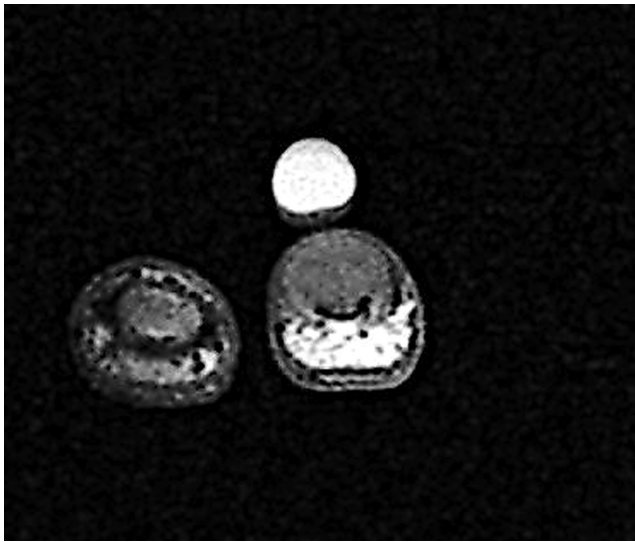


**Figure 6**

Sagittal plane MRI image of the right ring finger, with the tumor exhibiting low signal intensity in T1.

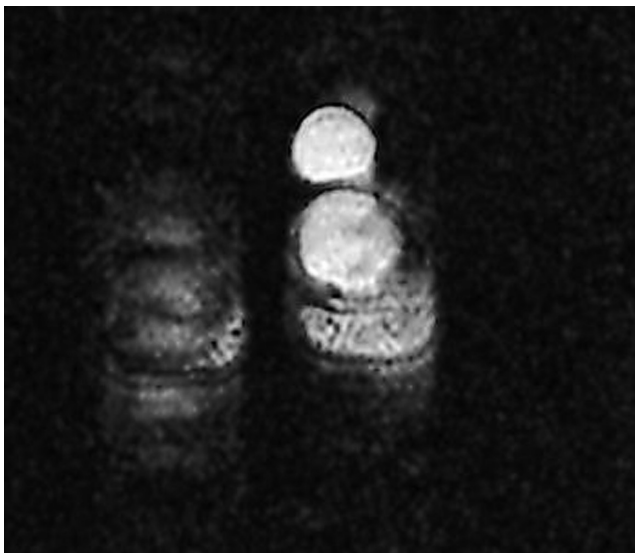
or the possibility of underreporting cases. Further future studies are required to establish the underlying rationale.

Histologically, superficial acral fibromyxoma presented as an unencapsulated tumor with star- or spindle-shaped fibroblasts in broad storiform growth patterns embedded in a myxoid or collagenous matrix. Mast cells and vasculatures are common in variable degrees. Nuclear atypia and mitotic figures are rare and atypical.

**Figure 7**

Axial plane MRI image of the right ring finger, with the tumor exhibiting low signal intensity in T1.

Immunohistochemically, in the majority of reported cases, the tumor cells showed strong and most consistent immunoreactivity to CD34 and negative for S100, actin, calponin, caldesmon, desmin, cytokeratin, and HMB-45, corresponding to our presenting case. Another less-mentioned protein, vimentin, proved to be of assistive value, as it was positive in all the previous literature when tested. The other proteins (EMA, CD99) were inconsistent (8, 9, 10, 11). Malignant transformation has never been reported in this tumor. There are other differential tumors with myxoid

**Figure 8**

Axial plane MRI image of the right ring finger, with the tumor exhibiting high homogeneous signal intensity in T2.

**Figure 9**

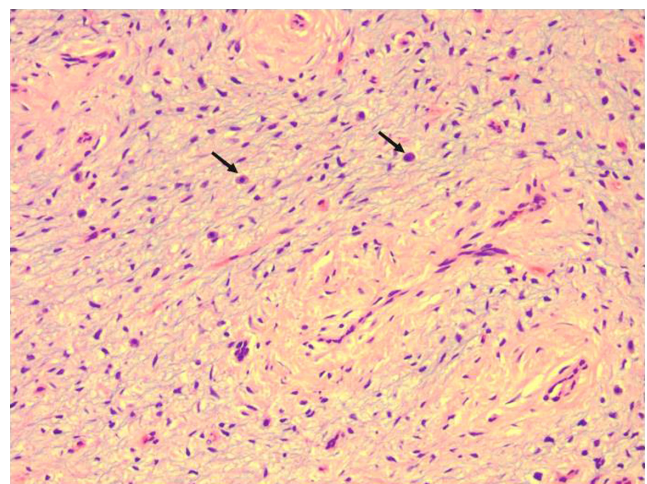
Macroscopic features of the nodule after excision. Measuring 2 cm × 1 cm × 0.8 cm. The nodule appeared well circumscribed, unencapsulated, and soft to firm in consistency.

histological appearance or tumors with distal extremity predilection (Tables 1 and 2).

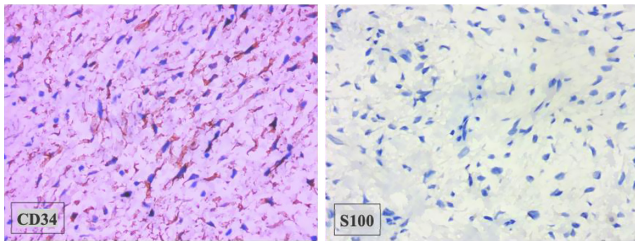
Currently, there are no proper diagnostic criteria for this tumor. We proposed a diagnostic feature based on the review and analysis of all 326 cases reported. From the analysis of all previously reported cases, CD34 is not diagnostic or unique to superficial acral fibromyxoma; it requires a combination of clinical, histological, and immunohistochemical features to diagnose. It is crucial to make an accurate diagnosis to plan for the management and prevent overtreatment (amputation) or undertreatment (risk of recurrence).

## Diagnostic features

Presentation:

**Figure 10**

Photomicrograph of tumor showing spindle and stellate cells in a loose fibromyxoid background, traversed by fine blood vessels. Scattered mast cells are present (arrow). Hematoxyllin and eosin 20×.



**Figure 11**  
 Photomicrograph showing CD34 cytopositivity (brown signals) and S100 negativity of the tumor cells. Immunohistochemistry 40×.

1. Mass/nodule of acral, palm, or foot in the location.
- Radiological investigation:
2. Scalloping of the underlying bone.
  3. Low T1 and moderate-to-high T2 signal under MRI.

**Histology features**

4. Star-/spindle-shaped fibroblasts in a storiform pattern embedded in a myxoid or collagenous matrix.

**Immunohistochemical features**

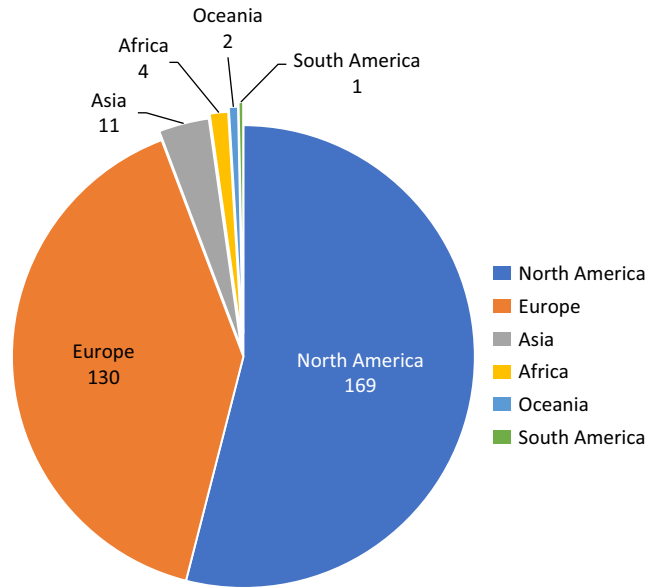
Positive – any one

1. Immunoreactive to vimentin.
2. Immunoreactive to CD34.
3. Immunoreactive to CD99.

Negative

4. Immunohistochemically negative for cytokeratin, smooth muscle, skeletal muscle, and melanocyte markers.

In our case, the excision was performed by experienced hand surgeons under optimal conditions, with adequate magnification and bloodless field, as well as gentle handling and excision of the tumor, yet the tumor recurred. Intraoperatively, it was noted that there were multiple sinuses over the bone surface where the tumor was situated (Fig. 13), which was believed to be the reason for the recurrence in our case. Despite extended curettage being performed using bone burr, recurrence occurred. Hence, based on our experience, a wider resection margin must be considered in the presence of bone scalloping. However, in these acral-prone tumors, the availability of bone stock can be a limiting factor; amputation would be the treatment of choice to achieve a clear margin. In the previous literature, out of eight amputations, none discussed the differences in management. For instance, two cases were reported by Varikatt *et al.* (6) with bone scalloping of the distal phalanx, one subjected to excision and the other to



**Figure 12**  
 Pie chart summarizing the incidence of superficial acral fibromyxoma based on continents.

amputation, while the differences in management and the follow-up were not mentioned in the study. Similarly, Luzar *et al.* (12) reported 1 case of amputation and 13 cases of local excision. Lastly, Fetsche *et al.* (2) reported that 4 out of 37 patients underwent amputation without further description of management specifics. Overall, the management of superficial acral fibromyxoma still needs to be standardized, though local excision was done for most of the cases in the literature (235 out of 250 patients). Based on the literature review and our



**Figure 13**  
 Intraoperative picture showing the tumor (curved arrow) and revealing the dorsum of distal phalanx with multiple sinuses (straight arrows).

**Table 1** Summarized list of tumors with myxoid histological appearances and their features (Source: WHO Classifications of Tumors 2020 (3)).

Tumor	Features
Neurofibromas	S100 positive CD34 negative
Superficial angiomyxoma	Predilection for trunk and head and neck region Inflammatory component usually seen Positive for CD34 Negative for S100, SMA, and muscle specific actin
Myxoid dermatofibrosarcoma protuberans (myxoid variant)	Uncommon With storiform pattern Positive for CD34 Negative for EMA Diagnostic: 17 and 22 translocations with COL1A1–PDGFB fusion gene
Low-grade fibromyxoid sarcoma	Deep-seated tumors Alternating collagenous and myxoid areas with rosette formation CD34 +/-
Digital mucous cyst/mucinous cyst	Ganglion-related lesion Commonly involving the synovium Loose, cystic area Low cellularity
Superficial angiomyxoma	Head and neck region Distinctive lobulated pattern Inflammatory cells are seen
Low-grade myxofibrosarcoma	Seen in extremities Curvilinear capillary pattern with myeloid matrix Perivascular condensation of tumor cells Marked nuclear atypia Variable for CD34 immunoreactivity EMA – negative

experience, marginal excision could be done in these acral-prone benign tumors. However, in the event of bone involvement/bone scalloping, depending on the extent of the tumor, extended excision with a clear bone margin or amputation is advocated for disease clearance.

The recurrence rate in the literature could be underreported due to the inconsistency and inadequacy in the follow-up duration. Based on our literature review, almost a third of the case reports/case series did not comment on follow-up/recurrence (19 out of 51 case reports/case series), 16 reported up to 1-year postoperative period, 9 reported until 2 years, and 4 reported beyond 2 years. For instance, Cogrel *et al.* (13) reported three cases with different follow-up periods for each subject (9, 12, and 22 months), and

the case series of two patients by Kazakov *et al.* (14) reported on the follow-up specifics in one case but not in the other. Hence, the actual recurrence rate could not be determined. Superficial acral fibromyxoma is a benign, slow-growing tumor that would take longer for the recurrence to occur. Therefore, a more extended follow-up period will be required to exclude early recurrences. In one of the few large case series reported by Fetsch *et al.* (2), two recurrences were revealed in their study, which occurred at 2 years 10 months and the second case at 10 years of follow-up. Similarly, Hollman *et al.* (15) reported recurrence occurred at a mean interval of 27 months in 10 patients, where the recurrence period is consistent with our study (at 2 years of follow-up). Hence, it is inevitable that in more than half of the case reports/case series where follow-up for less than 2 years, the recurrence

**Table 2** List of soft tissue tumors with distal extremity predilection and their features (Source: WHO Classifications of Tumors (3)).

Tumor	Features
Sclerosing perineurioma	Cell arranged in onion-skin pattern CD34 +/-, usually negative EMA positive
Acquired digital fibrokeratomas	Exophytic, paucicellular, with interwoven thick vertical-oriented collagen bundles CD34 +/- (not consistent) EMA - negative S100 - negative
Periungual fibroma	Multiple in nature Associated with tuberous sclerosis Abundant fibrous connective tissue Vertical-oriented collagen bundles
Acral myxoinflammatory fibroblastic sarcoma	Virocyte or Reed–Sternberg-like cells with hyaline areas Abundant inflammatory cells EMA - negative

status is considered unknown (38 out of 51 case reports/case series). Based on our literature analysis, all the recurrences reported were after 2 years of follow-up, with a mean of 26.5 months. The earliest recurrence was reported at 20 months post excision by Goo *et al.* (16), the only case where the recurrence was reported before 2 years of follow-up. Our literature review has covered the most significant number of reported cases. In contrast, the recurrence of the tumor was previously reported to be 10–20%; our analysis revealed an exact recurrence rate of 13% (22/164 of all reported cases) or 16% (22/135) where the analysis was taken from the reported cases follow-up beyond 2 years, including our case. However, the exact recurrence rate of the condition with bone involvement could not be analyzed separately.

### Limitation

Analyzing the recurrence rate of superficial acral fibromyxoma with bone involvement remains challenging as it is a rare and uncommon tumor. A future study on the long-term follow-up of the condition with bone involvement is recommended.

### Conclusion

In conclusion, we presented and summarized a poorly recognized entity with unique combinations of clinical and histopathological features. Superficial acral fibromyxoma is still uncommon to us. Due to the benign nature of the tumor, it would be commonly missed and ignored by the patient as it rarely disturbs the daily function. However, delayed cases could lead to

morbidity and loss of function of the affected joint/bone. In the presence of bone scalloping, a wider margin of resection or amputation will be warranted to avoid recurrence. Hence, this paper aims to introduce and assist in the management of future cases. Moreover, a more extended follow-up review is recommended.

#### Supplementary materials

This is linked to the online version of the paper at <https://doi.org/10.1530/EOR-23-0151>.

#### ICMJE Conflict of Interest Statement

The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of the case study reported.

#### Funding Statement

This work did not receive any specific grant from any funding agency in the public, commercial, or not-for-profit sector.

#### Patient perspective

The patient had consented to publish the article without the patient's identification for academic purposes and to assist in the future management of the uncommon condition. The patient is currently well with full hand function despite the recurrence. The patient is not keen on further surgery as the patient still has the full function of the hand and will consider further surgery later.

#### Acknowledgements

We are grateful to the NOCERAL at the University of Malaya for providing us with the resources and support we needed to complete this project. We would also like to thank our colleagues at the University of Malaya for their feedback and support throughout the research process.



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