Journal of Chemical Health Risks

www.jchr.org

JCHR (2023) 13(4s), 173-177 | ISSN:2251-6727

A Case of Superficial Acral Fibromyxoma

Dr. Bhushan Shah[1], Dr. Chaitanya Mahajan[2]*, Dr. Adithya Reddy Vijendra[3], Dr. Jayant Bajaj[4], Dr. Devendra S Yadav[5], Dr. Ritika Gupta[6]

[1]Professor and Head of Unit, Department of General Surgery, Dr. D.Y. Patil Medical College, Hospital and Research Centre, D.Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra-411018, Email: bhushan.shah@dpu.edu.in Orcid id-0000-0002-7709-2455

[2]*Resident, Department of General Surgery, Department of General Surgery Dr. D.Y. Patil Medical College, Hospital and Research Centre, D.Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra-411018,

[3]Resident, Department of General Surgery, Dr. D.Y. Patil Medical College, Hospital and Research Centre, D.Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra-411018.

[4]Resident, Department of General Surgery, Dr. D.Y. Patil Medical College, Hospital and Research Centre, D.Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra-411018.

[5]Resident, Department of General Surgery, Dr. D.Y. Patil Medical College, Hospital and Research Centre, D.Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra-411018.

[6]Resident, Department of General Surgery, Address: Dr. D.Y. Patil Medical College, Hospital and Research Centre, D.Y. Patil Vidyapeeth, Pimpri, Pune, Maharashtra-411018.

KEYWORDS

Superficial acral fibromyxoma, Soft tissue tumor, recurrence, mesenchymal cells, surgical excision, periungual.

ABSTRACT:

A rare benign tumor that mainly affects the distal extremities, such as the fingers and toes, is known as superficial acral fibromyxoma (SAFM). This distinct soft tissue tumor was first described in 2001 by Fetsch et al. and is characterized by slow-growing, painless nodules or tumors with a firm, rubbery texture and bluish appearance.

SAFM commonly involves the nail bed or periungual region, potentially leading to nail plate deformities. The exact etiology of SAFM remains unclear, although it is believed to originate from dermal dendritic cells. Histopathological examination and immunohistochemistry aid in confirming the diagnosis. Surgical excision with clear margins is the primary treatment, and long-term follow-up is advised to monitor for recurrence.

INTRODUCTION

The rare soft tissue tumour known as superficial acral fibromyxoma (SAF) was first recognised as a separate histopathologic entity in 2001 by Fetsch et al. It is also known as digital fibromyxoma and is derived from mesenchymal tissue. SAF typically occurs on the hands and feet, particularly on the fingers and toes. This tumor is characterized by its slow-growing nature and benign behavior. While it may cause local discomfort or pain, it is generally not associated with any significant health risks. However, due to its rarity and unique histopathological features, SAF can often be misdiagnosed or overlooked, leading to delayed treatment or unnecessary procedures.

The exact cause of superficial acral fibromyxoma is still unknown, but it is believed to be aresult of genetic mutations or alterations in the mesenchymal cells. SAF predominantly affects adults, with a slight male predominance. The tumor typically presents as a small, painless nodule that gradually increases in size over time. The color of the lesion can vary from flesh-colored to pink or brown. It may also have a gelatinous appearance due to the presence of myxoid material within the tumor.

Diagnosing superficial acral fibromyxoma can be challenging, as it shares similarities with other soft tissue tumors and lesions, such as dermatofibroma and ganglion cysts. A thorough clinical examination, combined with imaging techniques like ultrasound or magnetic resonance imaging (MRI), can help in establishing a definitive diagnosis. Histopathological analysis of a biopsy sample remains the gold standard for confirming SAF, as it reveals characteristic features such as spindled cells, myxoid stroma, and absence of mitotic activity.

Histopathological examination of SAF reveals a characteristic pattern of spindle-shaped cells embedded in a myxoid stroma. The tumor cells show low cellularity and minimal mitotic activity. Immunohistochemical staining can be used to confirm the diagnosis, with positive results for vimentin and negativity for S-100 protein and CD34.

Treatment options for superficial acral fibromyxoma

Journal of Chemical Health Risks
www.jchr.org
JCHR (2023) 13(4s), 173-177 | ISSN:2251-6727

include surgical excision and Mohs micrographic surgery. Complete excision of the tumor is often curative, and recurrence rates are generally low. However, due to the potential for misdiagnosis or incomplete excision, regular follow-up examinations are necessary to monitor for any signs of recurrence or metastasis.

CASE REPORT

A 63-year-old female patient reported to the department of general surgery with a chief complaint of swelling in her right little finger. The swelling had been present for 15 years, with an insidious onset and a gradual increase in size over the last 10–12 years. The patient reported no associated pain, trauma, or discharge. She denied any difficulty with finger movement. She had no known history of diabetes mellitus, hypertension, tuberculosis, or bronchial asthma. Her past medical and family history was unremarkable.

On general examination, the patient was conscious, cooperative, and well-oriented to time, place, and person. She was well-built and nourished. Vital signs were within normal limits, with no signs of pallor, icterus, clubbing, cyanosis, lymphadenopathy, or oedema.

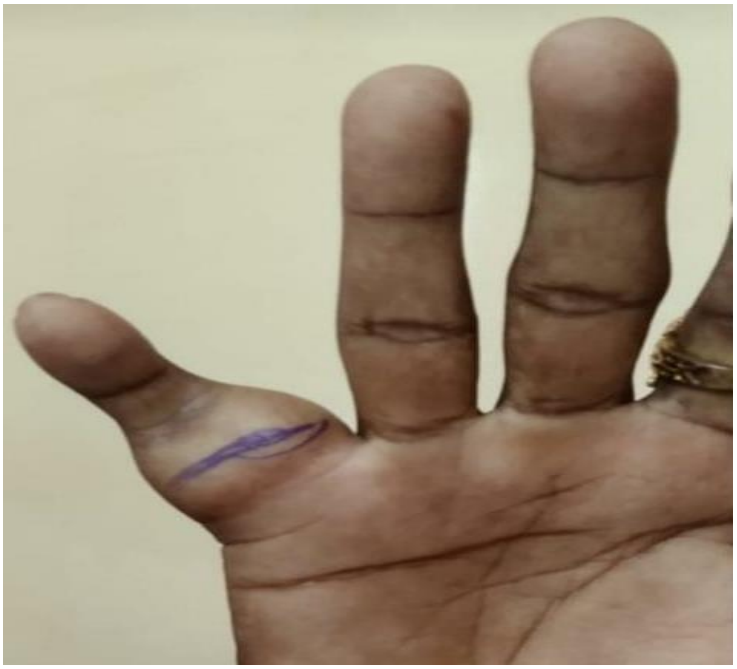

Figure 1. Swelling seen around little finger of right hand. (Pre-operative.)

Local examination of the right hand revealed a swelling at the middle finger tip, measuring approximately 3x4 cm. [figure 1] The swelling had a globular shape with thinning and broadening of the nail. The surface appeared smooth, and the skin over the swelling was normal. Finger movements were normal, and there was no redness. The rest of the right hand appeared normal on inspection. Palpation of the swelling revealed a firm to hard consistency.

The temperature was normal, and the swelling involved the right third distal phalanx. Systemic examination, including cardiovascular, respiratory, and central nervous system assessments, yielded unremarkable findings.

BIOCHEMISTRY LAB REPORTS

The patient's laboratory reports were within normal limits, indicating no significant abnormalities in the assessed parameters. The complete blood count (CBC) showed a hemoglobin (Hb) level of 12.5 g/dL, a total leukocyte count (TLC) of 6300 cells/mm³, and a platelet count of 3,08,000 cells/mm³. Liver function tests (LFT) revealed total bilirubin (TB) levels of 0.60 mg/dL, conjugated bilirubin (C) levels of 0.34 mg/dL, unconjugated bilirubin (UC) levels of 0.26 mg/dL, serum glutamic oxaloacetic transaminase (SGOT) levels of 14 IU/L, serum glutamic pyruvic transaminase (SGPT) levels of 40 IU/L, and alkaline phosphatase (ALP) levels of 22 IU/L. Renal function tests (RFT) displayed a urea level of 22 mg/dL and a creatinine level of 0.67 mg/dL. The levels of electrolytes and proteins in the serum were within normal limits. The prothrombin time-international normalized ratio (PT-INR) and random blood sugar levels were also found to be within normal range. These findings from the biochemistry laboratory reports suggest no significant

Journal of Chemical Health Risks
www.jchr.org
JCHR (2023) 13(4s), 173-177 | ISSN:2251-6727

abnormalities in the patient's blood parameters, liver function, renal function, electrolytes, coagulation profile, andblood sugar levels.

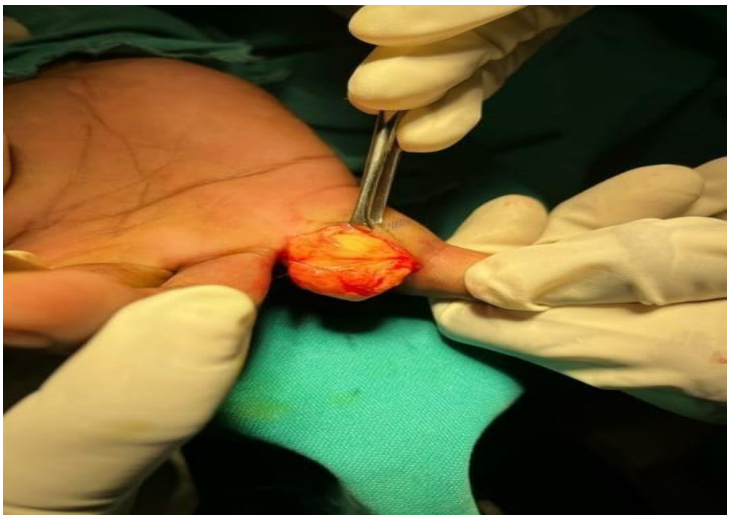

Figure 2. Surgical removal of SAFM. (Intra operative).

MANAGEMENT

The surgical excision was performed under regional anesthesia under appropriate aseptic conditions (Figure 2). Excised tissue sent for histopathological examination. (Figure 3) Antibiotics and analgesic drugs were prescribed. Proper wound care was also taken. Regular follow-up was done. After 15 days, satisfactory wound healing was seen. (Figure 4)

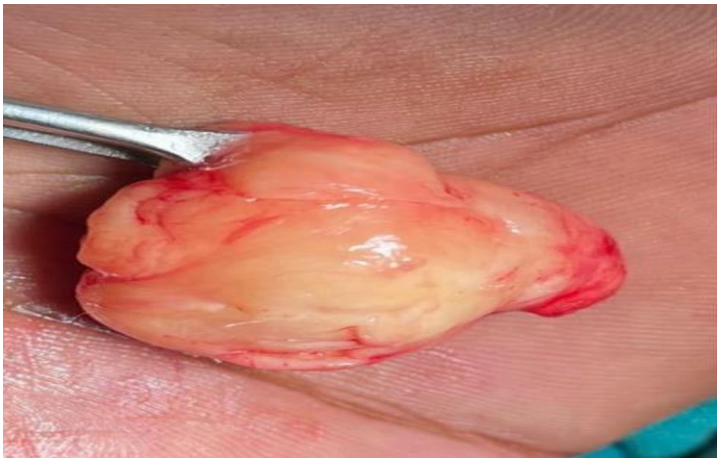

Figure 3. Surgically excised specimen (Post-Operative.)

Journal of Chemical Health Risks
www.jchr.org
JCHR (2023) 13(4s), 173-177 | ISSN:2251-6727

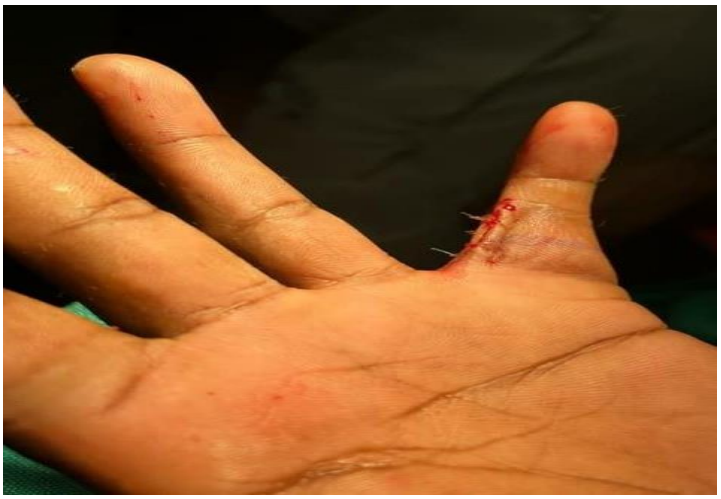

Figure 4. Closure of excised area after surgical removal of lesion.

DISCUSSION

Superficial acral fibromyxoma (SAFM) is a rare and slow-growing soft tissue tumor that predominantly affects the periungual and subungual regions of the fingers and toes in adults. This distinctive tumor was first described as a separate entity in 2001 by Fetsch et al. With slight male predominance, SAFM is a relatively uncommon condition, with fewer than 50 cases reported worldwide. Although it is considered a benign tumor, SAFM has a tendency to recur if inadequately excised. Therefore, accurate diagnosis, complete surgical excision with tumor-free margins, and regular follow-up are crucial for the management of this condition.[1]

Superficial acral fibromyxoma typically presents as a solitary, slow-growing mass in the periungual and subungual regions of the fingers and toes. It commonly occurs in middle-aged adults, with a slight male predominance. Prior to resection, the lesion may exist for a few months to several years. Patients may experience pain or discomfort when applying pressure to the affected area. Rarely reported and typically absent from the clinical history is antecedent trauma.[2]

Physical examination reveals a solitary nodule, which may manifest as an exophytic, dome shaped, polypoid, or verruciform lesion. In some cases, ulceration may be observed. The location of the tumor is primarily in the digits, with a high prevalence near the nail unit. However, rare cases have been reported on the ankle and leg.

-To establish a diagnosis of SAFM, histopathological examination is essential. Microscopic analysis of the lesion reveals a moderately cellular proliferation of stellate and spindle-shaped fibroblast-like cells within a myxocollagenous stroma. Mast cells are commonly found throughout the tumor. Multinucleated stromal cells can be present, however nuclear atypia and mitotic figures are rare. Immunohistochemical studies can aid in confirming the diagnosis, with positive reactivity for CD34, epithelial membrane antigen (EMA), and CD99.[3]

When encountering a superficial acral fibromyxoma, it is important to consider other differential diagnoses to ensure accurate classification. Neurofibroma, characterized by a fibromyxoid stroma, may be a potential differential diagnosis. However, immune histochemistry staining for S100 can help differentiate between the two tumors, as neurofibromas typically exhibit positive staining for S100.

Sclerosing perineurioma, which predominantly involves acral sites, may also be considered in the differential diagnosis. These lesions often lack myxoid changes and can be distinguished from SAFM by their negative staining for S100 and positive staining for EMA. Additional markers such as Glut-1 positivity and CD34 negativity favor the diagnosis of perineurioma.[4]

The treatment of superficial acral fibromyxoma involves complete surgical excision with clear margins. In cases of inadequate excision, the tumor may recur locally. Local recurrence is almost always linked to initial biopsy or subsequent excision margins that were positive.

However, malignant behavior or metastasis has not been reported in SAFM cases.[5] Regular follow-up is essential for patients with SAFM due to the potential for recurrence.

Long-term monitoring can help detect any local recurrence and ensure timely intervention if necessary. Although SAFM is a rare entity, it is important for clinicians to be aware of its existence, particularly in cases of recurring lesions in the periungual and subungual regions.

A rare and distinctive soft tissue tumor that mostly affects the periungual and subungual regions of the

Journal of Chemical Health Risks
www.jchr.org
JCHR (2023) 13(4s), 173-177 | ISSN:2251-6727

fingers and toes is called a superficial acral fibromyxoma.[6] While it is a benign condition, SAFM has a propensity for local recurrence if not adequately excised. Accurate diagnosis, complete surgical excision with tumor-free margins, and regular follow-up are essential for the management of this condition. By staying vigilant and knowledgeable about this rare tumor, clinicians can provide timely and appropriate care to patients with superficial acral fibromyxoma.[7]

SAF does not seem to be widely recognized. It was not described in the World Health Organization blue book of skin tumors but was included in the Armed Forces Institute of Pathology (AFIP) atlas of nonmelanocytic tumors. Also, similar tumors like this in the nail bed of fingers and toes have been reported as a cellular digital fibromas.[8]

A handful of case reports regarding SAFM have been published in the radiology literature also, but to our knowledge, only few studies has been documented with radiographic findings associated with the tumor. In 2008 Varikatt et al reported two cases of SAFM with erosion of underlying cortical bone of the distal phalanx [9]

Although rare, surgeons should be always aware of this condition, its presentation and its benign clinical features .[10]

In the majority of the literature, there have been no cases of cancer reported.

Conclusion

Superficial acral fibromyxoma is a single soft-tissue tumor with a high predilection for the subungual and periungual acral regions. It gives a scientific image and histopathological traits like different dermatological conditions, which makes its very last analysis difficult.

The strategies defined in a few literature that aid the diagnostic definition are scientific examination, imaging tests, histopathological evaluations, and immuno histochemical studies.

Currently, the only option for treating SAF is complete excision, with a high likelihood of recurrence and an undetermined likelihood of malignancy. To treat the tumors and prevent malignancy transformation and recurrence, the entire tumor must be surgically removed. Although there is no evidence of aggressive behaviour in the tumor, the possibility of malignant transformation is unclear because cytologic atypia is present in a small number of cases.

Declaration of competing interest:

There is no conflict of interest.

Author contributions:

All the authors had contributed in collecting data, preparing and proofreading the manuscript.

Ethical statement:

Informed consent was obtained from the patient included in the study. There is no information (names, initials, hospital identification numbers, or photographs) in the submitted manuscript that can be used to identify patients. All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national)

REFERENCES

1. Chiheb S, Mouradi M, Hali F. Superficial acral fibromyxoma: clinicopathologic analysis of five cases. Skin Appendage Disorders. 2021 Nov 2;7(6):468-74.
2. Sawaya JL, Khachemoune A. Superficial acral fibromyxoma. International Journal of Dermatology.2015 May;54(5):499-508.
3. Fetsch JF, Laskin WB, Miettinen M. Superficial acral fibromyxoma: a clinicopathologic and immunohistochemical analysis of 37 cases of a distinctive soft tissue tumor with a predilection for the fingers and toes. Human pathology. 2001 Jul 1;32(7):704-14.
4. Tardío JC. THIS ARTICLE HAS BEEN RETRACTED CD34-reactive tumors of the skin. An updated review of an ever-growing list of lesions. Journal of cutaneous pathology. 2008 Dec;35(12):1079-92.
5. Ashby-Richardson H, Rogers GS, Stadecker MJ. Superficial acral fibromyxoma: an overview. Archives of pathology & laboratory medicine. 2011 Aug 1;135(8):1064-6.
6. Crepaldi BE, Soares RD, Silveira FD, Taira RI, Hirakawa CK, Matsumoto MH. Superficial acral fibromyxoma: literature review. Revista Brasileira de Ortopedia. 2019 Nov 14;54: 491-6.
7. Perrin C. Tumors of the nail unit. A review. Part II: acquired localized longitudinal pachyonychia and masked nail tumors. The American Journal of Dermatopathology. 2013 Oct 1;35(7):693-712.
8. Al-Daraji WI, Miettinen M. Superficial acral fibromyxoma: a clinicopathological analysis of 32 tumors including 4 in the heel. Journal of cutaneous pathology. 2008 Nov;35(11):1020-6.
9. Bindra J, Doherty M, Hunter JC. Superficial acral fibromyxoma. Radiology Case Reports. 2012 Jan 1;7(3):751.
10. Quaba O, Evans A, Al-Nafussi AA, Nassan A. Superficial acral fibromyxoma. British journal of plasticsurgery. 2005 Jun 1;58(4):561-4.