ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research

Vol. 17, Issue, 01, pp.31568-31570, January, 2025
DOI: https://doi.org/10.24941/ijcr.48256.01.2025

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

RESEARCH ARTICLE

CHALLENGING DIAGNOSIS OF SUPERFICIAL ACRAL FIBROMYXOMA: CELLULAR ATYPIA AND UNUSUAL IMAGING FINDINGS

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ARTICLE INFO

Article History:
Received 20th October, 2024
Received in revised form
17th November, 2024
Accepted 24th December, 2024
Published online 31st January, 2025

Key Words:

Superficial Acral Fibromyxoma, Cellular Atypia, BoneLysis, Subungual Tumor.

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ABSTRACT

Introduction: Superficial acralfibromyxoma (SAF) is a rare benign soft tissue tumor typically occurring in acral regions, particularly the peri- and subungual areas. Submatrix localization is exceedingly uncommon and poses diagnostic and therapeutic challenges. **Case Presentation:** We report the case of a 21-year-old female presenting with a painful, progressively enlarging swelling of the left fourth toe over three years. Clinical examination revealed a subungual mass measuring 2.5 cm, firm and minimally mobile, with nail widening and pseudo-clubbing. Radiological findings demonstrated a well-defined lytic lesion in the distal phalanx, suggestive of a mucous or epidermoid cyst. Histopathological and immunohistochemical evaluation confirmed the diagnosis of SAF with cellular atypia and moderate mitotic activity. Given the rapid progression and significant bone involvement, a trans-phalangeal amputation was performed. **Discussion:** SAF typically exhibits slow growth and a benign course. However, in cases with atypical histological features and extensive bone involvement, as presented here, aggressive surgical management may be necessary to prevent recurrence. This case also highlights the importance of including SAF in the differential diagnosis of acral lesions with lytic bone involvement. **Conclusion:** This case underscores the rarity of SAF with submatrix localization and cellular atypia. Early diagnosis and adequate surgical margins are crucial to achieving favorable outcomes while minimizing recurrence risk.

Citation: Bachkira, E., Er-rejrragi, A., Ghannam, A., Bakdid, A., Bouhouche, M., Rajaallah, A., EL Kassimi, C., Rafaoui, A., Messoudi, A. et al. 2025. "Challenging Diagnosis of Superficial Acral Fibromyxoma: Cellular Atypia and Unusual Imaging Findings.*". International Journal of Current Research*, 17, (01), 31568-31570.

INTRODUCTION

Superficial AcralFibromyxoma (SAF) is a benign soft tissue tumor typically located in acral regions, often peri- or subungual. Submatrix localization is exceedingly rare. This relatively new entity was first described in 2001 by Fetsch et al. (1). SAF typically presents as a solitary, firm nodule that is painful in nearly half of the cases and exhibits slow growth. It is primarily found at the distal extremities of the fingers and toes, with a particular predilection for the nail region, especially the nail bed and lateral folds (2). We present the clinical, radiological, and histological features of a rare case of SAF with exclusive submatrix localization.

CASE PRESENTATION

A 21-year-old female patient, with no significant medical history, presented with a painful swelling of the left fourth toe that had been evolving over three years and had shown rapid growth recently. Clinical examination revealed a subungual mass overlying the distal phalanx of the fourth toe, measuring 2.5 cm in its longest axis.

The mass was firm, minimally mobile, and associated with nail widening, creating a pseudo-clubbing appearance [Fig. 1]. Imaging studies were performed. Standard foot radiography revealed a well-defined, rounded lytic lesion in the distal phalanx [Fig. 2]. Computed tomography (CT) of the foot identified a subungual cystic lesion in the fourth toe, suggesting the appearance of a mucous cyst [Fig. 3]. Magnetic resonance imaging (MRI) demonstrated a cystic formation, raising suspicion for an epidermoid cyst. [Fig. 4].

A biopsy was subsequently performed, revealing a poorly differentiated myxoid tumor proliferation with cellular atypia. Immunohistochemical analysis showed exclusive expression of CD99, with no staining for PS100 or EMA, findings consistent with superficial acralfibromyxoma exhibiting cellular atypia and moderate mitotic activity Given the rapid progression of the mass, the presence of cellular atypia, and significant bone lysis, we performed a trans-phalangeal amputation to prevent recurrence. [Fig. 5]. Histopathological examination of the resected specimen confirmed the initial diagnosis. After three year of follow-up, the patient showed no signs of recurrence.

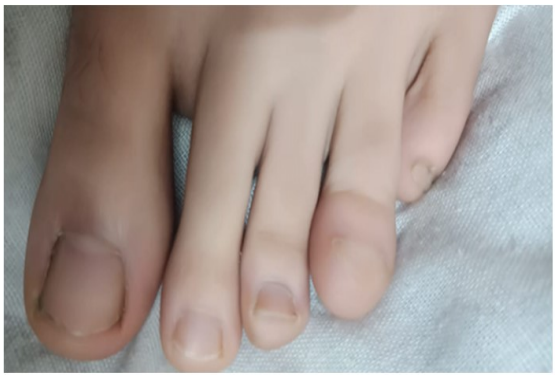

Fig. 1. Clinical image showing pseudo-clubbing of the fourth toe

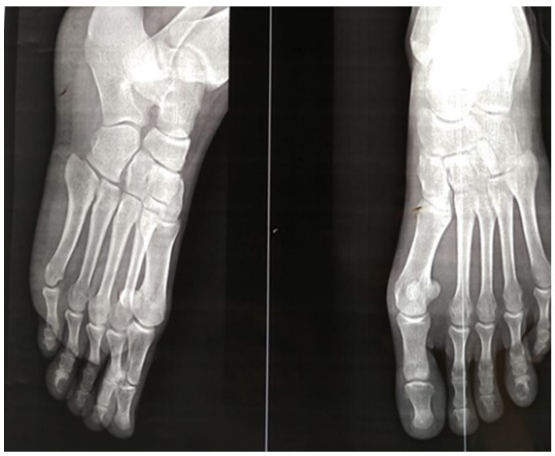

Fig. 2. X-ray of the left foot showing a lytic and erosive lesion of the distal phalanx

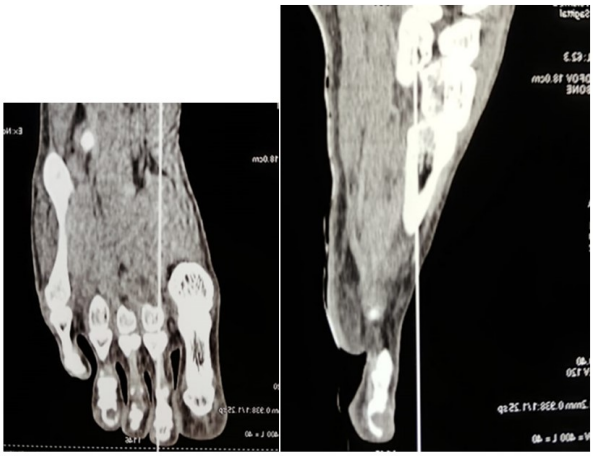

Fig. 3. CT scan of the foot showing a cystic lesion with bone lysis

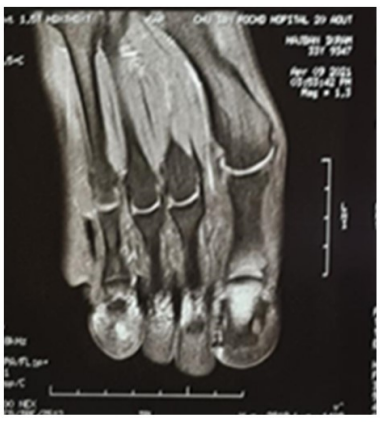

Fig. 4. MRI of the foot showing a cystic formation

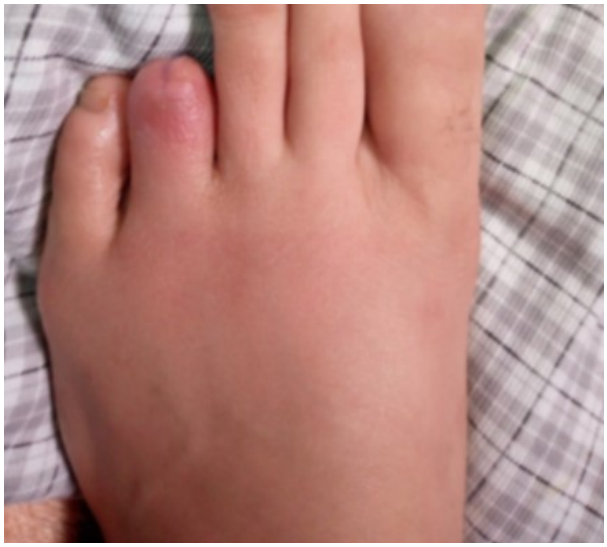

Fig. 5. Clinical image in the postoperative period

DISCUSSION

SAF typically affects the subungual region, with a predilection for the great toe, although the ventral surface of the fingers, the heel, the palm, and the ankle can also be involved. It is most frequently observed in young adults (mean age: 46 years) and exhibits a male predominance (male-to-female ratio: 2:1)(3). This is a non-encapsulated tumor, often delineated by a border referred to as the "green zone"(4), which separates the epidermis from the tumor. It is characterized by a proliferation of spindle-shaped or stellate cells arranged in various orientations, sometimes forming a storiform or fascicular architecture. The cells are embedded within a myxoid or myxocollagenous matrix, which is often highly vascularized and rich in mast cells (5). Mild to moderate nuclear atypia is sometimes observed, with no clear prognostic significance(1). Most SAF cases are located in the dermis or hypodermis, rarely involving the fascia or underlying bone(1,2). Radiographic imaging may reveal underlying bone erosion caused by the tumor's mass effect (3).

In reported series, most excisions have involved non-clear margins, yet only 24% of tumors recur, sometimes after several years.(2). The main clinical differential diagnoses for SAF include lipoma and schwannoma (5). Submatrix localization may suggest neurofibroma or pseudo-mucoid cyst (1). Chondromyxoid fibroma is a low-grade tumor that may exhibit nuclear atypia on histological examination, mimicking chondrosarcoma. Differentiating between these two entities is crucial to avoid overdiagnosis and overly aggressive treatment (6). Recurrence is common after marginal excision, particularly in young patients, as observed in our case. Complete resection remains the cornerstone of management. A conservative approach, with complete excision whenever possible, is recommended(2), However, amputation may be the treatment of choice to prevent recurrence. In a series of 37 SAF patients, Fetsch reported two cases with a significantly higher number of focal atypia and a low mitotic rate, suggesting that these cases might evolve into low-grade malignant neoplasms, although no evidence of aggressive progression was observed in these tumors(1). minimize the risk of recurrence, which remains low, wide margins at the resection site are advised

CONCLUSION

Superficial acral fibromyxoma is a rare and benign tumor predominantly found in acral regions, often with a favorable prognosis when managed appropriately. However, cases with cellular atypia and significant bone involvement, such as the one presented, highlight the need for a tailored therapeutic approach. Adequate surgical excision with clear margins remains the cornerstone of treatment to minimize recurrence risk. This case underscores the importance of considering SAF in the differential diagnosis of subungual and acral lesions, particularly when atypical clinical and radiological features are present. Long-term follow-up is essential to monitor for potential recurrence or unexpected progression.

Ethical approval: This case series got ethical approval from our institution. The patient was given consent form before the surgery.

Conflict of Interest: The authors declare no conflicts of interest.

Sources of funding: None.

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REFERENCES

1. Fetsch JF, Laskin WB, Miettinen M. Superficial acralfibromyxoma: a clinicopathologic and immunohistochemical analysis of 37 cases of a distinctive soft tissue tumor with a predilection for the fingers and toes. Hum Pathol. juill 2001;32(7):704-14.
2. Hollmann TJ, Bovée JVMG, Fletcher CDM. Digital fibromyxoma (superficial acralfibromyxoma): a detailed characterization of 124 cases. Am J SurgPathol. juin 2012;36(6):789-98.
3. Nayak C R., C, Swagata T. Superficial AcralFibromyxoma. Indian J Dermatol. 1 juill 2016;61:457.
4. Chabbab F, Metz T, Saez Beltran L, Theunis A, Richert B. Fibromyxome acral superficiel de localisation sous-matricielle : une forme clinique inhabituelle. Ann Dermatol Vénéréologie. 1 févr 2014;141(2):94-105.
5. Tan HL, Ahmad TS, Kumar CS, Adnan YK, Looi LM, Gunasagaran J. Superficial acralfibromyxoma: insights from case management and comprehensive literature review. 1 févr 2024 [cité 1 déc 2024]; Disponible sur: https://eor.bioscientifica.com/view/journals/eor/9/2/EOR-23-0151.xml
6. Riebesell SA, Lazaro JS, Kirby D, Rivlin M. A Case of Superficial Acral Fibromyxoma of the Index Finger. Cureus. mai 2024;16(5):e60518.