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Case Report

Superficial Acral Fibromixoma-A Rare Entity, A Novel Surgical Aproach

Joao Carvalho Pereira[1]*, Andre Moreira[1], Tiago Barbosa[1], Joana Azevedo[1], Aurea Canelhas[2], Elisabete Ribeiro[1,2], Pedro Varanda[1] and Luis Filipe Rodrigues[1,2]

[1]Department of Orthopaedic and Traumatology, Hospital de Braga, Braga, Portugal

[2] Hospitais Trofa Saude

***Corresponding Author:** Joao Carvalho Pereira, Department of Orthopaedics and Traumatology, Hospital de Braga, Braga, Portugal.

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Abstract

Superficial Acral Fibromyxoma (SAFM) is a rare and benign soft tissue neoplasia that most often affects the digits, particularly the nail apparatus. The pathology presents as a slow-growing mass associated with xanthonychia, nail thickening, and proximal paronychia, often leading to misdiagnosis with fungal infection. The mainstay of treatment is en-bloc resection, with a non-negligible recurrence rate. This case report describes a 48-year-old female with a 2-year history of right third fingernail discoloration and thickening. After unsuccessful treatment with anti-fungal agents, longitudinal partial proximal plate nail avulsion was conducted, revealing a consistent, gelatinous and well-demarcated flesh-coloured lesion. Histopathological examination confirmed SAFM, with CD34 positivity and negativity for pS-100 immunomarkers. After 12 months of follow up, the patient was completely asymptomatic, with no signs of lesion recurrence or nail dystrophia. Minimally damaging techniques as the one described in this case report should be preferred in order to preserve the maximum amount of nail matrix and provide the best clinical results for the patient. Awareness of this rare entity and its characteristic presentation is crucial for accurate diagnosis and timely surgical management.

Keywords: Superficial Acral Fibromixoma, Benign Neoplasms, Surgical Technique

Introduction

Superficial Acral Fibromyxoma (SAFM), first described in 2001 by Fetsch., *et al.* [1], is a rare and benign mesenchymal neoplasia that typically affects the upper and lower extremities [1-3]. Subungual presentation is common and constitutes a differential diagnosis with other benign subungual pathology [2,4]. The mainstay of treatment is *en-bloc* resection, with a non-negligible recurrence rate [1,3,4].

Case Description

A 48 years old Caucasian female, with no relevant medical history or associated traumatic events presented with a 2-year history of right third fingernail discoloration and thickening, with no associated pain. Due to onychomycosis misdiagnosis, several treatment protocols with anti-fungal agents were unsuccessfully performed.

Upon physical exam, a longitudinal lesion with well-defined borders, occupying the middle third of the nail was identified. The lesion presented with xantonychia, proximal nail thickening, *lúnula* bulging and proximal paronychia (Figure 1).

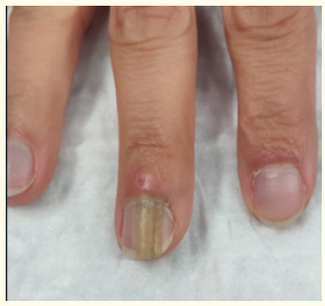

Figure 1: Initial clinical presentation - xanthonychia, proximal nail thickening and proximal paronychia.

Ultrasound exam showed an avascular hypoechoic subungual lesion, with several internal echoic foci (Figure 2).

In MRI T1 fat-saturated sequence, a contrast-enhanced lesion was visible. In T2 sequence a hyperintense signal was also described (Figure 3).

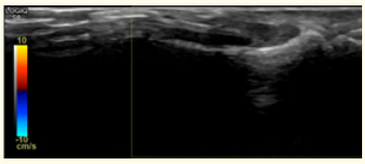

Figure 2: Ultrasound exam of the nail showing an hipoechoic, avascular lesion with internal echoic foci.

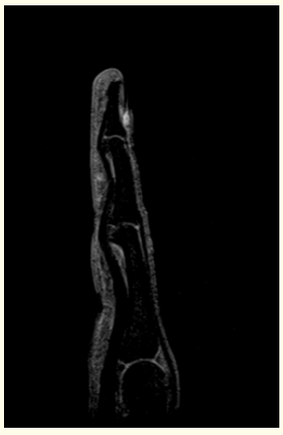

Figure 3: T1 Weighted contrast-enhanced MRI showing a lesion withing the nail matrix of the third finger.

The diagnosis of SAFM was presumed and longitudinal partial proximal plate nail avulsion was conducted.

Gross pathological evaluation of the partial nail avulsion revealed a consistent, gelatinous and well-demarcated flesh-coloured lesion (Figure 4).

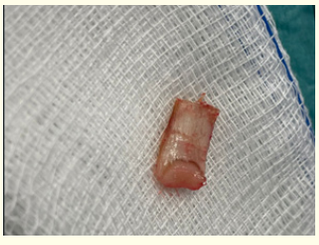

Figure 4: Excised lesion of the nail.

Histopathological examination showed a dermal expansion into the nail by a low/moderately cellular lesion composed of stellate/spindled cells with loose fascicular growth patter embedded in a myxocollagenous stroma. Immunohistochemical analysis demonstrated CD34 positivity and negativity for pS-100 immunomarkers (Figure 5). Periodic acid- Schiff (PAS) exam was negative for *fungii.*

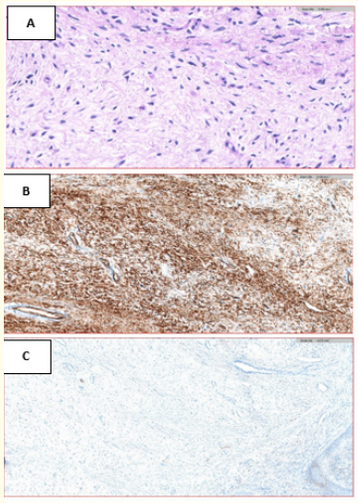

Figure 5: A - Histological examination showing fusiform cells within a myxoid stroma without cell atypia or mitotic figures. B- Positive immunohistochemical staining for CD 34. C -Negative immunohistochemical staining for Ps100.

After 12 months of follow-up the patient was completely asymptomatic, with no signs of lesion recurrence or nail dystrophia (Figure 6).

Figure 6: Clinical presentation at 1 year follow-up.

Discussion

SAFM is a rare soft tissue neoplasia, with little over 300 cases described in the literature [2]. This pathology is most often diagnosed in the fifth decade of life, with men accounting for around 60% of all cases [2,3]. Most cases occur in the digits, around the nail apparatus, with subungual cases being the most common clinical conditions [3]. The typical clinical presentation of the subungual SAFM is of a painless, slow-growing mass associated with xanthonychia, nail thickening, and proximal paronychia [2,4]. Due to this idle pathological onset, late diagnosis, or even, misdiagnosis with fungal infection often occur, postponing the correct surgical management [4]. History of previous trauma is described in a percentage of cases, although a causative relationship has not yet

been established [1-3]. No malignant transformation or metastatic disease has been reported. Recurrence is, however, a concern especially in positive margin surgical excisions [3].

Imaging methods may help in establishing a diagnosis and excluding similar lesions. SAFM typically presents in ultrasound studies as an avascular hypoechoic lesion with low level internal echoic *foci*. In MRI imaging T1 contrast enhancement is characteristic as well as T2 hyperintensity. Other imaging techniques such as plain radiography can be used to assess bone involvement [5].

Upon histological examination, SAFM is characterized by fusiform fibroblasts proliferating inside a myxoid/collagenous matrix. Microvasculature proliferation can often be observed. Mitotic figures/cellular atypia is rarely described in SAFM. Upon immunohistological evaluation, CD34 and CD99 positive staining and negative EMA and pS100 staining are characteristic of this lesion [1-3,6].

Differential diagnosis is made with glomus tumour, sclerosing perineuroma, acquired digital fibrokeratoma, periungeal fibroma (CD34 Negative), and superficial angiofibromyxoma, dermatofibromyxoma perturbrans and myxoid neurofibroma (CD 34 Positive) [2,4,6]. Definitive diagnosis is made upon analysing clinical, histological and immunohistochemical characteristics of the suspected lesion.

Surgical technique (Figure 7)

In order to achieve complete surgical excision of the lesion with clear margins and also to avoid recurrent pain and nail dystrophy, an adaptation of partial proximal nail plate avulsion technique was used [7]. Using a Nº 15 blade, two parallel longitudinal incisions were performed in the proximal nail fold in order to reflect the later and expose the lesion in the nail matrix. An English-anvil action nail splitter was used to cut and completely excise the lesion and nail plate around it. By doing this, complete inspection of the lesion embedded in the proximal nail matrix was permitted, allowing a clear-margin excision while preserving the lateral and distal aspect of the nail plate. The nail fold excisions were sutured with non-absorbable suture wire.

Figure 7: Post operative photograph showing 2 longitudinal incisions upon the proximal nail fold and the lesion excision preserving the healthy nail plate.

Conclusion

In conclusion, SAFM is a rare condition that should be considered in cases of slow-evolving xantonychia and paronychia. Partial proximal longitudinal resection may be used as a surgical technique providing good clinical and aesthetic results. Lesion recurrence should be avoided by clear margin excisions and close revaluation in clinic.

Bibliography

1. Fetsch JF., *et al*. "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* 32.7 (2001): 704-714.
2. Crepaldi BE., *et al*. "Superficial Acral Fibromyxoma: Literature Review". *Revista Brasileira de Ortopedia (Sao Paulo)* 54.5 (2019): 491-496.
3. Hollmann TJ., *et al*. "Digital fibromyxoma (superficial acral fibromyxoma): a detailed characterization of 124 cases". *The American Journal of Surgical Pathology* 36.6 (2012): 789-98.
4. Kalloniati E., *et al*. "Superficial Acral Fibromyxoma, Appearing as Retronychia: A Rare Clinical Entity". *Skin Appendage Disorders* 6.5 (2020): 312-314.
5. Lee S and MAR Reid. "Superficial acral fibromyxoma: a case report with radiological review". *Skeletal Radiology* 47.7 (2018): 1021-1028.
6. Ashby-Richardson H., *et al*. "Superficial acral fibromyxoma: an overview". *Archives of Pathology and Laboratory Medicine* 135.8 (2011): 1064-1066.
7. Collins SC., *et al*. "Alternatives to complete nail plate avulsion". *Journal of the American Academy of Dermatology* 59.4 (2008): 619-626.