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Case Report

Superficial Acral Fibromyxoma (SAF): A Case Report

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Abstract

Superficial Acral Fibromyxoma (SAF) is a benign soft tissue tumor characterized by the presence of stellate and spindle-shaped cells arranged in a storiform and fascicular pattern. This tumor is relatively rare, benign and slow-growing. If often involves peri- and subungual regions of fingers and toes in middle-aged adults, with a male predominance. This acral fibrous tumor is poorly known and its histology may suggest myxoid dermatofibrosarcoma, which carries a completely different prognosis and treatment. Thus, immunohistochemistry is extremely important to differentiate SAFM from other acral fibrous tumors. We present a case of SAFM that involved the nail matrix with canalicular dystrophy of the nail plate. This finding has not been described in the literature as of the date of publication of this case report. Hence, we believe that it is important to note this presentation so that this disease can be considered in the differential diagnosis of diseases that produce this type of nail dystrophy.

Keywords: Periungual Tumor; Nail Plate; Benign Tumor

Introduction

Superficial Acral Fibromyxoma (SAFM) is a rare, exophytic, benign tumor that was first described by Fetsch and colleagues in 2001. It usually affects the periungual and subungual regions, but there are also reported cases that involved the palms and ankles [1]. The differential diagnosis includes benign and malignant tumors, including myxoid dermatofibrosarcoma. Thus, a histopathological examination and immunohistochemical findings are needed for a definitive diagnosis and to determine the prognosis. SAFM has been observed in patients aged 14-75 years of age and shows a male predominance [2]. Here, we report the first case (to the best of our knowledge) of SAFM associated with canalicular dystrophy of the nail plate.

Case Report

A 53-year-old male, with no relevant history, sought a consultation for a painful papulo-nodular lesion that over 1 year had shown progressive growth and deformation of the nail plate of the fifth finger of the right hand. There were no triggering factors or associated systemic symptoms.

Physical Examination

We found a 5-mm papulo-nodular, erythematous, cupuliform lesion, with a soft consistency painful to the touch, involving the distal fold of the fifth fingernail of the right hand. There was loss of continuity of the "v" form in the proximal nail plate and distal canalicular dystrophy (Fig. 1).

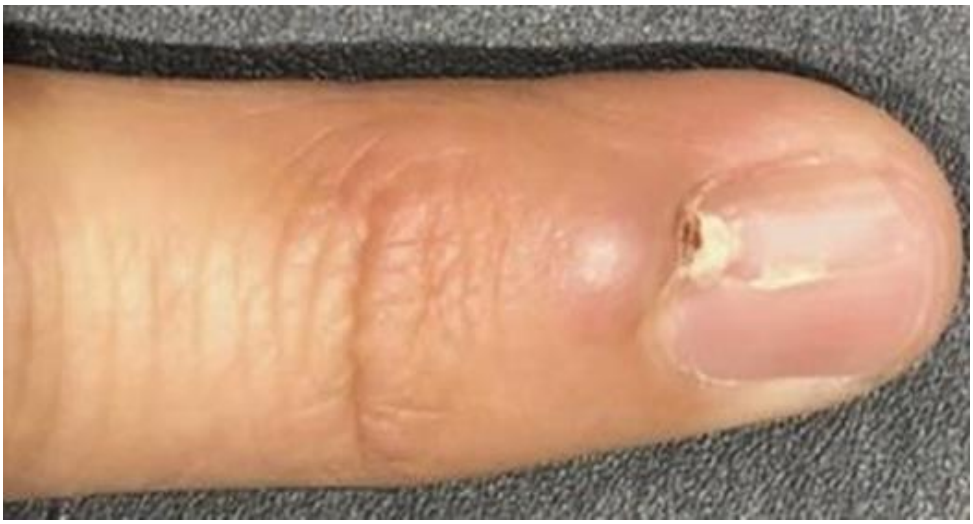

Figure 1: Clinical appearance of patient's fifth finger of the right hand.

Histopathology

Skin biopsy revealed a dermal non-encapsulated tumor that had resulted from the proliferation of stellate and spindle-shaped fibroblasts with a slightly storiform and fascicular pattern. It had a myxoid/collagenous stroma; dilated blood vessels; few lymphocytes, mast cells; and mitotic figures; and the absence of nuclear atypia (Fig. 2). Immunohistochemistry showed positive staining for CD34 and CD99 and negative staining for Epithelial Membrane Antigen (EMA), S-100, vimentin and smooth muscle actin.

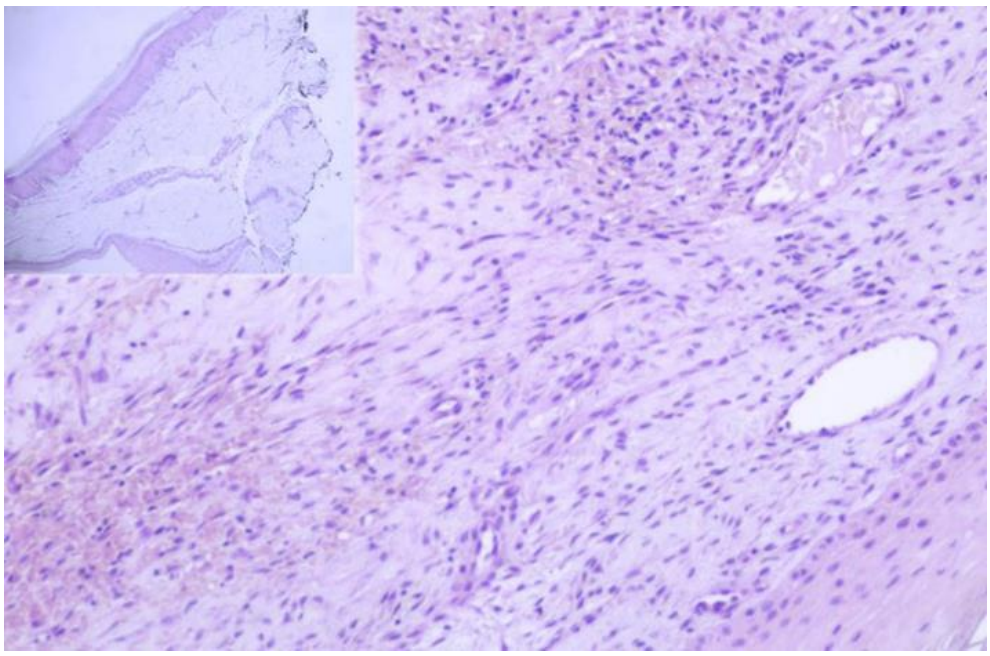

Figure 2: An image of the hematoxylin and eosin-stained skin biopsy (40× magnification). There are stellate and spindle-shaped fibroblasts with a slightly storiform and fascicular pattern, in myxoid/collagenous stroma, with dilated blood vessels. The inset in the upper left corner shows a panoramic view of the biopsy (4× magnification).

Other Complementary Tests

X-ray showed no bone alteration. Onychoscopy showed a circular cavity in the proximal nail plate, with keratinous material. Toward the proximal part of the eponychium, there was a rounded, non-pigmented, homogeneous pale pink tumor, furrowed by tortuous telangiectatic vessels. The distal part of the nail plate showed a depression-like "channel" delimited by two parallel hyperkeratotic ridges (Fig. 3).

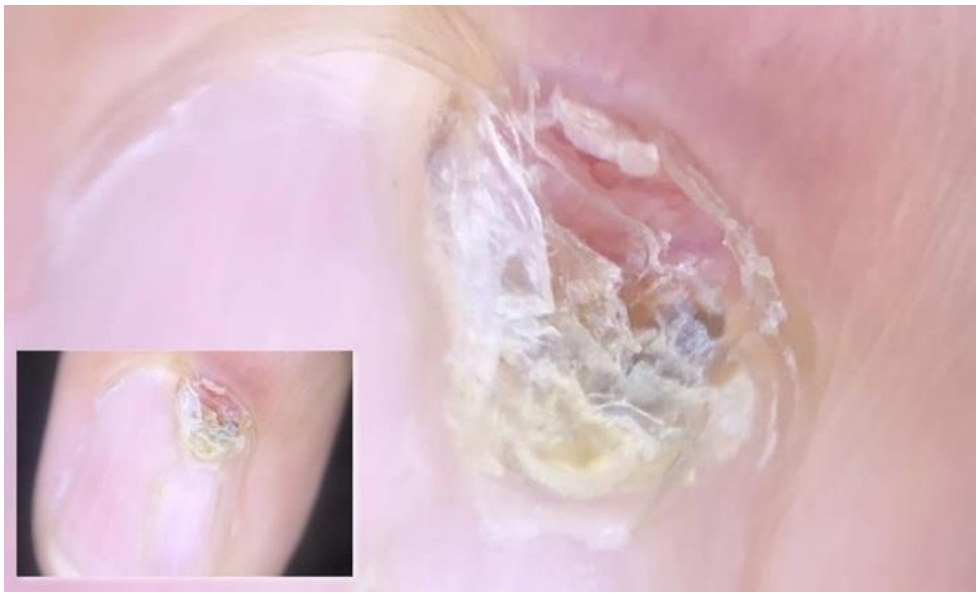

Figure 3: Onychoscopy with high magnification.

Diagnosis

We diagnosed the patient with SAFM.

Treatment and Evolution

We performed a window-shape avulsion of the skin of the proximal nail fold and found a well-demarcated tumor that formed a canaliculus by crushing the nail matrix. We debrided the tumor from the perilesional tissue and replaced the superficial skin. There was no recurrence during the 2-year follow-up.

Discussion

SAFM is a benign soft tissue tumor characterized by a solitary painless nodule, which varies in diameter. It is often located on the acral region, mainly the big toe. The prevalence is higher in men, with a sex ratio of 2:1 [3]. Clinically it presents as a solitary, asymptomatic nodule, usually located in acral areas such as the subungual and periungual region, although rare locations such as the heel, ankle, or palms of the hands have been reported. These tumors are usually slow growing (up to 30 years of evolution) and on average are 0.5-5 cm. While SAFM is usually confined to the dermis and subcutaneous tissue, 36% of cases may present bone lysis [4]. The nail plate could be affected in 50% of cases, but canalicular dystrophy has not been reported [1].

The diagnosis of SAFM requires histopathological analysis, which shows a well- circumscribed, unencapsulated dermal tumor, without contact with the underlying epidermis and with proliferation of spindle and/or stellate cells on a fibromyxoid stroma with increased vasculature. Mitotic figures are rare. Nuclear atypia, if any, is usually mild or moderate. The presence of hyperkeratosis or an epithelial collarette, mast cell infiltrate and/or cartilaginous and bone metaplasia have also been described [3]. Immunohistochemical analysis is important to reach a definitive diagnosis. The common finding is diffuse positive CD34 staining and focal EMA and CD99 staining. On the other hand, S100, HMB-45 and cytokeratin staining are negative; however, this staining can be variable [2,5,6].

Although there is no defined dermoscopic pattern, several findings have been described: a homogeneous pink color; small bright white striae; hyperkeratotic projections; red, brown and gray dots; and no vascular structures [1]. In our patient, high magnification onychoscopy revealed a pale pink tumor under a keratotic eponychium with evident vascularity on the surface.

The differential diagnosis includes tendon sheath fibroma, myxoid neurofibroma, glomus tumor, giant cell tumor of the tendon sheath and sclerosing perineurioma [4]. The treatment of SAFM is surgical. The recurrence rate is up to 20% in cases with incomplete extirpation. A 2-year follow-up is suggested [6].

Conclusion

SAFM is an acral tumor that remains difficult to diagnose, with a very long interval between the onset of symptoms and the final diagnosis, when we can see the deformity of the nail unit, as in our patient. There are two reasons for this diagnostic delay: the indolent character of the tumor and its very slow evolution and the common clinical and histological characteristics with other tumors of the ungual and periungual regions. A conservative surgical approach is recommended, with prolonged follow-up to prevent recurrence, eventual malignant transformation, or bone damage. However, no case of malignant transformation has been reported in the literature to date. In our patient, we performed a window-shaped avulsion of the skin of the proximal nail edge with tumor removal and repositioning of the superficial skin segment. Although there was no tumor recurrence during a 2-year follow-up, there was permanent residual canalicular dystrophic damage. Dermatologists and dermatopathologists should be aware of this entity in order to avoid misdiagnosis, which could lead to unwarranted mutilating surgery.

Conflicts of Interests

The authors declare that there is no conflict of interest for this paper.

References

1. Mejía-Rodríguez S, Maza de Franco A. Dermatoscopy of subungual digital fibromyxoma (superficial acral fibromyxoma). JAAD Case Rep. 2023;33:109-11.
2. 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. Hum Pathol. 2001;32:704-14.
3. Chiheb S, Mouradi M, Hali F. Superficial acral fibromyxoma: clinicopathologic analysis of five cases. Skin Appendage Disord. 2021;7:468-74.
4. Hashimoto K, Nishimura S, Oka N, Tanaka H, Ryosule K, Akagi M. Aggresive superficial acral fibromyxoma of the great toe: A case report and mini-review of the literature. Mol Clin Oncol. 2018;9:310-4.
5. Crestani L, Azevedo-Fasciani I, Kakizaki P, Sakai-Valente N. Case for diagnosis: Single-digit clubbing. An Bras Dermatol. 2020;95(4):524-6.
6. Sawaya J, Khachemoune A. Superficial acral fibromyxoma. Int J Dermatol. 2015;54:499-508.