CASE REPORT

Superficial Acral Fibromyxoma involving the nail's apparatus. Case report and literature review*

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DOI: http://dx.doi.org/10.1590/abd1806-4841.20142673

Abstract: Superficial Acral Fibromyxoma is a rare tumor of soft tissues. It is a relatively new entity described in 2001 by Fetsch et al. It probably represents a fibrohistiocytic tumor with less than 170 described cases. We bring a new case of SAF on the 5th toe of the right foot, in a 43-year-old woman. After surgical excision with safety margins which included the nail apparatus, it has not recurred (22 months of follow up). We carried out a review of the location of all SAF published up to the present day.
Keywords: Nails; Neoplasm recurrence, local; Neoplasms; Recurrence

INTRODUCTION

Superficial Acral Fibromyxoma (SAF) is a rare tumor of soft tissues with slow growth and acral location. It has a benign behavior, but it may persist or recur if not properly treated.

CASE REPORT

We present a 43-year-old woman, without any known allergies whose personal history reports beta thalassemia. She referred having had cutaneous changes not associated with any trauma for 8 years, consisting of swelling, partial nail loss and distal ulcerations with occasional bleeding on the 5th toe of the right foot. When the patient wore open shoes it was painless; however, it hurt and bled when she wore closed shoes. Upon examination, the distal end of the 5th toe presented a central ulcer with blood remains and partial onycholysis (Figure 1A).

Antero-posterior and oblique X-rays were requested of both feet, which showed subluxation of the distal phalanges of the 5th toes without signs of bone infiltration and a diagnostic biopsy was performed.

Histological results showed neoplastic dermal proliferation of fusiform cells without any relevant atypia, immersed in a myxoid stroma with collagenized areas and a prominent vascular weave (Figure 1B). Immunohistochemical studies reported positive results for CD34 and negative for S100, AME and AML (Figure 1C). The proliferation index, valued with Ki67 was low (less than 1%). These findings led to the diagnosis of Superficial Acral Fibromyxoma.

The subsequent therapeutic approach included complete removal of the tumor as well as the nail in order to avoid recurrence.

Histological examination of the surgical piece was similar to the previously described. The tumor was extirpated with wide margins, including the nail matrix, respecting the distal phalanx (Figure 1D). Resection margins were reported as tumor-free.

After a 22-month follow-up there was no recurrence of the tumor.

DISCUSSION

Superficial Acral Fibromyxoma (SAF) is a rare tumor of soft tissues, with slow growth and located in the subungual or periungual region of the hands and feet[1-7] (Table 1). However, the heel, palm and ankle can also be affected.[5] It affects young adults (mean age 43 years old), with higher frequency in men than in women in a 2:1 proportion. It is a relatively new entity described in 2001 by Fetsch *et al*.[4] It probably repre-

Received on 04.04.2013.
Approved by the Advisory Board and accepted for publication on 17.07.2013.
* *Work performed at the Hospital Universitario Virgen Macarena - Sevilla, Spain.
Financial Support: none
Conflict of Interests: none

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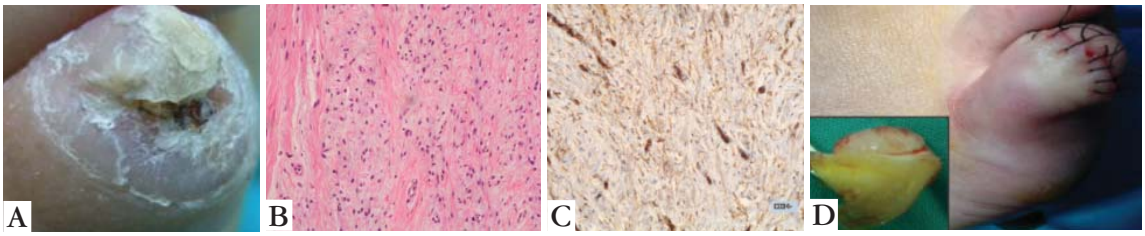

Figure 1: A: Loss of the distal end of the 5th toe of the right foot and partial onycholysis; **B:** Fusiform cell proliferation in a myxoid stroma (HE, x100); **C:** Fusiform cells show cytoplasmic positivity to CD34 (PAP, x200); **D:** Image after excision. The macroscopic part of the tumor along with nail apparatus can be seen

Table 1: Summary of all published cases and locations of SAF

References	Cases described	Location	Nº Cases	References	Cases described	Location	Nº Cases
Fetsch JF et al. Hum Pathol. 2001; 32:704-14.	37	Toes Fingers Palm	20 13 4	Tardío JC *et al*. Am J Dermatopathol. 2008; 30:431-5.	4	Big toe Middle finger Palm of hand Thumb of hand	1 1 1 1
Kazakov DV *et al*. Dermatology. 2002; 205:285-8.	2	Toes	2	Luzar B and Calonje E. Histopathology. 2009; 54:375-7.	14	Big toe ¿	8 6
Meyerle JH *et al*. J Am Acad Dermatol. 2004; 50:134-6.	1	Subungueal index finger	1	Pasquinelli G *et al*. Ultrastruct Pathol. 2009; 33:293-301.	1	Index finger	1
André J *et al*. Am J Dermatopathol. 2004; 26:472-4.	1	Great toe-nail	1	Wang QF et al. Zhonghua Bing Li Xue Za Zhi. 2009; 38:682-5.	1	Middle finger	1
Quaba O *et al*. Br J Plast Surg. 2005; 58:561-4.	1	Ring finger	1	Goo J *et al*. Ann Dermatol. 2010; 22:110-3.	1	Subungueal index finger	1
Abou-Nukta F *et al*. J Hand Surg Br. 2006; 31:619-20.	1	Nail of the thumb	1	Chattopadhyay M *et al*. Clin Exp Dermatol. 2010;35:807-9.	1	Subungueal index finger	1
Oteo -Alvaro A *et al*. Arch Orthop Trauma Surg. 2008; 128:271-4.	1	Toe	1	Cogrel O *et al*. Ann Dermatol Venereol. 2010; 137: 789-93.	3	Great toe Second Toe Finger	1 1 1
Misago N *et al*. J Eur Acad Dermatol Venereol. 2008; 22:255-7.	1	Tip of big toe	1	Fanti PA *et al*. G Ital Dermatol Venereol. 2011;146: 283-7.	12	Toes Fingers	¿ ¿
Varikatt W *et al*. Skeletal Radiol. 2008; 37:499-503.	2	Tip of index finger	2	Messeguer F *et al*. Actas Dermosifiliogr. 2012; 103:67-9.	1	Index finger	1
Al-Daraji WI *et al*. J Cutan Pathol. 2008; 35:1020-6.	32	Toes Fingers Heel	15 13 4	Ben Brahim E *et al*. Tunis Med. 2012;90:340-1.	1	Toe	1
Al-Daraji WI *et al*. Dermatol Online J. 2008; 28:14-27.	2	Subungueal big toe Index finger	1 1	Wakabayashi Y *et al*. Acta Dermatovenerol Croat. 2012; 20:263-6.	1	Great toe	1
Prescott RJ *et al*. Br J Dermatol. 2008; 159:1315-21.	41	Toes Fingers Palm	29 11 1	Wei C et al. Eplasty. 2013; 13: ic13.	1	Thumb	1

CHART 1: Differential Diagnosis of Superficial Acral Fibromyxoma

CD34+ Neoplasias	CD34- Neoplasias	Other lesions
Dermatofibrosarcoma protuberans	Giant cell tumor of tendon sheath	Fibroma of tendon sheath
Superficial angiomyxoma	Glomus tumor	Onychocryptosis
Myxoid neurofibroma	Sclerosing perineuroma	Cutaneous muxoma
Sclerosing fibroma	Benign fibrous histiocytoma	
Acral myxoinflammatory	Acral fibrokeratoma	
Spindle cell lipoma		

sents a fibrohistiocytic tumor with less than 170 described cases (SAF series and isolated case reports).[2]

Pain is not usually mentioned. Ungual involvement may be present. Only one case has been associated with previous trauma. X-rays rarely show bone alterations.[1,8]

Histologically, it is a well delimited, non-encapsulated dermal tumor that may extend towards the hypodermis. It is composed of a proliferation of cells from a fibroblastic line usually accompanied by many mast cells. The presence of a myxoid stroma with a rich vascular weave is very noticeable. Epidermis hyperplasia with hyperkeratosis is also frequent. CD34 positivity is characteristic but CD10, CD99, EMA, and nestin immunoreactivity are also common. Negative results for neural and muscular differentiation markers (S-100, HMB-45, SMA, desmin, actin), cytokeratin and apolipoprotein D are expected.[1-3, 9,10]

Although it is an infrequent event, it must be included in differential diagnosis of tumors present on the fingers and toes.[1-7] Differential diagnosis considerations are summarized in chart 1.

SAF has a benign behavior but may persist or recur if not properly treated.[7,8] Thus complete removal and follow-up is recommended. Up to this date, malignization has not been described.

In conclusion, this is the description of a rare case of Superficial Acral Fibromyxoma on the nail apparatus of a 43-year-old woman. There are less than 170 published cases. It is a benign tumor with slow growth and, although rare, it should be considered in differential diagnosis of acral lesions. Surgery is curative but requires adequate margins due to the high risk of recurrence. Malignization has never been described. ❑

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How to cite this article: Márquez García A, Mendonça FMI, Perea Cejudo M, Camacho Martínez FM, Ríos Martín JJ. Superficial Acral Fibromyxoma involving the nail 's apparatus. Case report and literature review. An Bras Dermatol. 2014;89(1):147-9.