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).

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.

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. 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. It probably repre-
### 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.                    | 37              | Toes          | 20       | Tardio JC et al.                    | 4               | Big toe       | 8        |
| Hum Pathol. 2001; 32:704-14         |                 | Fingers       | 13       | Am J Dermatopathol. 2008; 30:431-5. |                 | Middle finger | 1        |
| Kazakov DV et al.                   | 2               | Toes          | 2        | Luzar B and Calonje E. Histopathology. 2009; 54:375-7. | 14              | Big toe       | 1        |
| Meyerle JH et al.                   | 1               | Subungueal    | 1        | Pasquini G G et al. Ultrastruct Pathol. 2009; 33:293-301. | 1               | Index finger  | 1        |
| André J et al.                      | 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 | 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         | 1        |
| Varikatt W et al. Skeletal Radiol. 2008; 37:499-503. | 2              | Tip of index finger | 2 | Messegueur F et al. Actas Dermosifiliogr. 2012; 103:67-9. | 1               | Fingers       | 1        |
| Al-Daraji WI et al. J Cutan Pathol. 2008; 35:1020-6. | 32             | Toes          | 15       | 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 | 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          | 29       | Wei C et al. Eplasty. 2013; 13: ic13. | 1               | Thumb        | 1        |
|                                    |                 | Fingers       | 11       |                                    |                 |               |          |
|                                    |                 | Palm          | 1        |                                    |                 |               |          |

*An Bras Dermatol. 2014;89(1):147-9.*
sents a fibrohistiocytic tumor with less than 170 described cases (SAF series and isolated case reports). 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.

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.

Although it is an infrequent event, it must be included in differential diagnosis of tumors present on the fingers and toes. SAF has a benign behavior but may persist or recur if not properly treated. 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.

**REFERENCES**

1. Ashby-Richardson H, Rogers GS, Stadecker MJ. Superficial acral fibromyxoma: an overview. Arch Pathol Lab Med. 2011;135:1064-6.
2. Wakabayashi Y, Nakai N, Takemaka H, Katoh N. Superficial acral fibromyxoma of the great toe: case report and mini-review of the literature. Acta Dermatovenerol Croat. 2012;20:263-6.
3. Messegue F, Nogere E, Aguix-Mejias A, Traves V. Fibromixoma acral superficial, un tumor periungueal CD34 positivo. Actas Dermosifiliogr. 2012;103:67-9.
4. 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.
5. Wei C, Fleeger EJ. Superficial acral fibromyxoma of the thumb. Eplasty. 2013;13:e13.
6. Prescott BJ, Husain EA, Abdellau9 A, Al-Mahmoud RM, Khan M, Salmin WD, et al. Superficial acral fibromyxoma: a clinicopathological study of 41 new cases from the UK: should myxoma (NOS) and fibroma (NOS) continue as part of 21st-century reporting? Br J Dermatol. 2008;159:1315-21.
7. Al-Daraji Wl, Miettinen M. Superficial acral fibromyxoma: a clinicopathological analysis of 32 tumors including 4 in the heel. J Cutan Pathol. 2008 Nov;35(11):1020-6.
8. Goo J, Jung YJ, Kim JH, Lee SY, Ahn SK. A Case of Recurrent Superficial Acral Fibromyxoma. Ann Dermatol. 2010;22:110-3.
9. Quaba O, Evans A, Al-Nafussi AA, Nassan A. Superficial acral fibromyxoma. Br J Plast Surg. 2009;58:561-4.
10. Frierson HF, Cooper PH. Myxoid variant of dermatofibrosarcoma protuberans. Am J Surg Pathol. 1983;7:445-50.

**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.