Superficial Acral Fibromyxoma of the Great Toe: Case Report and Mini-Review of the Literature

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SUMMARY Superficial acral fibromyxoma (SAF) is a rare, distinctive, benign soft tissue tumor with a predilection for the hands and feet, first described by Fetsch *et al.* in 2001. Histologically, SAF tumors are unencapsulated, mostly located in the dermis or subcutis, and composed of spindled and stellate-shaped cells with random, loose storiform and fascicular growth patterns. The stromal component of the tumor was Alcian blue-positive. The tumor cells were mostly immunopositive for CD34, vimentin, and CD99; often immunopositive for EMA; and immunonegative for S-100, HMB-45, SMA, desmin, and keratin. There have been 19 reports of 149 SAF cases in the English language literature. However, SAF is not widely recognized because it is an uncommon occurrence that has been described only relatively recently. Herein, we report a case of SAF and describe the clinicopathologic characteristics based on a review of published SAF cases from July 2001 to July 2011.

KEY WORDS: superficial acral fibromyxoma, benign soft tissue tumor, mini-review, clinicopathologic characteristics

INTRODUCTION

Superficial acral fibromyxoma (SAF) is a rare, distinctive, benign soft tissue tumor with a predilection for the hands and feet, first described by Fetsch *et al.* in 2001 (1). There have been 19 reports of 149 SAF cases in the English language literature (1-19), including two recent review articles (9,13). However, SAF is not widely recognized because it is an uncommon occurrence that has been described only relatively recently. Herein, we report a case of SAF and describe the clinicopathologic characteristics based on a review of published SAF cases from July 2001 to July 2011.

CASE REPORT

A 51-year-old Japanese woman was referred to our department for diagnosis of a slight hyperkeratotic, pinkish, round nodule of 10 mm in diameter located in the distal aspect of her right great toe (Fig. 1). She had no nail deformity and had not experienced any subjective symptoms for the past 14 months. She had no particularly relevant medical history, except for diabetes mellitus. Laboratory data, including liver and kidney function, were normal. A biopsy speci-

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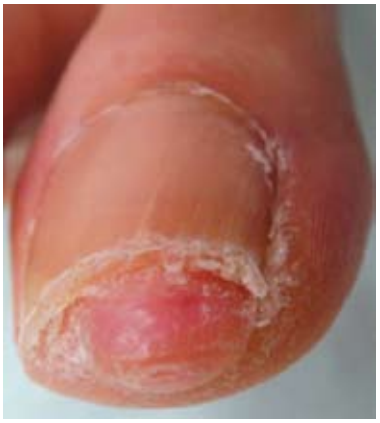

Figure 1. A slight hyperkeratotic, pinkish, round nodule of 10 mm in diameter was located in the distal aspect of the right great toe. Nail deformity was not seen.

men of the nodule showed no malignancy. En bloc excision with a 2-mm margin was performed under local anesthesia because the nodule was causing problems with her daily activities. In addition, histologic examination of the entire tumor was considered to be crucial for accurate diagnosis and understanding of the possible spread of the tumor. The tissue defect after excision of the tumor healed in 3 weeks with conservative treatment.

Excised specimens showed an unencapsulated fibromyxoid tumor with hyperkeratosis located in the deep dermis (Fig. 2a). Abundant stromal mucinous material was detected by Alcian blue staining. A higher magnification showed stellate and spindle cells with a fascicular pattern embedded in myxoid stroma with scattered small vessels and mast cells, but no nuclear atypia or mitotic figures (Fig. 2b). Immunohistochemically, the tumor cells were positive for CD10, CD34 (Fig. 2c), CD99, and vimentin, and negative for S-100, HMB-45, epithelial membrane antigen (EMA), and smooth muscle actin (SMA). On the basis of these histologic features, we diagnosed the nodule as SAF. The tumor was excised completely, but a close margin from the tumor excision was detected. Therefore, we have continued careful observation of the tumor, and there has been no evidence of recurrence for over 1 year postoperatively.

DISCUSSION

Including our case, there have been 20 reports of SAF (149 patients) in the literature (1-19). The patients (96 males, 47 females, and 6 patients of unidentified gender) had an age range between 14 and 91 years

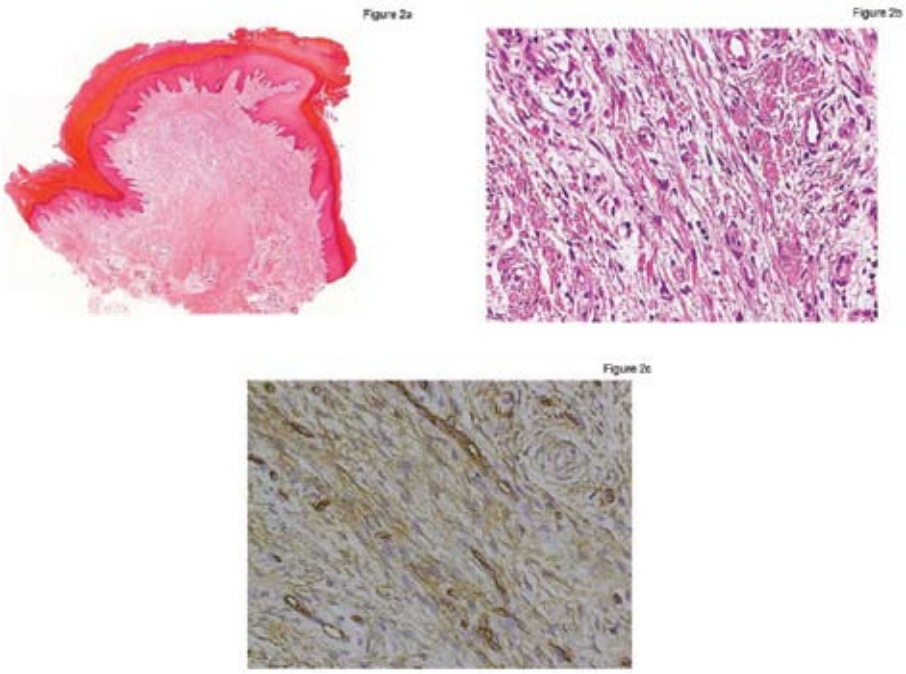

Figure 2. Histopathologic examination of the tumor: (a) an unencapsulated fibromyxoid tumor with hyperkeratosis was located in the deep dermis (H&E; ×40); (b) stellate and spindle cells with a fascicular pattern were embedded in myxoid stroma with scattered small vessels. There were neither nuclear atypia nor mitotic figures (H&E; ×200); (c) the tumor cells were positive for CD34 (×200).

old (mean 48.2, median 49.0 years). The tumor mostly presented as a painless solitary mass or nodule with an average size of 17.8 mm (range 5-50 mm; median 15.0 mm). The tumor involved a toe (n=82, 55.1%), heel (n=4, 2.7%), finger (n=46, 30.9%), palm (n=6, 4.0%), and unknown site (n=11, 7.4%). The most frequently affected site was the great toe (n=62, 43.1%). In available records, 35 tumors involved the nail region or were located close to the nail, and 6 tumors caused deformity of the nail. The underlying bone was affected in 11 tumors, but there was no evidence of systemic involvement. A history of trauma was reported in only 13 cases. The duration of the tumor ranged from 3 months to 30 years (mean, 5.1 years; median, 4 years).

Histologically, SAF tumors are unencapsulated, mostly located in the dermis or subcutis, and composed of spindled and stellate-shaped cells with random, loose storiform and fascicular growth patterns (1). The tumor cells are embedded in a myxoid or collagenous matrix, often with a mildly to moderately accentuated vasculature and an increased number of mast cells (97.8% of 46 evaluable cases). Slight to moderate cytologic or nuclear atypia was detected in some cases, but clinically aggressive behavior was not observed.(1,2,9,13,14). The stromal component of the tumor was Alcian blue-positive (100% of 5 evaluable cases). The tumor cells were mostly immunopositive for CD34 (96.3% of 135 evaluable cases), vimentin (100% of 80 evaluable cases), CD99 (81.3% of 48 evaluable cases); often immunopositive for EMA (59.2% of 120 evaluable cases); and immunonegative for S-100 (99.2% of 126 evaluable cases), HMB-45 (100% of 102 evaluable cases), SMA (99.0% of 99 evaluable cases), desmin (99.2% of 120 evaluable cases), and keratin (100% of 119 evaluable cases).

In 143 cases from available records, excision (n=131), biopsy or partial excision (n=6), or amputation of the affected digit (n=6) was performed. Follow up information was available for 71 cases (mean follow up, 47.9 months; median follow up, 18.4 months). Tumor recurrence was found in 9 of 131 excised cases after a mean period of 15.1 months. In 3 evaluable cases treated by biopsy or partial excision, one showed stable disease and 2 showed local tumor growth. The mean duration was 26.0 months. The relationship between the margin clearance at excision and the recurrence rate of SAF was unclear because of a lack of detailed information. However, Luzar and Calonje found no recurrence of the tumor in 7 patients with incomplete tumor excision over a mean follow up period of 18.4 months (16). Fetsch *et al*. (1) suggest that SAF rarely progresses to a low-grade neoplasm based on the cytologic atypia in histopathologic findings.

CONCLUSION

We report a case of SAF. The true malignant potential of SAF is not clear at present, and careful observation for at least two or three years after complete excision is required on the basis of the mean recurrence period.

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By bad weather – Nivea cream; year 1936.
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