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Superficial Acral Fibromyxoma

CASE REPORT

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ABSTRACT

Superficial acral fibromyxoma (digital fibromyxoma) is a rare tumor with a predilection for the subungual or periungual regions of the hands and feet. Generally, it states at dermis and has no capsule. Proliferation of the spindle and stellate-shaped fibroblastic cells in storiform and a scattered or fascicular pattern are characteristic features of this tumor. There are mast cells in myxoid or collagenous stroma. Tumor cells are often positive for CD34, CD99, and EMA immunohistochemically. In the treatment of these tumors, complete excision is recommended because of the potential recurrence. Here, we present a 60-year-old male patient with a papillomatous lesion on the second finger of his right hand. The tumor was extending into the nail bed. After excision, it was diagnosed as superficial acral fibromyxoma. The histopathological and immunohistochemical features of these rare tumors are discussed.

Keywords: Fibroma, myxoma, fibromyxoma

INTRODUCTION

Superficial acral fibromyxoma (SAF) was first defined in 2001 by Fetsch as a tumor with histological and immunohistochemical features located in acral extremities (1). It is generally a slow-growing soft tissue tumor in men. Since it causes no pain, patients usually seek medical help late. Patients state that the lesion has been present for many years. Few cases have a trauma history (2).

Microscopic examination reveals a lesion in the dermis with indistinct borders. The lesion consists of spindled and stellate-shaped cells in the myxocollagenous stroma. In spindle-shaped cells, CD34, CD99 and vimentin are positive immunohistochemically (1). In the differential diagnosis, angiomyxoma, fibrokeratoma, dermatofibroma, dermatofibrosarcoma protuberans, subungual acral malignant melanoma, and particularly neurofibroma are observed. It is important to use immunohistochemical methods in the diagnosis of these tumors, since there is a high recurrence risk after incomplete excision.

CASE REPORT

After the physical examination of a 60-year-old male patient who presented with a lump on his right index finger, a papillomatous mass 1 cm in diameter in the second nail bed was detected. In the histopathological examination performed after the excision, under the epidermis, nodular lesions with indistinct borders was observed (Figure 1). The lesion had a hypocellular appearance. Loose myxoid stroma, cells with spindled-shaped nuclei, and scattered cytoplasmic extensions showing no distinct fasciculations attracted attention (Figure 2, 3). A great number of mast cells were detected histochemically through toluidine blue (Figure 4). Immunohistochemically, spindled-shaped cells were positive for CD34 and EMA but negative for CD99 (Figure 5). Immunoreactivity with actin, desmin, and S-100 was not observed (Figure 6). The case was determined as superficial acral fibromyxoma with these findings. Written patient consent was received for the case presented in the article.

DISCUSSION

Superficial acral fibromyxoma (SAF) is a recently defined and relatively rare tumor. While defining this new tumor, Fetsch reevaluated 280 cases in the archives of AFIP (The Armed Forces Institute of Pathology) for the years 1970-2000 that had been diagnosed as fibroma, fibromyxoma, fibromyxoid nodules, fibrous histiocytoma, digital fibrokeratoma, angiofibroma, angiomyxoma, myxolipoma, and myxoma. By presenting 37 cases that had similar histological views, Fetsch thought that these tumors were proper for superficial acral fibromyxoma terminology (1). After that, tumor series with similar studies from different archives that fit the histological view defined by Fetsch were published (1-10).

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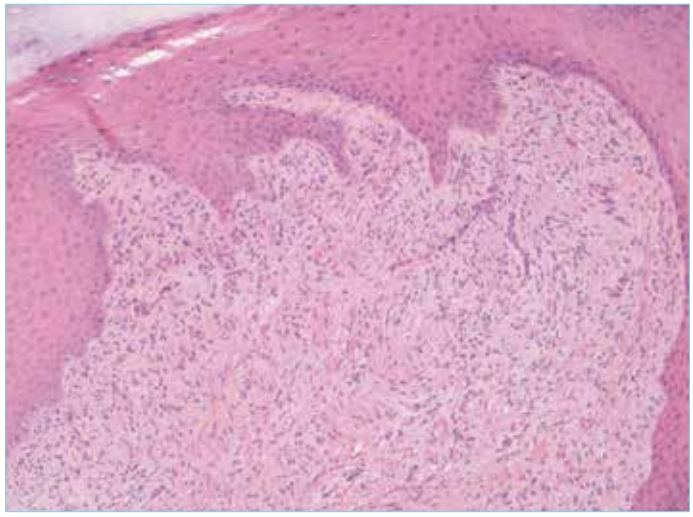

Figure 1. Nonencapsulated lesion under the epidermis (H&E x100)

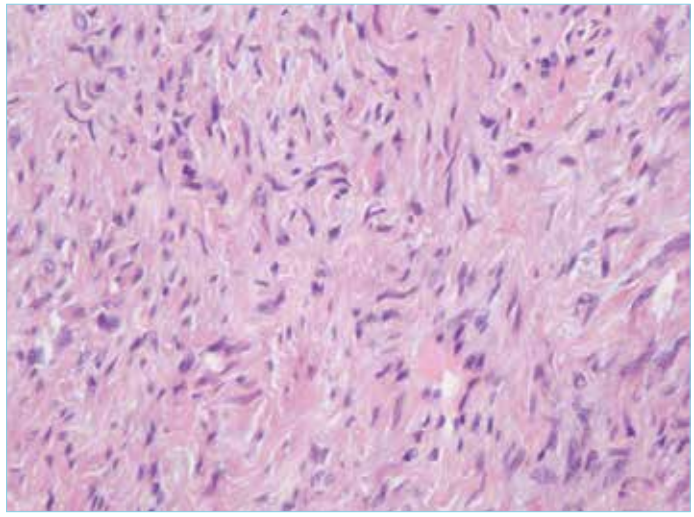

Figure 2. Spindle-shaped cells forming short bundles (H&E x200)

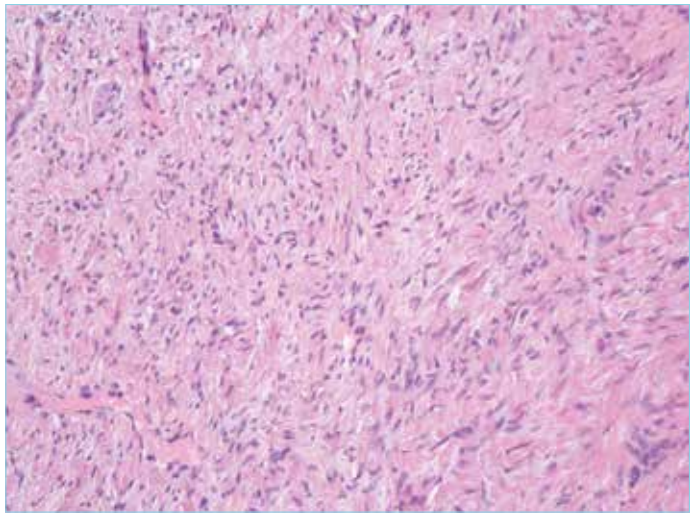

Figure 3. Mild nuclear hyperchromasia and pleomorphism in cells (H&E x400)

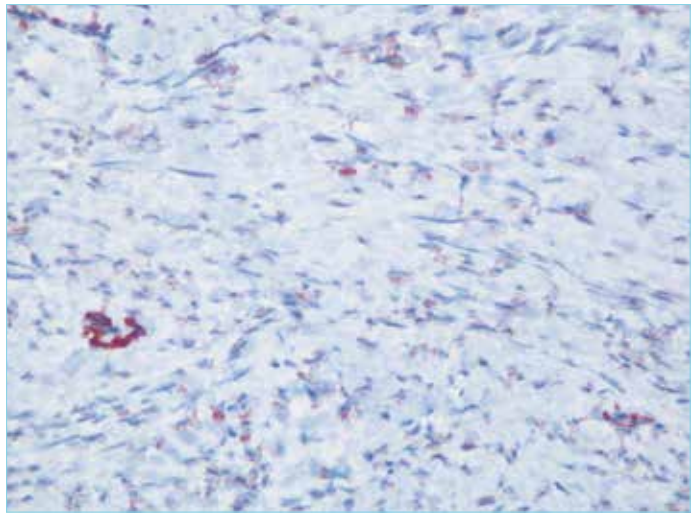

Figure 4. A large number of mast cells in the tumor showing metachromatic staining (Toluidine blue x200)

The average age is 43-50 and is twice as common in men (1,3). Most tumors occur on the toe and the finger. The tumor is located in the nail bed in half of the cases. There is no family history related to other soft tissue lesions in the cases. Just a few cases have a trauma history (1, 3). Fetsch reported recurrence in 3 out of 18 patients on whom he could perform a follow-up, and Prescott reported recurrence in 5 of his 12 patients. Our case was a 60-year-old patient, and a nodular lesion was present under the nail bed of his second finger. There was no evidence of recurrence during his 1-year follow-up.

Histopathologically, these tumors are non-encapsulated and have no clear circumscription. The tumor is located in the dermis and subcutaneous tissue. Extension to the fascia and bone is rare. It can be seen that spindled-shaped cells may be arranged in a loose storiform or fascicular pattern. The stroma might be collagenous or myxoid. It was reported that myxoid tumors are mostly located in the hands, whereas tumors with collagenous stroma are located in the feet (1). Mast cell growth is remarkable. Necrosis has not been defined in any cases. Mitosis is rare (<1/50 major growth fields). Mild to moderate nuclear atypia might be found. Hyperkeratosis in the epidermis, irregular acanthosis, and papillomatosis might be observed (1, 3, 4, 7).

Superficial acral fibromyxoma (SAF) stains extensively and is intensely positive for CD34 and vimentin immunohistochemically. Tumor cells stain positive for CD99. Keratin, actin, desmin, and S-100 are negative. Neurofibroma, angiomyxoma, fibrokeratoma, dermatofibroma, dermatofibrosarcoma protuberans, low-graded myxofibrosarcoma are considered in the differential diagnosis, and immunohistochemical methods play an important role in the differential diagnosis. Myxoid or myxocollagenous forms of SAF can be confused with neurofibroma. However, unlike SAF, neurofibroma does not have an appearance of vascularity, and it is S-100 positive. Superficial angiomyxoma is located mostly in the trunk and head-neck. It has a multilobule appearance. As a diffuse, it is myxoid and contains neutrophils. The storiform growth pattern in dermatofibroma is well developed. The epidermis is hyperplastic. The lesion has more irregular borders. Factor 13a and actin positivity with CD34 negativity-focal positivity are helpful in

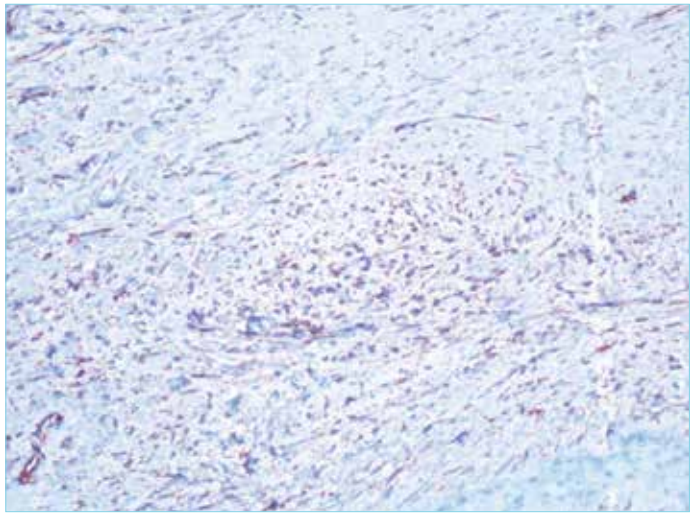

Figure 5. **(CD34 x200)**

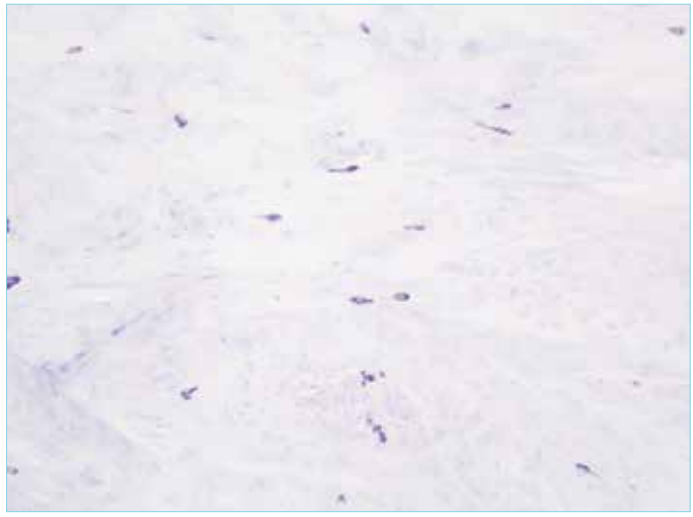

Figure 6. **(Actin x200)**

the diagnosis. Acral fibrokeratoma (acquired digital fibrokeratoma, periungual fibroma, periungual fibrokeratoma) has smaller, more superficial, and exophytic lesions. Cellularity and vascularity are lower, and collagenated stroma is more evident. Factors are 13a (+), CD34 (+/-), EMA (-), and S-100 (-). Dermatofibrosarcoma protuberans (DFP) and SAF have immunohistochemically similar profiles. CD34, CD99, and CD10 are positive in both of them. However, DFP is rarely superficial-acral located. It forms larger masses. The storiform pattern is distinct. Infiltration to subcutaneous adipose tissue is typical. Myxoid may vary focally. In difficult cases, the determination of the COL1A1-PDGFB fusion gene that is formed through translocation between chromosomes 17 and 22 may confirm the diagnosis. Low-graded myxofibrosarcoma has a tendency to be located in the extremities, and CD34 is positive in varying proportions. Significant nuclear atypia, the typical pattern of capillaries, perivascular concentration of tumor cells, and EMA negativity might be useful in the diagnosis (3, 8).

CONCLUSION

Superficial acral fibromyxoma is a rare and relatively new entity but must be kept in mind in the diagnosis of tumors. The biological behavior is not clear, due to the scarcity of patients who can be followed up for a long time.

However, it is reported that recurrence rates might be up to 42% (3). Malignancy criteria have not been defined yet. The importance of the significant nuclear atypia that is observed in some patients is not known (4). Therefore, complete excision of the tumor and follow-up of the patients are recommended.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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Authors' Contributions: Conceived and designed the experiments or case: DA, FC, YK. Performed the experiments or case: DA, FC, YK. Analyzed the data: DA, FC, YK. Wrote the paper: DA, FC, YK. All authors have read and approved the final manuscript.

Conflict of Interest: No conflict of interest was declared by the authors.

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