

A Case of CD34-Negative Superficial Acral Fibromyxoma

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ABSTRACT

Superficial acral fibromyxoma (SAF) is a tumor that occurs on the distal phalanges of the digits. As it does not spontaneously regress and is often associated with pain, the primary treatment is surgical resection. It is often associated with the nail component of the affected digit, and thus cosmesis is an important goal of the operation. We herein describe a case of SAF on the distal phalanges of the fifth digit of the foot beneath the nail, which was successfully resected with the nail component kept intact. Moreover, although SAF is most commonly CD34-positive, the present case was CD34-negative except for endothelial cells within the tumor. While CD34-negative SAF has been previously reported, the current case further indicates that CD34-positivity is not essential for the diagnosis of SAF.

Keywords: Superficial Acral Fibromyxoma; CD34; Nail

1. Introduction

There are a variety of tumors that can affect the acral extremities including superficial acral fibromyxoma (SAF), fibroma, exostosis, enchondroma, tylosis, and glomus tumor. The growth of some of these is accompanied with pain and digital deformity. SAF is a tumor observed in the apical area and is usually accompanied with pain secondary to the stiffness of tumor within its capsule. When the tumor is associated with the nail, cosmesis is an important factor during tumor resection. We report a case of SAF growing beneath the nail of the right fifth toe which was successfully removed with the nail component kept intact. Immunohistochemical analysis indicated that the resected tumor did not stain positive for CD34, which is rare in cases of SAF.

2. Case Report

A 40-year-old woman noticed a small growth on her right fifth toe five years prior to her presentation with us. The tumor regressed, but it eventually grew again and was accompanied by pain. As the pain increased in severity it became hard for her to wear a shoe on the affected foot, and she subsequently presented to our hospital. There was no history of trauma or other event related to the onset of the tumor. The mass measured 8 mm in length and consisted of greyish-white tissue with well-defined margins, and felt cartilaginous in consistency (**Figure 1**). The epidermis did not show parakeratinization or hyperkeratinization and was grossly intact. X-ray

images showed no calcification. Magnetic resonance imaging (MRI) of the mass demonstrated low signal intensity on T1 and T2-weighted images (**Figure 2**).

As the tumor did not invade the surrounding tissue on these images and was presumed to be benign, a simple resection was performed. Under spinal anesthesia, a transverse incision on the toe was made with the nail bed kept intact. The tumor was resected and the excess skin was trimmed away. Then the skin was approximated with 5 - 0 nylon. The contour of the toe after the operation did not affect of the shape of the nail (**Figure 3**). Histological examination showed a proliferation of collagen, fibroblastic cells, and edematous change compatible with SAF (**Figure 4(a)**). Immunohistochemical examination for the presence of CD34 in the specimen was negative except for within endothelial cells (**Figure 4(b)**).



Figure 1. Macroscopic view before operation.



(a)

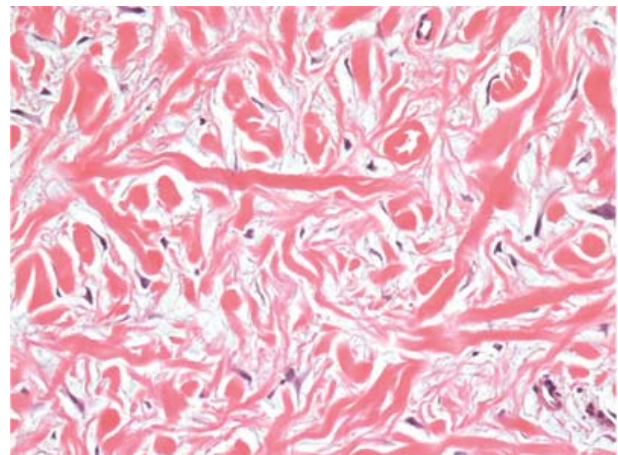


(b)

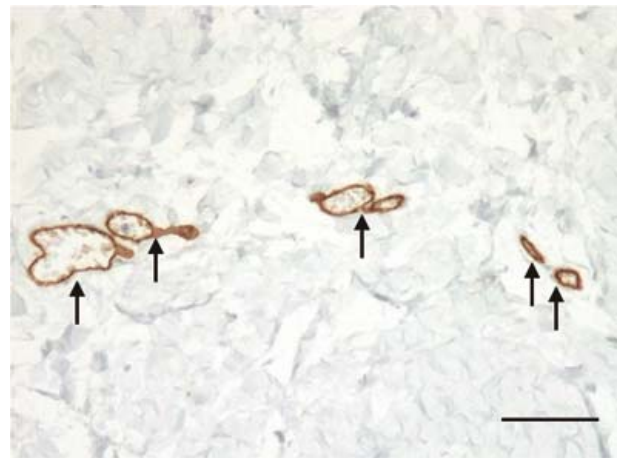
Figure 2. MRI images. MRI of the mass demonstrated low signal intensity on T1 (a) and T2 (b)-weighted images.



Figure 3. Macroscopic view three months after operation. The contour of the toe was preserved.



(a)



(b)

Figure 4. Histological section of SAF. H-E staining showing that cells in the tumor were spindle and stellate in shape and associated with the myxoid matrix and a delicate vascular network (a). Immunohistochemical staining of the tumor for CD34 shows that no cells except for endothelial cells (arrow) were positive for CD34 (b). Bar = 100 μ m.

3. Discussion

Fibromyxoma is a fibroma that has undergone myxomatous degeneration. Superficial acral fibroma was reported by Fetsch *et al.* in 2001 [1] as an uncommon tumor of the superficial soft tissue of the acral extremities. Most cases present in middle-aged adults as a long-standing, solitary mass measuring between 1 and 2 cm on the hands or feet, and are often observed in a subungual location [1,2].

In regards to operative management, the tumor should be resected at the margin of the tumor as SAF is benign. Although the tumor exists in close proximity to the nail bed, it is of mesenchymal origin and can be removed without affecting the nail component [3].

Pathologically, SAF is characterized by a slight to moderate cellular proliferation of spindle and stellate-shaped fibroblastic cells arranged in a random or fas-

cicular growth pattern, with myxoid stroma and prominent vascularity [2,4]. It has been reported that the tumor cells of the SAF are diffusely positive for CD34 and may be focally positive for EMA, CD10, and CD99 [4,5-8].

In the present case, most of the tumor consisted of fibrous tissue with some areas showing myxomatous change. This is associated with the deposition of mucin among the fibrous part. From these histological findings, we diagnosed this tumor as SAF.

Immunohistochemical examination for CD34 was negative except for the endothelial cells within the tumor. CD34-negative SAF has been reported previously [4], and as supported by another example in the present case, the diagnosis of SAF should not be made solely on the basis of immunostaining for CD34.

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