

Solitary Nodule on the Proximal Nail Fold

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A 62-year-old man presented for evaluation of a slowly growing, nonpainful nodule on the first proximal toenail fold of the right foot of 6 years' duration. He reported that the nail plate of the affected toe was thickened and malaligned. He denied a history of trauma. Physical examination revealed a 2.0×1.6-cm, flesh-colored, nontender, well-defined, rubbery nodule with prominent overlying tortuous telangiectases on the medial aspect of the first proximal toenail fold of the right foot. The associated nail plate was yellow, thickened, and angled laterally into the second toe. Radiograph of the right hallux identified a soft tissue density contiguous with the dorsal aspect of the distal portion of the phalanx. There was no evidence of bony involvement. A shave saucerization biopsy specimen was obtained and sent for hematoxylin and eosin and immunohistochemical staining. The spindle cells were diffusely positive for CD34.

WHAT'S THE DIAGNOSIS?

- dermatofibroma
- dermatofibrosarcoma protuberans
- infantile digital fibromatosis
- neurolimmoma
- superficial acral fibromyxoma

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The authors report no conflict of interest.

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THE DIAGNOSIS: Superficial Acral Fibromyxoma

A shave biopsy revealed an uninvolved grenz zone and mildly cellular spindle cell dermal proliferation in a collagenous and myxoid background (Figure 1). Spindle cells were seen in a myxoid background among dense coarse collagen (Figure 2A). Spindle cells also were seen in a myxoid background with mast cells and capillary network (Figure 2B). Histopathologic examination of the biopsy specimen revealed spindle cells that were diffusely positive for CD34 (Figure 3); focally positive for epithelial membrane antigen; and negative for melanocytic markers, smooth muscle markers, and cytokeratin. A diagnosis of superficial acral fibromyxoma (SAFM) was made based on clinical, histopathologic, and immunohistochemical findings.

Superficial acral fibromyxomas, also known as digital fibromyxomas, are soft, slow-growing tumors that have a predilection for subungual or periungual regions of the hands and feet. Superficial acral fibromyxomas most frequently occur on the hallux and rarely occur on the ankle or leg. They can present as nodular, dome-shaped, polyploid, or verrucous masses. They can be soft to firm, gelatinous or solid, off-white to gray-white and can have fasciculate cut surfaces. Superficial acral fibromyxomas can be either painful or painless and present with a deformed nail in 9% of cases. Superficial acral fibromyxoma is a superficial lesion with frequent infiltration of the dermal collagen and subcutaneous tissue and may even erode or infiltrate into the underlying bone in rare cases.¹⁻⁴ Although SAFMs are rare tumors, documented

cases of SAFM have been reported at an increasing rate since the first published report by Fetsch et al² in 2001.

Patients often delay seeking medical treatment and present with a solitary mass that has been slowly growing for months to years. In a study of 124 patients, Hollmann et al¹ found that symptoms exist for a mean of 35 months and present with a small mass with a mean tumor size of 1.7 cm before biopsy or excision. Although the age range is broad, SAFM mostly affects middle-aged adults (median age, 49 years).¹ Hollmann et al¹ also reported a male predominance (1.3:1 ratio), and preexisting local trauma is reported in 25% of cases.²⁻⁴

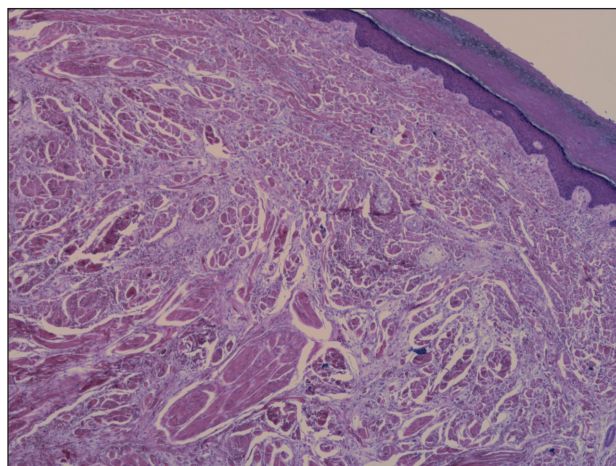


FIGURE 1. Superficial acral fibromyxoma with an uninvolved grenz zone and mildly cellular spindle cell dermal proliferation in a collagenous and myxoid background (H&E, original magnification $\times 40$).

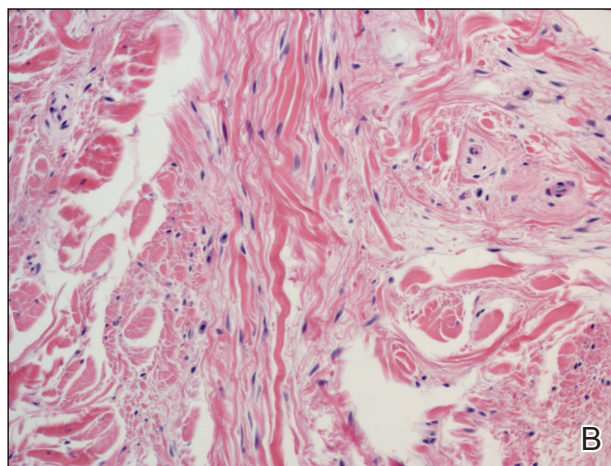
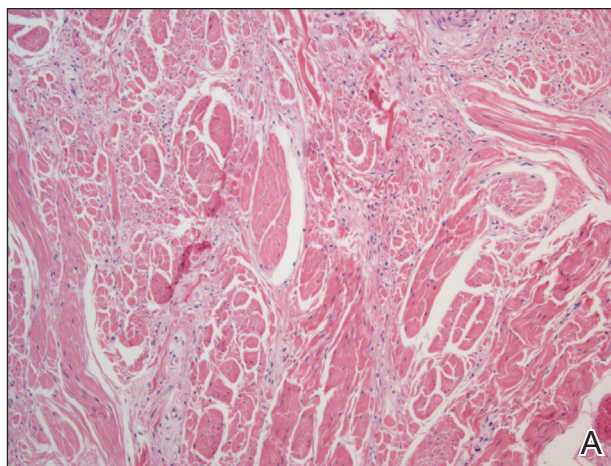


FIGURE 2. Superficial acral fibromyxoma with spindle cells seen in a myxoid background among dense coarse collagen (A) as well as with mast cells and capillary network (B)(H&E, original magnifications $\times 200$ and $\times 400$).

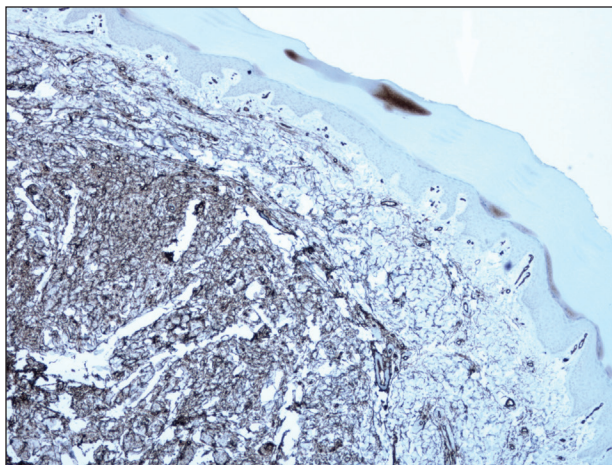


FIGURE 3. Superficial acral fibromyxoma immunohistochemical staining showed spindle cells that were diffusely positive for CD34 (original magnification $\times 40$).

The differential for SAFM should include dermatofibroma, keloid, dermatofibrosarcoma protuberans, acquired digital fibrokeratoma, infantile digital fibromatosis, neurolemmoma, sclerosing perineurioma, superficial angiomyxoma, low-grade fibromyxoid sarcoma, and acral myxoinflammatory fibroblastic sarcoma.¹⁻⁴

Superficial acral fibromyxomas are composed of CD34⁺ spindle or stellate-shaped cells that are embedded in a myxoid and/or dense hyalinized collagenous stroma in a random or loosely fascicular growth pattern. The spindle or stellate-shaped cells in SAFMs also have

been found to be focally positive for epithelial membrane antigen and CD99. Lesions have accentuated microvasculature and increased mast cells.⁵⁻⁸

Conservative management is reasonable, but patients presenting with persistent pain and/or local deformity should be definitively treated with complete excision and follow-up. Hollmann et al¹ found that 24% of tumors recurred locally upon incomplete excision after a mean interval of 27 months. All recurrent tumors had positive margins at excision or initial biopsy.¹ To date, no reports of tumors metastasizing have been documented.¹⁻⁴

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