

# Toe Nodule Obliterating the Nail Bed

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A generally healthy 30-year-old man presented with a 3-cm exophytic, yellowish red, subungual nodule of the left great toe of 1 year's duration that was obliterating the nail plate. Ten years prior, a similar nodule in the same location was removed via laser by a podiatrist. Medical records were not retrievable, but the patient reported that he was told the excised lesion was a benign tumor. Plain radiographs were performed at the current presentation and demonstrated an inferior cortical lucency of the distal phalanx as well as a lucency over the nail bed region with extension of calcification to the soft tissues. Magnetic resonance imaging showed a mass with a proximal to distal maximum dimension of 2.1 cm that involved the dorsal surface of the proximal phalanx. Magnetic resonance imaging also demonstrated bone erosion from the overlying mass. A 4-mm incisional punch biopsy was performed prior to surgical excision.

## What's the diagnosis?

- a. dermatofibroma
- b. dermatofibrosarcoma protuberans
- c. neurofibroma
- d. superficial acral fibromyxoma
- e. superficial angiomyxoma

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## The Diagnosis: Superficial Acral Fibromyxoma

Superficial acral fibromyxoma (SAF) was first described in 2001 by Fetsch et al.<sup>1</sup> Subsequently, the term *digital fibromyxoma* was proposed in 2012 by Hollmann et al<sup>2</sup> to describe a distinctive, slow-growing, soft-tissue tumor with a predilection for the periungual or subungual regions of the fingers and toes. The benign growth typically presents as a painless or tender nodule in middle-aged adults with a slight male predominance (1.3:1 ratio).<sup>1,2</sup> In a case series (N=124) described by Hollmann et al,<sup>2</sup> 9 of 25 patients (36%) who had imaging studies showed bone involvement by an erosive or lytic lesion. Reports of SAF with bone involvement also have been described in the radiologic and orthopedic surgery literature.<sup>3,4</sup> Radiographically, the soft-tissue invasion of the bone is demonstrated by scalloping on plain radiographs (Figure 1).<sup>3</sup>

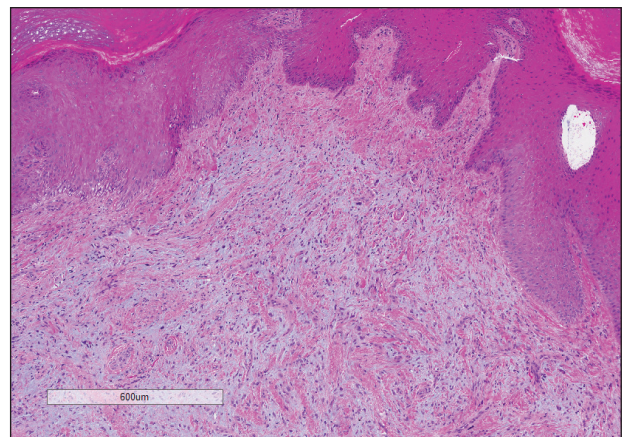
Histologically, SAFs are moderately cellular with spindled or stellate fibroblastlike cells within a myxoid or collagenous matrix (Figure 2).<sup>1</sup> The vasculature is mildly accentuated and an increase in mast cells usually is observed. The nuclei have a low degree of atypia with few mitotic figures, and the stellate cells exhibit positive immunohistochemical staining for CD34 (Figure 3), epithelial membrane antigen, and CD99.<sup>1</sup> Hollmann et al<sup>2</sup> found that 66 of 95 tumors (69.5%) infiltrated the dermal collagen, 26 (27.4%) infiltrated fat, and 3 (3.2%)

invaded bone. Of the 47 cases that were evaluated on follow-up, 10 tumors (21.3%) recurred locally (all near the nail unit of the fingers or toes) after a mean interval of 27 months. Although invasion of underlying tissues and recurrence of the tumor has been demonstrated, this growth is considered benign. The histologic differential diagnosis includes neurofibroma, myxoma, fibroma, low-grade fibromyxoid sarcoma, dermatofibroma, superficial angiomyxoma, and dermatofibrosarcoma protuberans.<sup>2</sup>

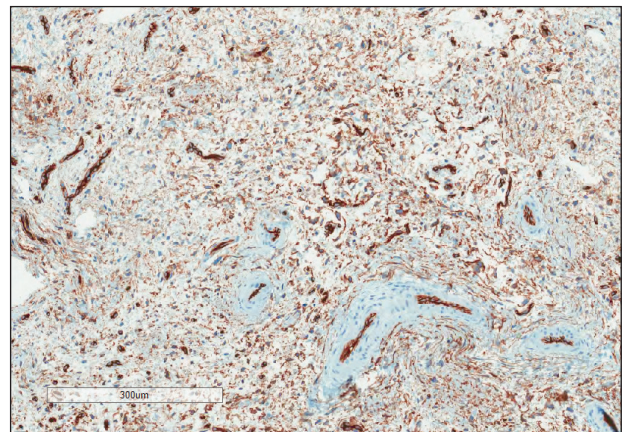
The primary treatment of SAF is local excision. The incidence of local recurrence found in the case series by Hollmann et al<sup>2</sup> was directly linked to positive margins after the first excision (10/47 [21.3%])



**Figure 1.** Plain radiographic evidence of inferior cortical lucency on the left great toe.



**Figure 2.** Benign epidermis with a low-grade myxoid dermis (H&E, original magnification  $\times 4$ ). Reference bar denotes 600  $\mu\text{m}$ .



**Figure 3.** Higher magnification of the myxoid stroma and CD34 positivity (original magnification  $\times 7.4$ ). Reference bar denotes 300  $\mu\text{m}$ .

recurrent lesions had positive margins). To date, there are no known reports of metastatic disease in SAF.<sup>2</sup> Our case manifested with a late recurrence of the tumor and bone involvement requiring surgical excision, which illustrates the role of adjuvant imaging and close follow-up following excision of any soft-tissue tumors of the fingers and toes that have been histologically confirmed as SAF, particularly those of the periungual region.

## REFERENCES

1. 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;32:704-714.
2. Hollmann TJ, Bovée JV, Fletcher CD. Digital fibromyxoma (superficial acral fibromyxoma): a detailed characterization of 124 cases. *Am J Surg Pathol.* 2012;36:789-798.
3. Varikatt W, Soper J, Simmon G, et al. Superficial acral fibromyxoma: a report of two cases with radiological findings. *Skeletal Radiol.* 2008;37:499-503.
4. Oteo-Alvaro A, Meizoso T, Scarpellini A, et al. Superficial acral fibromyxoma of the toe, with erosion of the distal phalanx. a clinical report. *Arch Orthop Trauma Surg.* 2008;128:271-274.