Superficial Acral Fibromyxoma on the Thumb-Nail Bed

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ABSTRACT

Superficial acral fibromyxoma, also known as digital fibromyxoma, is a benign soft tissue tumor. The acral regions, including the palms, soles, fingers, toes, and nail units, are the commonly affected locations. The subungual region of the great toe is the most common site reported in current literature. The tumor is slowly progressive and benign in nature. Histology commonly reveals a fibromyxoid neoplasm with immunoreactivity to CD34 and CD99 markers.[1,2,3] We present the case of a 39-year-old female with a nine-year history of repetitive digital trauma presenting with superficial acral fibromyxoma of the thumb-nail bed. Our case is unique due to the tumor location and the patient's prior long history of trauma to the tumor site.

KEYWORDS: digit, fibromyxoma, nail, neoplasm, superficial acral fibromyxoma

CASE REPORT

A 39-year-old woman presented with a painful, slowly progressive growth on her right thumb-nail bed. She reported a nine-year history of repetitive trauma to the thumb from both physical and chemical factors. On physical examination, a skin-colored nodular growth was present on the right thumb-nail bed with overlying nail plate fragility and onycholysis (**Figure 1**). A nail-bed biopsy was conducted to rule out malignancy as well as to provide therapeutic relief to the patient.

The histopathology and clinical findings confirmed the diagnosis of superficial acral fibromyxoma (SAF). Histopathology showed a dermal proliferation of spindle cells embedded in a myxoid stroma with scattered mast cells. (**Figure 2**). There was no significant cytologic atypia. An Alcian-blue stain highlighted markedly increased mucin. Immunostains for CD34 and CD99 showed spotty positivity (**Figures 3,4**). An immunostain for SOX-10 was negative. Epithelial membrane antigen was negative. An immunostain for Ki-67 showed no significant increase in the dermal proliferation index.

Figure 1. Nodular growth with overlying nail plate fragility and onycholysis on the right thumb-nail bed.

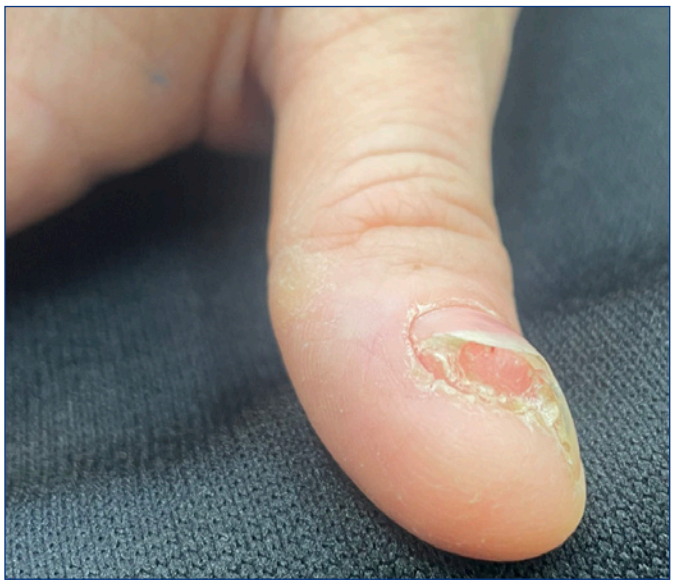


Figure 2. A dermal proliferation of spindle cells embedded in a myxoid stroma with scattered mast cells is present on tissue pathology (H&E, original magnification X 40).

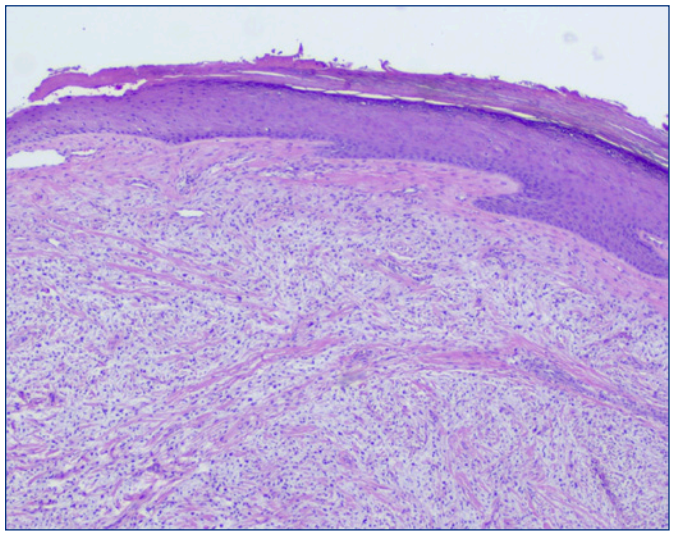


Figure 3. Tumor cells demonstrated spotty positivity for CD34 staining (original magnification X 200).

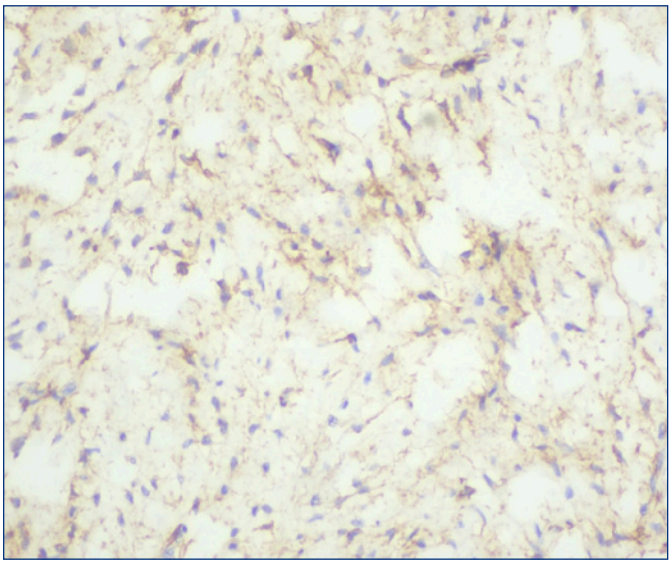
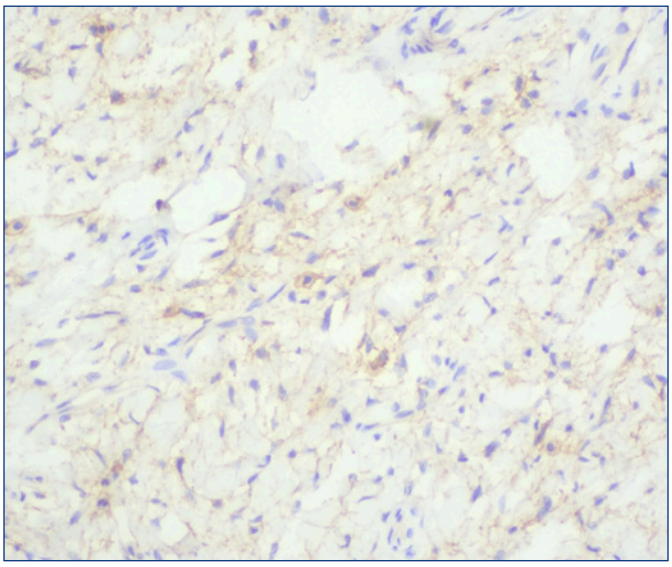


Figure 4. Tumor cells demonstrated spotty positivity for CD99 staining (original magnification X 200).



DISCUSSION

SAF is a rare benign soft tissue tumor first described in 2001 by John Fetsch et al.[4] There are currently 314 reported cases in the literature.[5] It presents as a pink to flesh-colored nodule that is slow-growing and asymptomatic.[1,2] SAF is located most commonly on the acral regions, mainly the toes (45.8%) and fingers (39.1%).[5] This neoplasm occurs most commonly in men (male:female ratio of 2:1) in their 40s (range = 4–91 years, mean = 47 years).[4,5] The growth is dome-shaped, verrucous, or polypoid, and the mean size of these lesions is 1.75 cm in diameter (range = 0.5 to 5.0 cm).[6] Dermatoscopy may show yellow-white hyperkeratotic areas, white and red structureless areas, small arborizing vessels, and vascular structures at the periphery of the lesion.[1,2]

Due to the nonspecific features and rarity of SAF, it is difficult to diagnose clinically. Malignant processes in the differential diagnosis include squamous cell carcinoma, subungual melanoma, and rarely, dermatofibrosarcoma protuberans and myxofibrosarcoma. Benign lesions in the differential diagnosis include acquired digital fibrokeratoma, periungual fibroma, neurofibroma, glomus tumor, sclerosing perineurioma, osteoma, or subungual exostosis. Biopsy and histopathology findings are necessary to confirm the diagnosis.[1,2,3]

Histology of SAF may show a fibromyxoid neoplasm covered by hyperkeratosis or an epithelial collarette. In the dermis, elongated, spindle- or star-shaped neoplastic cells with a myxoid stroma without atypia are seen.[4] Mast cells are usually present. Tumor cells are typically positive for CD34, epithelial membrane antigen, and CD99. Tumor cells are usually negative for S100, desmin, CK40, and CK48. Immunoreactivity to these immunohistochemical markers has been reported to vary among lesions, with the strongest historical reactivity to CD34 and CD99 markers.[2,3]

Imaging studies may be useful in the work-up of superficial acral fibromyxoma as bone involvement has been reported in 36% of cases.[6] Plain radiography may show underlying bone erosions and scalloping.[4] Magnetic resonance imaging with T2-signal may show hyperintensity, consistent with myxoid content. Fine needle aspiration will show a cluster of loose spindle cells in myxoid material.[1,2]

Management of SAF is with complete surgical excision but with no specific margin guidelines in the literature other than obtaining a clear histological margin.[7] Recurrence of the tumor is rare if initial margins are negative. However, recurrence is approximately 20–25% in cases where excision had initially positive margins. The average time of local recurrence is 27 months.[6] Follow-up after excision is recommended and recurrences can be managed with re-excision.[4,5] Mohs surgery has been proposed as an alternative treatment for greater control of margins and to reduce the risk of recurrence.[1-3,7]

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