Painful subungual nodule

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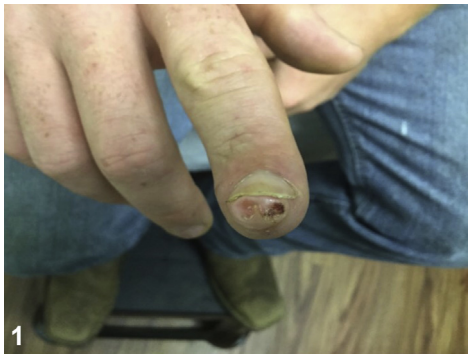
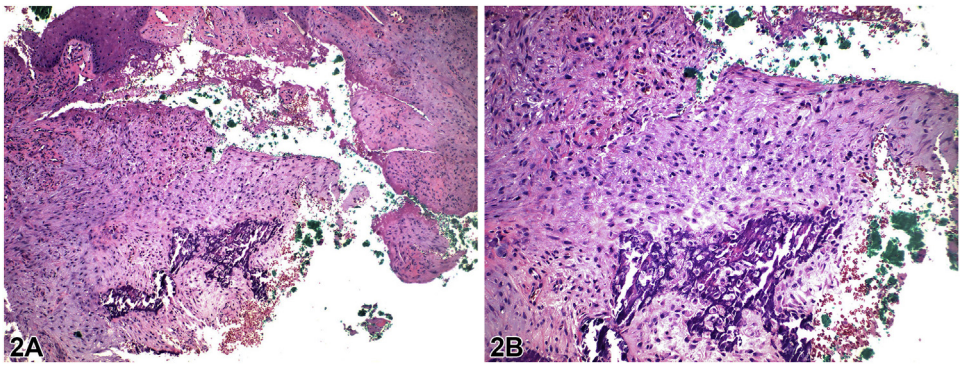
Key words: cartilaginous metaplasia; nails; subungual tumor; superficial acral fibromyxoma.





CASE PRESENTATION

A 22-year-old man with no significant medical history presented with a hard, subungual nodule on his right distal index finger that had been slowly enlarging for 7 years. He complained that the lesion was tender particularly while working with his hands. The patient denied any trauma to the area prior to the lesion appearing. The lesion was unsuccessfully treated by a primary care physician with liquid nitrogen. On physical examination, the lesion appeared as a 1.3-cm hard, traumatized, skin-colored subungual nodule involving the right distal index finger (Fig 1). A punch biopsy was obtained with hematoxylin-eosin staining (Fig 2).

Question 1: What is the most likely diagnosis?

A. Subungual osteochondroma

B. Phalangeal chondrosarcoma

C. Superficial acral fibromyxoma (SAFM)

D. Subungual exostosis

E. Subungual keratoacanthoma

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Funding sources: None.
Conflicts of interest: None disclosed.

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JAAD Case Reports 2020;6:531-3.
2352-5126

https://doi.org/10.1016/j.jdcr.2020.03.019

Answers:

A. Subungual osteochondroma — Incorrect. Subungual osteochondromas are rare tumors affecting the distal fingers or toes. Histologically, they consist of a central area of mature bone surrounded by a cartilaginous capsule. Early lesions may only show cartilaginous changes, as ossification has not yet occurred; however, an abundance of spindle cells is not seen in osteochondromas.

B. Phalangeal chondrosarcoma — Incorrect. Chondrosarcomas are malignant tumors of bone that rarely metastasize but may require extensive surgical resection. Histologically, these tumors will feature encasement of cartilage with new reactive bone and mild-to-severe nuclear atypia, which are not apparent in the slides of this case.

C. SAFM — Correct. SAFM is a benign fibrotic neoplasm classically presenting as a pink-to-flesh–colored nodule commonly affecting the periungual and subungual regions with insidious onset.[1-3] Although pre-existing trauma to the affected site has been reported in some cases, the causative pathophysiology has yet to be determined.[2,3] Histologic findings of a SAFM, as seen in our case, involve a nonencapsulated tumor comprised of proliferative spindle-shaped cells embedded in a variably myxoid dermis.[1-3] Accurate histologic diagnosis of this lesion is imperative, as malignant mimickers often require more extensive surgical resection or adjuvant therapy.

D. Subungual exostosis — Incorrect. Subungual exostosis is a benign exophytic bony proliferation affecting the distal digits. Histologically, they will present with a fibrocartilage cap but will not have proliferations of spindle-shaped fibroblasts as seen in Fig 2.

E. Subungual keratoacanthoma — Incorrect. Subungual keratoacanthomas present as rapidly progressive, painful subungual nodules showing well differentiated squamous epithelium with a central core of keratin on histology. These findings are not present in our case.

Question 2: Which of the following is a unique histologic finding for our case as seen in Fig 2?

A. Necrosis

B. Psammomatous calcification

C. High number of mitotic figures

D. Tendinous ossification

E. Cartilaginous metaplasia

Answers:

A. Necrosis — Incorrect. Necrosis presents with pale-staining collagen caused by degradation and cellular injury often with surrounding granulomatous inflammation. These findings are not present in our specimen but can frequently be seen in epithelioid sarcomas.

B. Psammomatous calcification — Incorrect. On histology, psammomatous calcification will appear as concentric lamellated calcification commonly seen in meningiomas and papillary thyroid carcinoma. Although rare, this finding may also be present in some mesenchymal tumors.

C. High number of mitotic figures — Incorrect. The histologic specimen does not appear to have a high degree of mitotic activity or pleomorphism, indicating a more benign process. This finding would be more characteristic for an epithelioid sarcoma or phalangeal chondrosarcoma given the location.

D. Tendinous ossification — Incorrect. Despite the presence of a cartilaginous matrix, there is no sign of bone formation or tendon involvement within the specimen.

E. Cartilaginous metaplasia — Correct. To our knowledge, SAFM involving cartilaginous metaplasia, as seen in Fig 2, *A* and *B*, has been reported only twice in the literature prior to the present case.[3,4] Chondrocytes are apparent within lacunae, but the absence of a cartilaginous capsule helps rule out osteochondromas and subungual exostosis.

Question 3: What is the optimal treatment for this lesion?

A. Monthly intralesional steroid injections

B. Complete surgical removal of the lesion

C. Intralesional methotrexate injection

D. Observation with close follow-up

E. Sclerotherapy

Answers:

A. Monthly intralesional steroid injections — Incorrect. There is currently no evidence showing resolution of SAFM with intralesional steroid injections.

B. Complete surgical removal of the lesion — Correct. Complete surgical excision often provides definitive management of SAFM; however, recurrences

can occasionally occur. In a previously published retrospective study, 24% of SAFM recurred over a mean follow-up period of 27 months. All recurrences in this study showed positive margins on initial biopsy or excision and were near the nail unit.[3] Although rare, malignant transformation has been reported in 2 cases, which emphasizes the need for complete surgical removal.[5]

C. Intralesional methotrexate injection — Incorrect. Intralesional methotrexate injections have been used for the treatment of keratoacanthomas. There is no evidence to support its use in treating SAFM.

D. Observation with close follow up — Incorrect. SAFM may become locally destructive with rare erosive or lytic bone involvement reported. Atypical cases of malignant transformation have also been documented.[5] For these reasons, conservative observation is not recommended.

E. Sclerotherapy — Incorrect. There has been no evidence to support the use of sclerotherapy in treating SAFM.

The authors thank James Patterson, MD and Mark Wick, MD for reviewing the slides of this case.

Abbreviation used:

SAFM: superficial acral fibromyxoma

REFERENCES

1. Cullen D, Diaz Recuero JL, Cullen R, Rodriguez Peralto JL, Kutzner H, Requena L. Superficial acral fibromyxoma: report of 13 cases with new immunohistochemical findings. *Am J Dermatopathol*. 2017;39(1):14-22.
2. Sawaya JL, Khachemoune A. Superficial acral fibromyxoma. *Int J Dermatol*. 2015;54(5):499-508.
3. 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.
4. Moulonguet I, Goettmann S, Zaraa I. Superficial acral fibromyxoma with cartilaginous metaplasia. *Am J Dermatopathol*. 2019;41(4):316-317.
5. Fetsch J, Laskin W, Miettinen M. Superificial 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(7):704-714.