A large growth on the toe

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CLINICAL FINDINGS

A 65-year-old man presented to the dermatology clinic with a lesion on his left little toe. (Fig. 1) He reports that the lesion first appeared more than 10 years ago and has been gradually increasing in size. It was largely asymptomatic, but he felt pain sometimes, resulting from its compression from the adjacent toe. He did not take any treat for the lesion in the past. On cutaneous examination there was a 4 x 2-cm skin-colored to erythematous firm plaque over the distal left little toe, impinging on the nail, with associated distal hyperkeratosis.

What is your clinical differential diagnosis?

Giant cell tumor of tendon sheath
Myxoid neurofibroma
Superficial acral fibromyxoma
Dermatofibrosarcoma protuberans

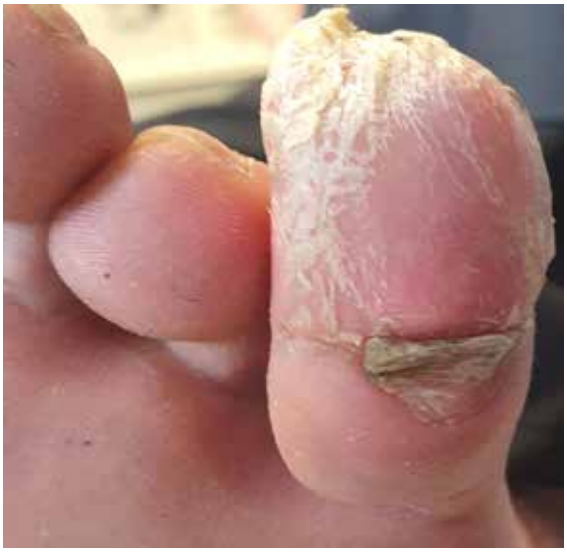

Fig. 1 A 4 x 2-cm mass on the left little toe

PATHOLOGICAL FINDINGS

A 4 mm punch biopsy was taken for histopathological analysis (Fig. 2). Histopathology revealed a dermal proliferation of CD 34+ spindle shaped cells within a fibrous and myxoid stroma.

DIAGNOSIS

Superficial acral fibromyxoma (SAF)

COMMENT

First described in 2001 by Fetsch et al, SAF is a benign myxoid tumor with a predilection for the periungual and subungual areas of the fingers and toes.[1] There is an approximately 2:1 male to female ratio, with a large variation in the age of onset, mostly middle-aged adults.[1-3] The mean age of diagnosis is 47 years and 7 months according to a recent review of the literature.[1-3]

The majority of tumors present as a solitary, slow-growing, painless mass with a gelatinous to firm consistency, and the size may vary between 0.5 and 5cm.[1] Other presentations may include pain

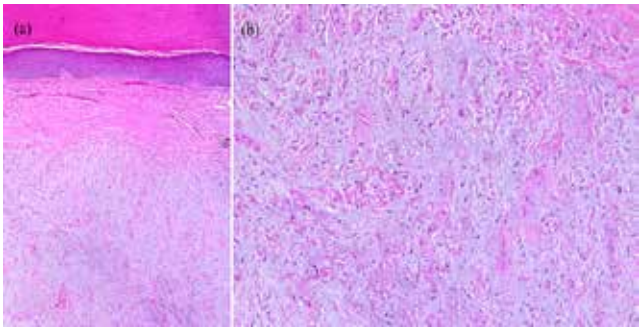

Fig. 2 Dermal proliferation of spindle shaped cells within a fibrous and myxoid stroma. (hematoxylin-eosin a. x100; b. x400)

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Table 1: The clinicopathological challenges of Superficial acral fibromyxoma.

Diagnosis	Clinical	Pathological
Giant cell tumor of tendon sheath	• It is a slow growing soft tissue mass that develops over a period of months to years. It is the second commonest tumor of the hand. Trauma, inflammation, metabolic disease and a neoplastic etiology are considered as etiological factors.	• It is a well circumscribed nodule arising in the deep dermis or subcutis, comprised of a population of oval cells set in a minor condensed eosinophilic fibrous stroma. Scattered multinucleated giant cells are present. In most cases small clusters of lipid laden cells can be seen. It is CD68 and Vimentin positive, hinting a fibrohistiocytic origin
Myxoid neurofibroma	• A benign peripheral nerve sheath tumor, which is S-100 protein positive. The most common locations are the face, shoulders, arms, periungual and in the feet.	• Interlacing bundles of elongated cells with wavy darkly stained nuclei • Interspersed variably sized collagen bundles • Interspersed mast cells • Stroma contains variable mucin and collagen • CD34 positivity in cells of unclear histogenesis • S100 positivity in neural cells • Monomorphic "comma-shaped nuclei" • Divergent differentiation, including melanin pigmented cells, may occur rarely
Dermatofibrosarcoma protuberans	• Classically an exophytic, nodular cutaneous mass; however, often initially presents as a flat plaque Initially may show persistent slow growth, often for many years, then sudden progression • Progression may occur with fibrosarcomatous transformation.	• Generally centered in the dermis or subcutis and characterized by spindle cells with a storiform to whorled pattern. • The cytoplasm is generally abundant and eosinophilic; the nuclei are monomorphic and ovoid to elongated with variable mitotic activity. • Tumors infiltrate and expand fibrous septa; interdigitation amongst lobules of fat yields a so called "honeycomb" pattern. • Adnexal structures typically spared. • The stroma may be myxoid, collagenous, or microcystic.

(41%) and less commonly nail deformity, bleeding, or infection, as well as locations like the legs, hands, and ankles.[1-3] This tumor is difficult to diagnose and delays in its diagnosis may occur due to its indolent nature, low morbidity, and the common histopathologic characteristics it shares with other pathologies.[1-3]

On histology, SAF appears as a well-circumscribed unencapsulated dermal tumor, generally without an intraepidermal component. The tumor can also extend down into the subcutaneous tissue. Cytologically, the cells of the tumor appear spindle-shaped or stellate-shaped. These cells are usually immunoreactive for CD34, EMA, and CD99. Cellular atypia is not common, but it has been described.[1-3] The stroma surrounding these cells exhibits both fibrous and myxoid components, and one may predominate the other.[1-3]

Radiologic studies, such as simple radiography, ultrasonography and nuclear magnetic resonance (NMR), may provide additional diagnostic and prognostic information, especially regarding bone

involvement which has been reported in around 36% of cases.[1-3] Malignancy arising from the tumor has not been reported in the literature, but the potential for malignancy is not clear due to cytologic atypia in a small number of cases.[1-4]

The differential diagnosis of SAF includes various fibromyxoid/myxoid-appearing proliferations or acral tumors.[2-5] Myxoid dermatofibrosarcoma protuberans (DFSP) does not commonly affect acral sites and clues favoring its diagnosis include its storiform configuration and subcutaneous tissue infiltration.[4] Myxoid neurofibroma can be easily distinguished by its S100-immunopositivity.[4] While also showing mucinous areas, digital myxoid pseudocyst typically exhibits much lower cellularity compared to SAF.[4] While also presenting as an slowly growing acral nodule, giant cell tumor of the tendon sheath (GCTTS) can be distinguished histologically by its cellular composition of monocytic polyhedral stromal cells and multinucleated osteoclast-like giant cells.[5]

Management of SAF usually consists of surgical excision; however, local recurrence may occur in up to 24% of cases, especially when there are positive margins.[1-3] Hence, regular follow-up is required after excision. Also, Moh's micrographic surgery may provide therapeutic relief while decreasing the rate of recurrence and preserving functions of the digit and nail apparatus.

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