

Superficial Acral Fibromyxoma 2nd Toe: A Rare Entity

Saumya Gupta ^{1#}, Shreya Bhardwaj ¹, Pratibha ², Navneet Mishra ³, Pranav Kumar Dave ^{4*}, and Vandana Agrawal ⁵

¹ Resident Surgery, L N Medical College and J K Hospital, Kolar Road, Bhopal-462042, MP, India.

² Assistant Professor Surgery, L N Medical College and J K Hospital, Kolar Road, Bhopal-462042, MP, India.

³ Professor Surgery, L N Medical College and J K Hospital, Kolar Road, Bhopal-462042, MP, India.

⁴ Professor Radiology, L N Medical College and J K Hospital, Kolar Road, Bhopal-462042, MP, India.

⁵ Professor & HOD, Pathology, L N Medical College and J K Hospital, Kolar Road, Bhopal-462042, MP, India.

Refers to Main Author.

* Refers to Corresponding Author.

***Corresponding Author:** Pranav Kumar Dave, Prof. Department of Radiology, E-7/540, MIG Senior, Arera Colony, Bhopal-462016, MP, India.

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Abstract

Superficial Acral Fibromyxoma is a benign and rare tumor of the soft tissues. It's also called digital fibromyxoma and was first described by Fetsch in 2001, as a localized tumour in the acral extremities. Superficial acral fibromyxoma often presents on either the fingers or toes, and has been shown to involve the nail in 97% of fingers, and 96% of toes [1]. Incidence of acral fibromyxoma is not noted in the literature, and because of an unusual diagnosis, surgeons should be aware of this tumor, which requires complete surgical excision and follow up for recurrence. Since they are generally painless, intervention is late and a history of trauma is described in several cases.

Keywords: superficial acral fibromyxoma; 2nd toe; rare tumor; digital fibromyxoma; nodule; cytology; spindle cells; DIP; fascicles; nail

Introduction

Superficial Acral Fibromyxoma is a benign and rare tumor of the soft tissues. It's also called digital fibromyxoma and was first described by Fetsch in 2001, as a localized tumour in the acral extremities. Superficial acral fibromyxoma often presents on either the fingers or toes, and has been shown to involve the nail in 97% of fingers, and 96% of toes [1]. Incidence of acral fibromyxoma is not noted in the literature, and because of an unusual diagnosis, surgeons should be aware of this tumor, which requires complete surgical excision and follow up for recurrence. Since they are generally painless, intervention is late and a history of trauma is described in several cases.

Case Report

A 48-year-old healthy man presented with an asymptomatic, slowly growing nodule with a 15-year history on the tip of his second right toe. Examinations revealed a hemisphere, irregular-shaped growth, measuring 3.0 cm × 2.3 cm × 1.5 cm in size, on the tip of his second right toe (Fig 1). The nail was not visible and the nail bed was distorted. X ray right leg AP and lateral view was suggestive of excess of soft tissue components distally in the 2nd toe, bones and joints appeared normal (Fig 2). The USG local site was suggestive of lobulated echogenic mass lesion in the descending phalanx of the 2nd toe. Cytology was suggestive of scattered benign oval to spindle cells with no evidence of malignant cells. The growth was completely excised, with DIP disarticulation (Fig 3).



Figure 1(a).



Figure 1(b).



Figure 2



Figure 3

Histopathologically, the section showed dermal and subcutaneous tumour composed of bland spindle and stellate cells within myxoid and collagenous stroma. Scattered small calibre blood vessels and mast

cells are seen. Cells are arranged in a random loose pattern and in focal areas forming short fascicles. No mitosis was seen. No nuclear atypia was noted (Fig 4).



Figure 4

Discussion

Superficial acral fibromyxoma is a benign and slow-growing solitary soft-tissue neoplasm [2]. The often-asymptomatic presentation of acral fibromyxomas means that there is often a delay in their diagnosis and subsequent treatment [3]. SAFM, a rare slow growing myxoid tumor in the subungual area that was first described in 2001 [4]. Clinically, neurofibroma, dermatofibrosarcoma protuberans (DFSP), low grade fibromyxoid sarcoma, and myxofibrosarcoma should all be considered in the differential diagnosis and evaluation of SAF [5]. It is also called digital fibromyxoma. It has a slight male predominance. The mean age of the patients falls in the 5th decade with patients ranging from 4 to 86 years of age. About 25% of cases recur [1,6].

It usually manifests itself through a painless mass of slow growth that affects mainly males in the fifth decade of life. It usually affects the distal region,

with a polypoid or dome-shaped appearance [7]. The differential diagnosis of superficial acral fibromyxoma includes fibroma of the tendon sheath, myxoid neurofibroma, glomus tumor, giant cell tumor of the tendon sheath, sclerosing perineuroma, acral fibrokeratoma, cutaneous myxoma, myxoinflammatory acral fibroblastic sarcoma, fibrous histiocytoma, and dermatofibrosarcoma protuberans [8].

Conclusion

In conclusion, if a soft tissue tumor occurs under the nail, we should suspect superficial acral fibromyxoma and we also should keep in mind that such tumors can grow aggressively. Awareness of this entity is helpful in distinguishing SAF from other myxoid soft tissue tumors occurring there. Complete excision with clear resection margins is the mainstay of treatment [9].

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