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Case Report

A Rare Occurrence of Digital Fibromyxoma to the Hallux Distal Phalanx

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Abstract

A paucity of literature exists for superficial acral fibromyxoma, which typically affects the acral regions of the hands and feet. Because of its indolent nature and clinical mimicry of other ungual and periungual tumors, delays in diagnosis are common. This report aims to augment our understanding of this pathology and improve recognition for prompt diagnosis, as well as provide an addition to the literature.

Keywords: *Digital Fibromyxoma; Hallux Distal Phalanx; Superficial Acral Fibromyxoma*

Introduction

Superficial acral fibromyxoma or digital fibromyxoma, first described by Fetsch., *et al*. is a rare benign tumor commonly found on the periungual and subungual regions of fingers and toes [1]. Although primarily localized to upper and lower extremities, these tumors can be found on the palms, heels, or ankles albeit infrequent. There is a higher predilection in men and age of onset is approximately 45 - 50 years old [2]. The lesions present as flesh-colored, solitary or erythematous papules usually smaller than five centimeters. Nail bed involvement is seen in 50% of all cases; however, there are no current reports of invasion beyond the mid-reticular dermis, and less frequently, reports of prior trauma to the affected region [2,3].

Radiographically, these tumors can cause erosive changes to the underlying bone, however, penetration to bone is unlikely [4]. On MRI, these tumors are hypertensive on T2 and STIR and isointense to muscle on T1. Correct diagnosis is contingent on appropriate clinical evaluation, advanced imaging and immunohistological staining analysis [4]. Histologically, these lesions appear as fibrous, well-circumscribed myxoid tissue composed of mucin and gelatinous material, often spindle-form, ovoid or stellate-shaped cells within a myxoid stroma [5].

Differential diagnoses include superficial acral fibromyxoma, digital fibrokeratoma, sarcoma, superficial angiomyxoma, chondroid tumor and myxoid neurofibroma. If the tumor is noted to be painless and slow-growing, the diagnosis is likely superficial acral fibromyxoma [6]. To further differentiate between digital fibromyxoma and other ungual masses, microscopic evaluation is required via

sampling/excision of the tumor. Immunohistological staining demonstrates CD34-positive, SMA-positive, and S100-negative cells in cases of superficial acral fibromyxoma [5].

The treatment of choice is wide surgical excision. Digital fibromyxoma is benign in nature, however even with wide resection, rate of recurrence remains relatively high ranging from 10% to 25% [7]. Therefore, it is essential to follow up with patients for a minimum 12 - 20 months to determine need for further treatment [8].

Case Report

This case study looks at a 72-year-old female who presented initially with complaints of a slow-growing, soft tissue mass to the left great toe of one year duration. Although mostly painless, she reported discomfort with direct pressure and detailed occasional clear fluid drainage from the area. She reported scratching the lesion but was advised not to due to her diabetes. The patient had past medical history significant for Type II diabetes mellitus, hypothyroidism, and breast cancer in remission.

On clinical evaluation, the skin lesion appeared raised, well-circumscribed, firm, and nodular with a serous fluid-filled proximal border, measuring approximately 2.8 x 2 x 1 cm. There was evidence of concomitant nail dystrophy although no invasive properties were noted (Figure 1).

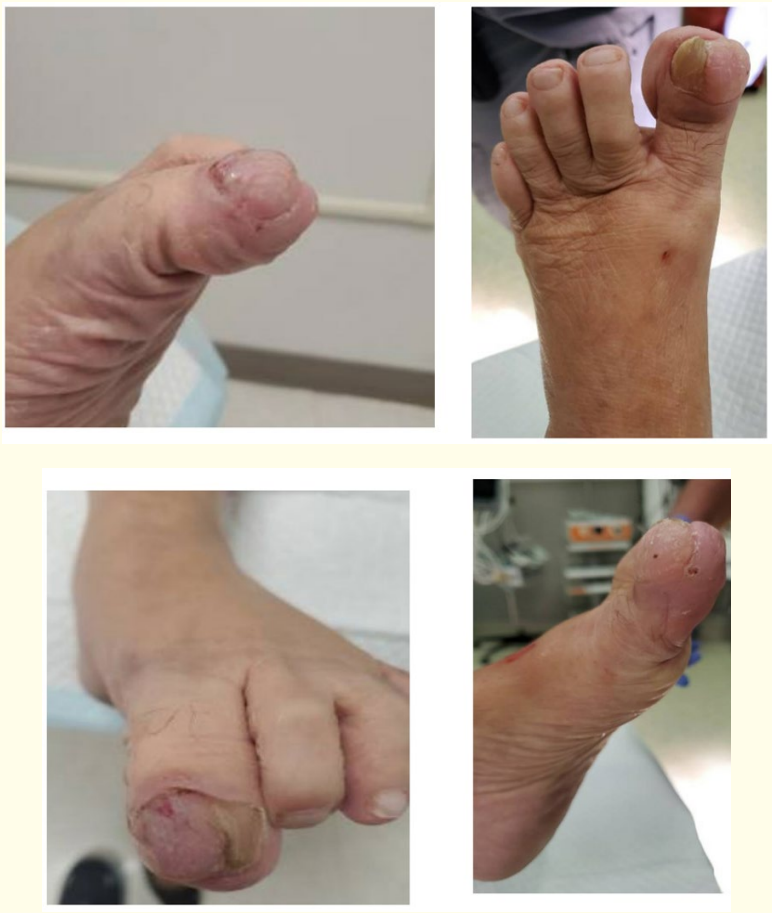
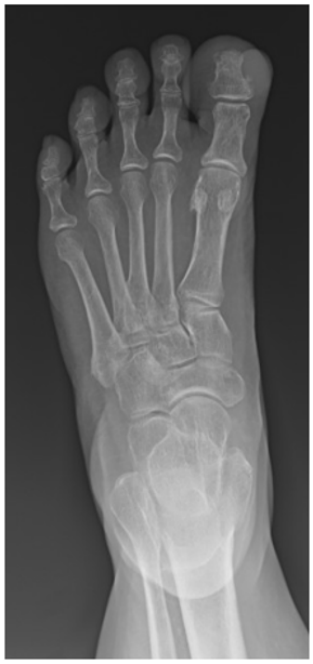
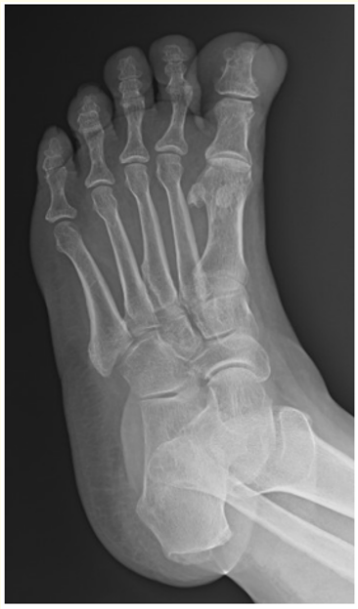


Figure 1a-1d: *Clinical presentation of digital fibromyxoma to the left hallux.*

Radiographic evaluation was performed to assess the underlying deformity and potential osseous involvement. The plain films demonstrated a well-circumscribed, dorsal hallux soft tissue mass without osseous invasion or other evidence of osseous fracture or dislocation (Figure 2 and 3). Advanced imaging with MRI evaluation was additionally performed for surgical planning to assess margins for resection. The MRI results demonstrated a solid enhancing round mass at the dorsal medial distal great toe possibly arising from the nail bed or adjacent soft tissue structures measuring 1.2 x 1.9 x 1.8 cm. The mass appeared isointense to muscle on T1 and hyperintense on T2 with uniform enhancement consistent with a solid tumor. Additionally, there was no evidence of osseous erosions which was suggestive of an indolent process (Figure 4 and 5).





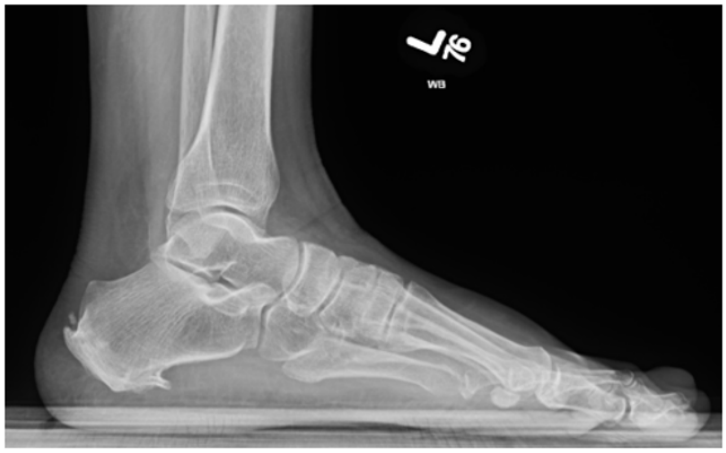
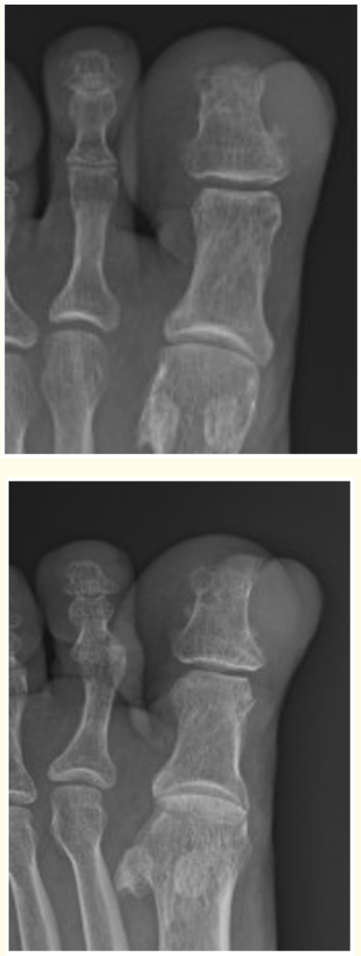


Figure 2: *Anteroposterior, medial oblique, and lateral radiographs demonstrating a soft tissue lesion to the left hallux.*



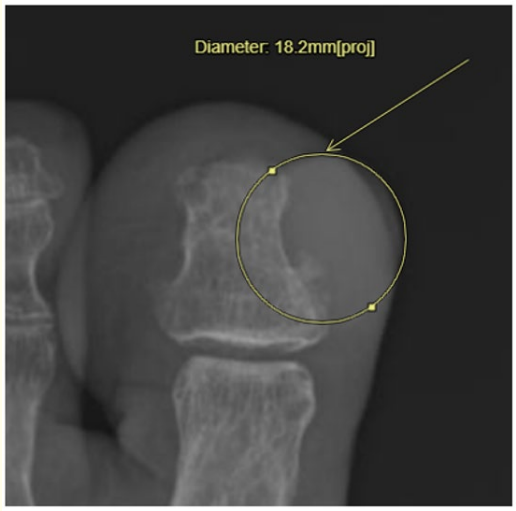
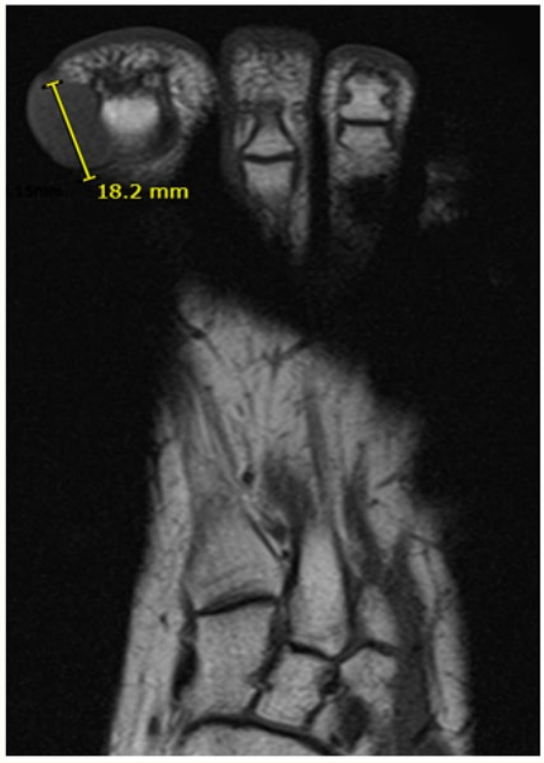


Figure 3: *Radiographs of the left hallux demonstrating soft tissue mass to the medal distal phalanx with no underlying osseous involvement.*



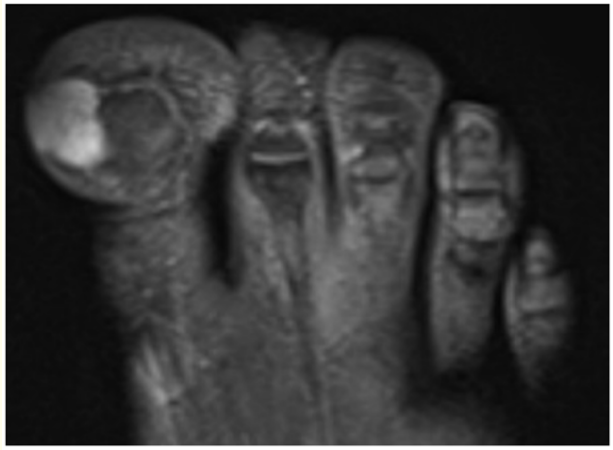


Figure 4a and 4b: *Coronal views of the lesion on MRI, T1 and T2-weighted respectively.*

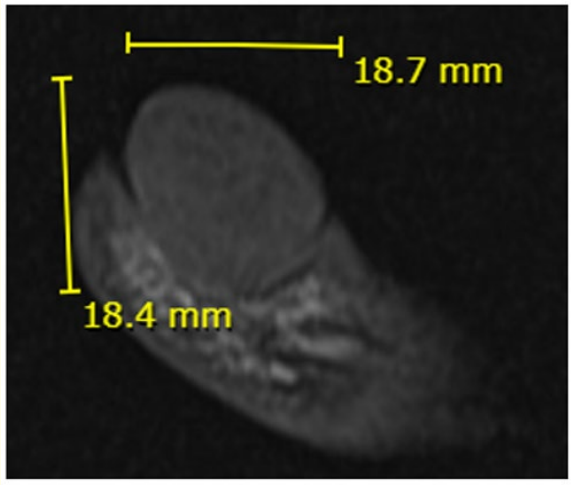
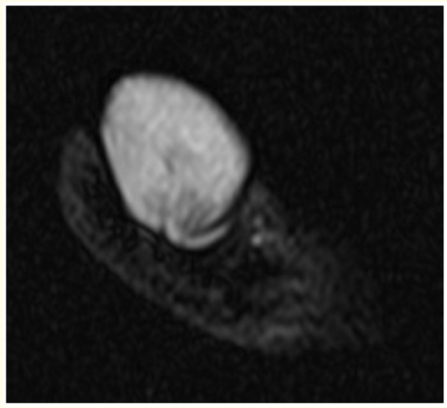


Figure 4c and 4d: *Sagittal views of the lesion on MRI.*



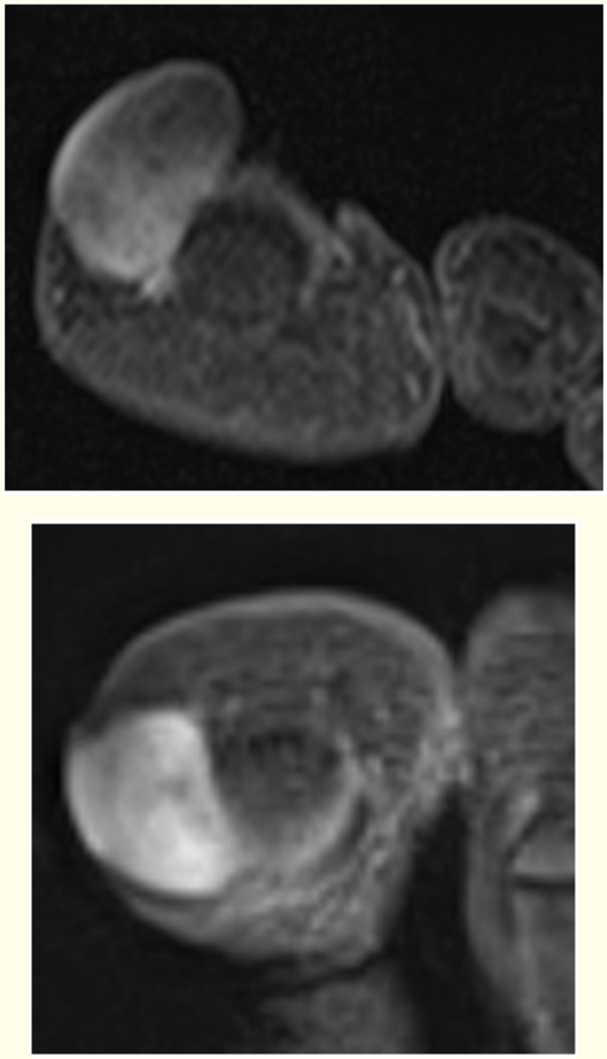


Figure 5a-5c: *Sagittal, axial and coronal views of the soft tissue mass, respectively, on MRI.*

At this time, the patient underwent surgical resection including a left foot soft tissue mass excision, skin biopsy, and partial phalangectomy (Figure 6). A partial hallux amputation was advised to limit infection risk, capture clear margin, limit postoperative wound complications due to the gross size of the mass as well as the patient's uncontrolled diabetes and treat already damaged nail. The patient elected against amputation at the interphalangeal joint and worked on her diabetes to a controlled level. Surgical pathology demonstrated a 2.0 x 1.7 x 1.2 cm tan-white and gritty to myxoid nodule consistent with a digital fibromyxoma. Immunostains with appropriate controls were performed with lesional cells positive for CD34 and SMA but negative for S100, consistent with digital fibromyxoma. The patient had an unremarkable postoperative course with no recurrence to date.

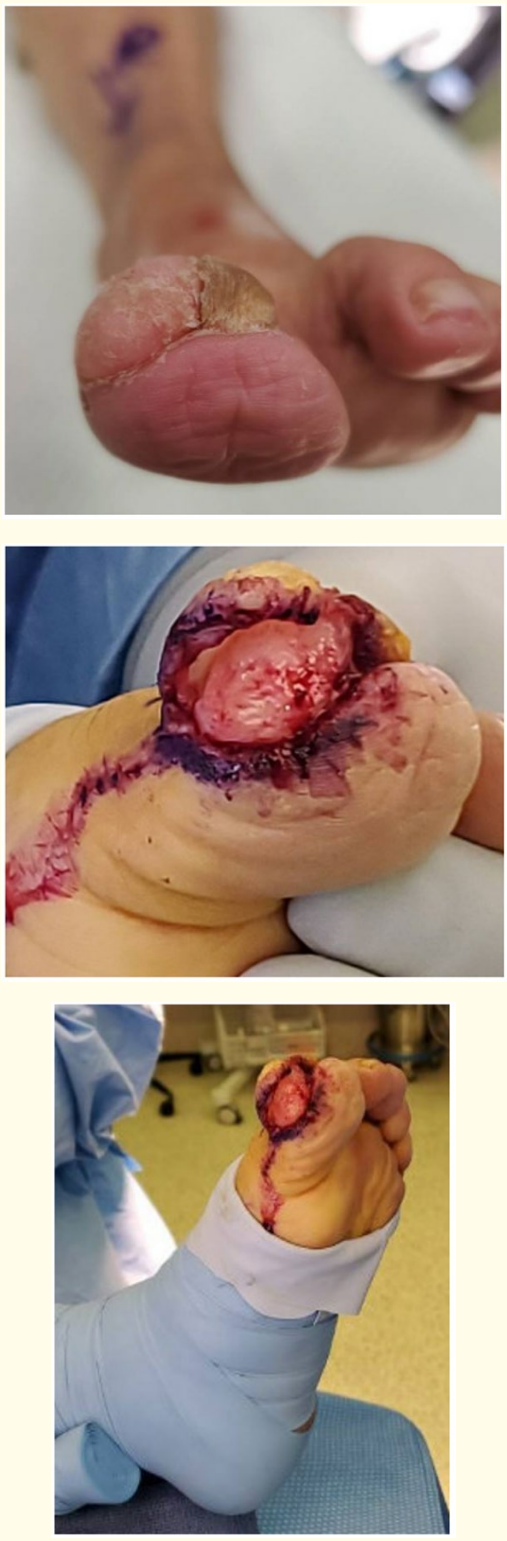


Figure 6a-6c: *Intraoperative wide resection of the lesion.*

Discussion and Conclusion

Superficial acral fibromyxoma is a relatively rare benign tumor commonly found on the acral regions of the upper and lower extremities with just over 300 reported cases thus far [9]. Although there can be several differential diagnoses, certain unique clinical and histological distinctions can help differentiate it from other periungual neoplasms. In conclusion, digital fibromyxoma is a rare condition that should be considered in cases with painless, ungual or periungual, slow-growing lesions. Wide surgical resection is often the treatment of choice; however, clinicians should remain vigilant postoperatively as recurrence rates are high.

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