Case Report

Superficial acral fibromyxoma: A case report

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ABSTRACT

There are a wide variety of benign and malignant soft tissue tumors. Digital superficial acral fibromyxoma is a rare, benign, expansive soft tissue growth which often involves the distal aspect of the digit and the nail plate. While it is often asymptomatic in nature, it may cause a great deal of distress due to involvement of the nail and the resultant cosmetic deformity. Unlike wide excision or amputation needed for more aggressive tumor types, local excision is often curative. We review the literature and describe the presentation of this unique pathology to our clinic in an otherwise healthy 33-year-old female.

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1. Introduction

Superficial acral fibromyxoma (SAF), also known as digital fibromyxoma, was first described by Fetch et al. during a retrospective review of archived pathology from the Armed Forces Institute of Pathology from 1970 to 2000.¹ They identified 37 cases, with the mass typically presenting on the fingers and toes. All patients presented with a solitary mass ranging from 0.6 to 5.0 cm in size, and 67.6% of patients were male.¹ Clinically, SAF generally presents on the distal aspect of the hands or feet with a polypoid or dome shaped appearance.¹ It is slow growing and has been reported to present between 3 months and 30 years after the patient first noticed the mass.² While the delay in presentation is generally due to its relatively asymptomatic course, the main complaint in up to 41% of patients is associated pain.³

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.³ Imaging will reveal lytic lesion of the bone in 36% of cases.³ There are no known risk factors for SAF and it has been reported in patients ranging in age from 4 to 86.^{3,4} Due to its relatively general presentation and lack of any known risk factors definitive diagnosis is typically made via pathological examination. Histologically it appears as a non-encapsulated soft tissue tumor of the dermis with spindle shaped fibroblasts in a storiform or fasciculated pattern.^{1,3} Nuclear atypia and mitotic figures are

uncommon due to the benign nature of SAF.² Additionally it has a myxoid collagenous stroma which frequently contains mast cells.¹ On immunohistochemistry SAF is commonly positive for CD34, and CD99 with variable positivity for epithelial membrane antigen (EMA).^{1,3} Importantly it is negative for S100 aside from one reported case from Fetsch et al., distinguishing it from other pathology such as neurofibromas.¹

Despite the evidence of specific immunohistochemical markers SAF may still get confused with other types of soft tissue pathology of the hand. Hollmann et al. reviewed the pathological findings in 124 cases of SAF submitted to them for review from other pathologists and found that only 2/124 clinicians correctly identified the mass with some of the most common incorrect diagnoses being: soft tissue tumor (12), fibroma (7), ganglion (5), cyst (5), and granuloma (5).³ Submitting pathologists did not fare much better, with only 4/124 pathologists correctly identifying the tumor as SAF. The most common incorrect diagnoses were: neurofibroma (18), myxoma (8), fibroma (7), low grade fibromyxoid sarcoma (6), dermatofibroma (6), dermatofibrosarcoma protuberans (5).³ The high incidence of misdiagnosis by both clinicians and pathologists reveals the need for the appropriate suspicion in making the diagnosis of SAF. This will allow for both clinicians and pathologists to ensure the correct immunohistochemical stains are used in the evaluation of the tissue sample.

2. Case report

A 33 year old otherwise healthy female presented to hand clinic with an enlarging mass on the distal aspect of her thumb for the last year. She denied any pain but did complain of cold sensitivity

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Fig. 1. T2 weighted image showing hyperintense soft tissue mass with some erosion of the distal phalanx.

and noted a cosmetic deformity of her thumbnail. She reported normal sensation in the digit. The physical exam was significant for tenderness over the mass, which measured approximately $1.5\text{ cm} \times 1.5\text{ cm}$ and protruded from the radial aspect of the thumb. There was no drainage, fluctuance, or sign of infection. Her IP joint was restricted in its range of motion. The patient was sent for MRI evaluation (Fig. 1) which showed a lobular hyperintense soft tissue mass on T2 measuring $1.9\text{ cm} \times 1.7\text{ cm} \times 1.3\text{ cm}$ with some erosion of the distal phalanx. At this point diagnosis was unclear and plan was made for nail removal and biopsy to determine diagnosis prior to final surgical planning (Fig. 2). Biopsy

Fig. 2. Clinical photo of the patients thumb mass following nail remove and prior to biopsy.

Fig. 3. (A) Preoperative image of the patients thumb. (B) Intraoperative photo showing development of plane between germinal matrix and the soft tissue defect following debulking. (C) Debulked specimen. (D) Closure of the thumb.

Fig. 4. Clinical appearance 2 months postoperatively shows good cosmetic appearance of the thumb as well as some regrowth of the nail.

revealed superficial acral fibromyxoma and definitive debulking was performed with the goal of maintaining the nail plate in order to allow for future nail regrowth. Intra-operatively it was possible to dissect a plane between the normal germinal matrix and tumor (Fig. 3). At 2 month follow up the patient denied any cold intolerance or pain at the site of her tumor. She was non tender and was beginning to experience nail regrowth (Fig. 4).

3. Discussion

Superficial acral fibromyxoma is a rare entity that is often confused with other soft tissue tumors which may be more malignant in nature. Periungual fibroma or acquired digital fibrokeratoma may be initially diagnosed however they typically extend into the dermis and have a more cellular appearance on microscopy.^{5,6} Clinically neurofibroma, dermatofibrosarcoma protuberans (DFSP), low grade fibromyxoid sarcoma, and myxofibrosarcoma should all be considered in the differential diagnosis and evaluation of SAF.⁷ Neurofibroma is most similar in presentation to SAF however it displays S100 positivity on immunohistochemical

staining.⁴ Location can also help distinguish these various types of soft tissue masses. DFSP often presents on the trunk as opposed to the usual occurrence of SAF at the distal extremity. There has been one case of SAF on the palm.⁸ Also while both DFSP and SAF stain positive for CD34, DFSP will be negative to EMA.³ Low grade fibromyxoid sarcoma can be a more aggressive tumor and will present with a less organized growth pattern than SAF, often times infiltrating deeper into the soft tissue, and is less commonly seen on acral surfaces.⁷ Ultimately there are many soft tissue tumors of the extremity. It is important to consider SAF as the testing and treatment differ when compared to more aggressive soft tissue tumors.

The management of SAF consists of complete surgical excision. Hollman et al. reported that in the evaluation of 73 tumors 89% had positive margins, displaying the ability of the tumor to interdigitate with normal tissue.³ Despite inability to achieve negative margins with high success, recurrence has been reported to only be approximately 20–25% at a mean follow up of 27 months. Recurrence has only been reported in cases in which initial margins were positive.³ There has been one case of a second recurrence following re-excision.¹ Additionally there are no known cases in the literature of malignant transformation or metastasis of SAF.^{3,7} Given the non-aggressive nature and the low rate of recurrence, SAF can best be managed with local excision. While SAF is relatively uncommon, it is an important addition to the differential diagnosis of tumors of the digits, as successful identification can alleviate the need for more aggressive surgical treatment.

Conflicts of interest

The authors have none to declare.

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