Acral myxoinflammatory fibroblastic sarcoma: A case report and literature review

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Acral myxoinflammatory fibroplastic sarcoma is an extremely rare soft-tissue sarcoma. It typically presents as an inflammatory mass in the distal extremities of adult patients. The authors present a review of the available literature as well as a discussion on the surgical management of a patient with acral myxoinflammatory fibroplastic sarcoma who originally requested conservative management but ultimately required a two-digit ray amputation after local recurrence.

Le sarcome fibroblastique myxo-inflammatoire acral : rapport de cas et analyse bibliographique

Le sarcome fibroblastique myxo-inflammatoire acral est un sarcome des tissus mous d'une extrême rareté. En général, il prend la forme d'une masse inflammatoire des extrémités distales chez des patients adultes. Les auteurs présentent une analyse des publications et un exposé sur la prise en charge chirurgicale d'un patient atteint d'un sarcome fibroblastique myxo-inflammatoire acral qui a commencé par demander une prise en charge prudente, mais a finalement apté pour l'amputation de deux orteils après une récurrence locale.

Key Words: *Acral; Fibroblastic; Myxoinflammatory; Soft tissue sarcoma*

Soft-tissue sarcomas represent a group of anatomically and histologically diverse tumours. These tumours are relatively rare, accounting for approximately 1% of all adult malignancies (1,2). According to the American Cancer Society, the number of new cases of soft-tissue tumours in the United States was more than 11,000 in 2012 (3), with only an estimated 15% of these occurring in the upper extremity (4). Due to the scarcity of these tumours, few hand surgeons are well versed in their diagnosis and treatment. A certain degree of suspicion must be entertained because the clinical presentation of these tumours as a painless mass can mimic infection, ganglion or lipoma (5). Unfortunately, misdiagnosis can lead to inappropriate management or delay in treatment, with grave consequences including major amputation or death. We present a review of the current literature and a sample case of a distal extremity sarcoma presenting as a chronic inflammatory mass.

CASE PRESENTATION

The patient was a 33-year-old right-hand dominant female schoolteacher referred by her primary care provider for evaluation of an 'abscess of the right index finger'. The patient reported experiencing a spider bite to the area approximately two months previously, after which she developed a persistent mass in the finger that slowly increased in size. Although initially painless, the patient described sharp pain at the site that progressively worsened. No other constitutional symptoms were reported. She failed multiple attempts at conservative therapy, including antibiotics and steroids, provided by her primary care provider.

Physical examination revealed a soft tissue mass 2 cm in size on the ulnar aspect of the right index finger at the level of the proximal phalanx and adjacent to the web space (Figure 1A). The finger was neurovascularly intact and demonstrated normal range of motion and strength. Standard radiographic plain films of the hand revealed no evidence of fracture, foreign body or bony invasion. Given the atypical history, an incisional biopsy was offered but the patient declined, opting for excision.

The patient was subsequently taken to the operating room. The mass was identified subcutaneously at the ulnar border of the right index finger proximal phalanx and was found to invade the second web space. The mass exhibited multiple cyst formations and appeared to be a combination of fat, tissue necrosis and liquefaction (Figure 1B). The entire mass was dissected free, widely excised and sent to pathology.

Postoperatively, the patient did well and initial pathology revealed an unspecified neoplasm with myxoid features. The specimen was sent to two separate institutions including the Armed Forces Institute of Pathology (Washington DC, USA), where a diagnosis of acral myxoinflammatory fibroblastic sarcoma (AFMS) was confirmed. AFMS is a low-grade sarcoma known to be locally aggressive with rare distant metastases. A literature review yielded no standardized treatment protocol. In light of the potential for local aggressiveness, the patient was offered ray amputation but elected conservative therapy with close observation.

The patient returned 16 months later with palpable recurrence of the soft-tissue tumour at the ulnar aspect of the proximal second phalanx (Figure 1C). After consultation with multiple hand surgeons and oncologists, the patient agreed to the recommendation of ray amputation of the index and long fingers, which was performed without complications (Figure 1D). Pathology demonstrated recurrent AFMS. The patient has now been tumour free for more than five years postoperatively, with negative follow-up magnetic resonance imaging (Figures 1E and F).

DISCUSSION

AMFS is an extremely rare form of sarcoma first described in 1998 by Meis-Kindblom and Kindblom (6), and further characterized that same year (6-8). Since then, there have been a total of 21 articles published relating to cases of AMFS, with the majority of these from the pathology literature, with only two clinical orthopedic case reports (6-27). As such, the pathological characteristics are well described: histologically, the lesions are poorly circumscribed, multinodular tumours that often invade into adjacent structures including tendon sheaths, synovial lining, subcutaneous fat and muscle. The lesions contain various myxoid zones with poor cellularity and vascularity interspersed between solid tumour areas. The myxoid matrix has been described as containing numerous types of inflammatory cells, fibrosis

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and large, scattered, bizarre tumour cells with vesicular nuclei, prominent inclusion-like nucleoli and abundant eosinophilic cytoplasm with features of modified fibroblasts. The inflammatory infiltrate typically includes polymorphonuclear leukocytes, lymphocytes, granulocytes, eosinophils and plasma cells in addition to germinal centres.

The 21 articles describe 179 cases (6-27). There appears to be no predilection for a specific age group or sex. All of the tumours were localized exclusively to the extremities, with 65% of the cases occurring in the upper extremity. Of the tumours discovered in the upper extremity, 81% occurred in the fingers or hand. AMFS acted as a low-grade malignant sarcoma with a high rate of local recurrence and a typically low rate of metastasis.

In the original report by Meis-Kindblom and Kindblom (6), long-term follow-up was available for 36 patients diagnosed with AMFS and 24 of these (67%) expereinced local recurrence after excision. Approximately 50% of these recurrences were in multiple locations and 10 patients (28%) required amputation for definitive treatment after multiple recurrences. Local recurrences were significant in that they demonstrated an apparent neoplastic progression with an increase in atypical cells and extent of tumour cell clustering compared with the initial resections (6). In addition, there was a 6% metastasis rate in the initial report, with one case metastasizing to the inguinal lymph nodes 1.5 years after the initial resection while the second case developed pulmonary metastasis five years after resection. Of the other published reports, Sakaki et al (24) reported metastasis to a regional lymph node three months after resection in one patient, and Hassanein et al (14) reported two cases in which there were distant metastases, one to the lung and the other to the skull at three months to five years, respectively. One should interpret this seemingly low incidence of metastisis with some degree of caution because the majority of published data do not include follow-up beyond two years.

In their clinical case series, Lang et al (19) described patients initially being preoperatively diagnosed with various benign conditions including ganglion cyst, giant cell tumour of the tendon sheath and benign tumour of the hand. Three of their patients described a firm, painless mass noted incidentally while two patients described the mass as initially painless but becoming progressively more painful before seeking medical care. A similar presentation was noted by Leti Acciaro et al (20) in a subsequent report. These mimic the scenario in the present report, with the patient originally presenting with a diagnosis of chronic abscess and painless mass that became progressively more painful. Although the significance of the patient's report of a spider bite is unknown and may be unrelated, Meis-Kindblom and Kindblom (6) state that the intimate relationship with the synovium, the frequent association with tenosynovitis and the prominent inflammatory infiltrate suggest that inflammation plays a role in the pathogenesis of AMFS.

The histological differential diagnosis of AMFS is as broad as its clinical presentation. The variability can be associated with the myxoid, inflammatory and atypical features of AMFS. The differential diagnosis includes such conditions as tenosynovitis, giant cell tumour of tendon sheath, inflammatory myofibroblastic tumour, liposarcoma, epithelioid sarcoma and myxoid malignant fibrous histiocytoma (19).

Given that the majority of lesions in the hand are unlikely to harbour a malignancy, an incisional or core needle biopsy is an important consideration in the management of any lesion. Although routine biopsy does commit the patient to a second procedure for definitive management of what are normally benign lesions, it is imperative to discuss the consequences of a marginal excision with any patient who elects to omit this diagnostic procedure. Given the atypical history in the presented case, an incisional biopsy was offered and recommended to the patient, which she declined in favour of a single procedure. At this time, a thorough discussion regarding the aforementioned consequences was held with the patient, who stated understanding of the associated risks. Unfortunately, the discussion held with the patient did not explicitly include an option to convert the original resection into an incisional biopsy once the lesion had been visualized, which would have been a reasonable option given the atypical intraoperative appearance of the lesion. As highlighted by the present case, the patient's pathology dictated further excision, perhaps to a greater extent than what may have been necessary had a tissue diagnosis been ascertained before initial resection.

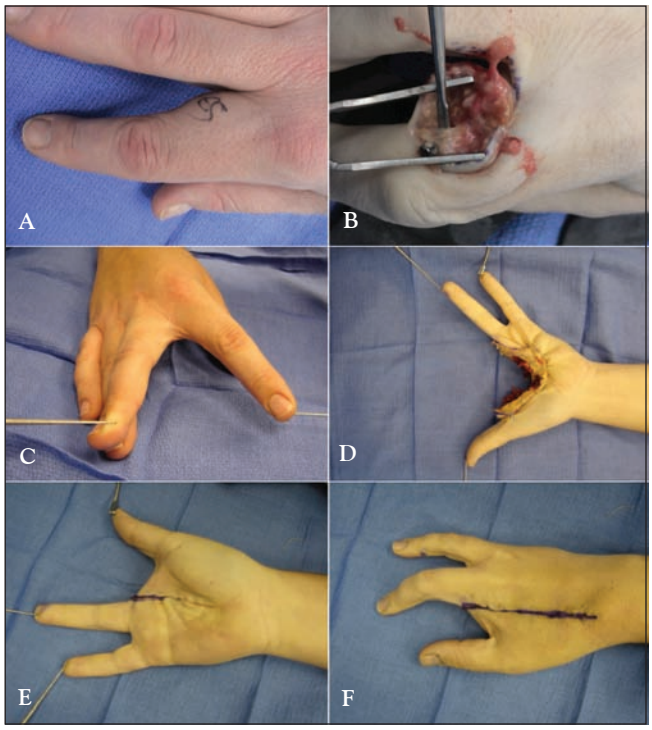

Figure 1) A *Appearance of the mass at initial presentation*. **B** *Appearance of the mass at initial excision. Notable for multiple cyst-like formations and areas of fat necrosis*. **C** *Appearance at subsequent presentation. The previous incision extends into the first web space*. **D** *Extent of the two-digit ray amputation to ensure resection of the entire web space*. **E** *and* **F** *Final appearance after closure of the amputation defect*

Although no formal standardized treatment protocol exists, the general low-grade nature of AMFS suggests that it should be treated with wide surgical excision because of its high rate of local recurrence (>50%). Although there is a paucity of literature from which to draw sufficient conclusions regarding the ideal extent of excision and anatomical structures that may be spared, there has not been any reported bone invasion, yet the lesion is commonly found to be attached to the underlying fascia and, in many cases, connected to tendons and ligaments, and rarely into muscle (5). The propensity to involve the dermis has also been well documented (1,2,5,6). In light of the lesion's potential to involve these surrounding structures, we cannot condone the sparing of anatomical structures within the planned field of excision. Close postoperative tumour surveillance also necessitates evaluating for the presence of local recurrence, regional nodal metastasis and the rare potential for distant metastasis including chest x-ray to evaluate for potential pulmonary metastasis, especially if physical examination findings, such as localized wheezing, are identified. More recently, at the Center for Sarcoma and Connective Tissue Oncology at Massachusetts General Hospital (Boston, USA), Tejwani et al (25) reported excellent local control with preoperative radiotherapy in 14 patients with only one recurrence in almost three years of follow-up. It must be cautioned, however, that these data were from a specialized sarcoma centre and the diagnosis of AMFS was known preoperatively, whereas in the majority of these cases, the final diagnosis is not known until postoperative pathological confirmation. In some patients, including our presented case, local amputation may be necessary for definitive treatment.

CONCLUSION

AMFS is an extremely rare sarcoma that often presents in the hands and fingers, and may easily be confused with many benign lesions. Because AMFS is a low-grade sarcoma with frequent local recurrence and the possibility of regional and distant metastasis, all surgeons involved in treatment of the hand should include AMFS in their differential diagnosis of slow-growing, painless masses to prevent misdiagnosis leading to inappropriate management or a delay in treatment that may have grave consequences.

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