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Case Report

Recurrent myxoid liposarcoma of the hand

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ABSTRACT

Malignant hand tumors are rarely observed in clinical practice which makes distinguishing them from benign soft tissue masses particularly challenging. A 41-year-old male presented with joint pain and swelling of his left index metacarpophalangeal joint. Radiological studies showed pronounced swelling around the affected joint with no bony abnormality, but incision and drainage of the site yielded negative cultures. Subsequent gross total resection confirmed the diagnosis of myxoid liposarcomas after cytogenetic pathology examination. Recurrence occurred after 1 year, followed by second resection and adjuvant radiotherapy. At 2 years follow-up, the patient was recurrence free. In conclusion, myxoid liposarcoma should be in the differential diagnosis of an older patient with erythema and swelling surrounding a subfascial mass of the hand.

Introduction

A facile understanding of the management of hand tumors requires knowledge of common and uncommon clinical etiologies of soft tissue masses of the hand [3]. Liposarcomas are a common subtype of soft tissue sarcomas with several histologic subtypes. The most common type is myxoid liposarcoma which has a greater propensity for the extremities. Malignant hand tumors are rarely observed in clinical practice, comprising just 2%-5% of all hand tumors [1]. In addition, there is also a paucity of literature describing them. To the best of our knowledge, less than 5 cases of myxoid liposarcoma presenting in the hand have been described in literature [2,3]. This case highlights the importance of having a high suspicion for these rare diagnoses in the differential for hand pathologies. Decreasing delays in diagnosis will lead to better outcomes for patients because proper management with imaging, surgical excision, and adjuvant therapies there is a low rate of local recurrence.

Case report

A 41-year-old right-hand-dominant African American male initially presented to the emergency department (ED) with swelling, pain, and erythema of the first metacarpophalangeal

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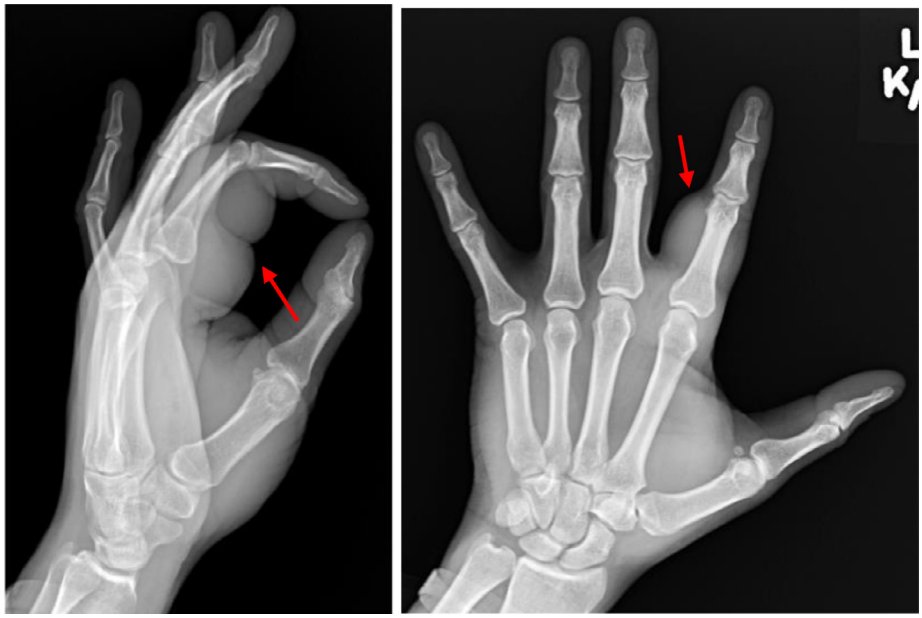

Fig. 1 – Initial radiographs of left hand with red arrows pointing to soft tissue swelling.

Fig. 2 – Intraoperative exposure of the tumor and final pathologic specimen.

(MCP) joint of the left hand. The patient reported that 10 years prior to presentation he suffered a laceration to the left index MCP joint in a work-related accident. The injury site healed with a "knot" (persistent mass), which he attributed to scarring, but the patient had full return of function in that area. For 2 months prior to presentation, the patient noticed rapid growth and extension of the mass from his left index MCP joint to the palmar aspect of his hand distally. Two weeks prior to admission, he developed progressive pain, edema, and induration of the joint, limiting his ability to grasp and make a fist. Examination revealed an erythematous, edematous, and tender left index MCP joint. There was a fluctuant mass extending from the palmar aspect of the left index MCP joint to the proximal phalanx. During this initial ED visit, an x-ray of the hand revealed no bony abnormalities (Fig. 1), and a soft tissue infection was suspected. Bedside incision and drainage was performed in the ED, which yielded scant purulence and mucinous gelatinous material. Microbiology studies revealed no growth at 24 hours. Given these suspicious findings, surgical exploration was performed a few days later which yielded gross total resection of a 6 × 8 cm tan-yellow multiloculated mass extending from the dorsum of the left index finger MCP joint to the proximal phalanx of the left index finger (Fig. 2). There was no involvement of the joint or tendons. Imaging and final pathology with fluorescence in situ hybridization analysis classified the tumor as stage IIIA myxoid liposarcoma measuring 7 × 5.5 × 2.1 cm, based on The American Joint Committee on Cancer. There was no round cell component to the tumor, and both macroscopic and microscopic excision margins were tumor free. Excellent cosmetic and functional outcomes were seen at regular postoperative clinic visits. The patient was then referred to a radiation oncologist which resulted in consensus decision for no further adjuvant therapy given the morbidity of radiation to the hand. Three months

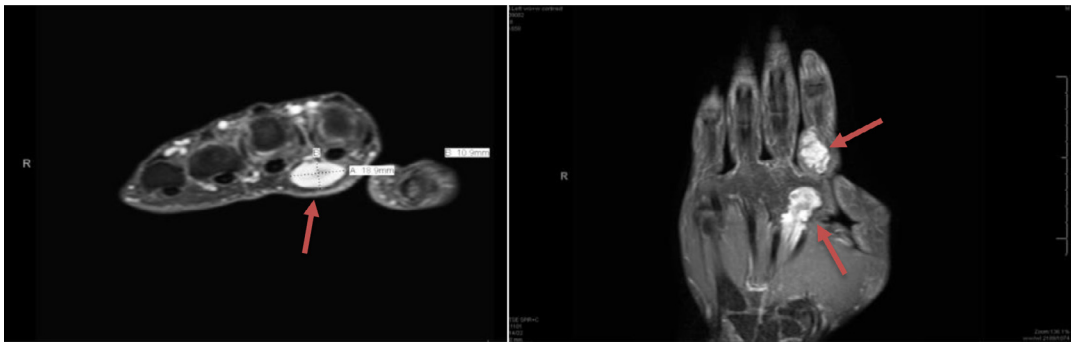

Fig. 3 – MRIs obtained after second presentation with red arrows pointing to the abnormal masses.

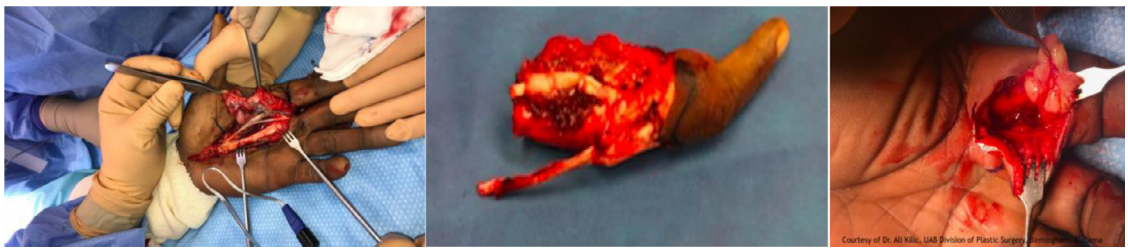

Fig. 4 – Intraoperative photos from reoperation for local tumor recurrence.

later, a PET scan was performed and showed no distant disease.

One year after the surgical removal of the tumor, the patient returned to clinic with complaints of increased swelling and pain exacerbated by flexion of the left index finger associated with paresthesia of the left index finger for the past month. An MRI revealed a 2 × 1.5 × 3 cm mass at the level of the proximal phalanx and a 1 × 2 × 5 cm mass at the level of the metacarpal of the left index finger which abuts the flexor digitorum superficialis, consistent with recurrence of the tumor (Fig. 3). Due to the proximity of the tumor to the flexor digitorum superficialis, the patient was scheduled for a ray amputation of the left index finger the following week (Fig. 4). Pathology revealed recurrent myxoid liposarcomas, measuring 3.5 cm with negative margins. The patient was then referred to radiation oncology for adjuvant local radiation to prevent morbid amputation due to tumor recurrence. Left hand MRI done 3 months after completion of adjuvant radiation revealed no evidence of macroscopic residual or recurrent malignancy within the left hand (Fig. 5). The patient has since complained of joint stiffness attributable to radiation therapy but endorses improvement with hand physical therapy. With 2 years of follow-up, he has had no additional local tumor recurrence and no evidence of distant disease per serial CT chest imaging.

Discussion

Liposarcomas are the second most common subtype of soft tissue sarcoma, comprising approximately 20% of soft tissue sarcomas. They have a higher incidence in males in their fifth decade of life [4]. Proposed etiologies for tumor development include genetic contributions, metabolic imbalances, and trauma [2]. They are found most frequently in the extremities, with a greater incidence in the lower extremities, particularly the thigh [5].

The 2013 World Health Organization guidelines defined 3 histologic subtypes of liposarcoma including: well differentiated, myxoid and/or round cell, pleomorphic, and dedifferentiated. Prognosis and treatment of liposarcoma are largely dependent on histologic classification, underscoring the need for early identification of subtype [5]. Well-differentiated liposarcoma is the most common histologic subtype. This subtype is a low-grade tumor with a large adipose component. Myxoid liposarcomas are the second most common subtype. Recently, the World Health Organization combined the myxoid and round cell subtypes of liposarcoma due to their association with the same gene fusions: FUS-DDIT3 or EWSR1-DDIT3. A round cell component of >5% within a myxoid liposarcoma has been associated with worse outcomes [5]. Myxoid liposarcomas are defined as well-circumscribed, multinodular masses, and gross histologic appearance depends on the degree of myxoid and round cell components. Myxoid liposarcomas involve the intermuscular space between 70% and 80% of the time vs intramuscular or subcutaneous locations [6]. The myxoid component of these tumors has a large mucopolysaccharide matrix with high water content, which contributes to the gelatinous appearance. This myxoid matrix contains well-circumscribed lobulated lipoblasts in varying states of differentiation in a plexiform vascular network.

As in the case of this patient, a myxoid liposarcoma has a more benign appearance on physical exam and may be mistaken for a soft tissue infection or other benign etiology. However, a subfascial mass greater than 5 cm in an extremity

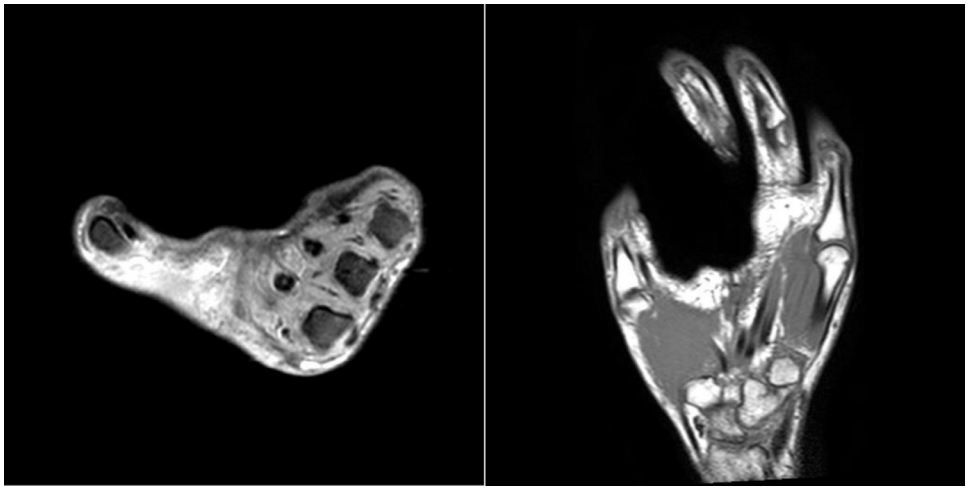

Fig. 5 – MRIs obtained after ray amputation of the left index finger and adjuvant radiation.

should raise suspicion and neoplastic etiology should be ruled out. As seen in this case, histopathology, cytogenetics, and imaging modalities are necessary for tumor staging, which dictates treatment. X-ray and MRI are reliable and often utilized methods of liposarcoma identification. MRI studies may also be used to subtype liposarcomas as they indicate the location and concentration of lipomatous components [6]. However, a recent study by Crombe et al indicated that homogeneous myxoid liposarcomas may be mistaken for cysts on MRI studies, which presents a challenge for accurate and early diagnosis [5].

Treatment of liposarcoma of the extremity without distant metastases usually consists of limb sparing surgery with neoadjuvant or adjuvant radiotherapy and/or chemotherapy. Indications for neoadjuvant or adjuvant therapies include large tumor size, involvement of the neurovascular bundle, and positive resection margins [7]. Additionally, prior analyses of myxoid subtype have found it to be the most chemosensitive and radiosensitive subtype of liposarcoma [8]. Well-differentiated and myxoid subtypes have a more favorable prognosis than other subtypes. The National Comprehensive Cancer Network recommendations for stage II-III myxoid liposarcoma include surgical resection with adjuvant radiotherapy. The lack of adjuvant radiotherapy in the patient's initial presentation and postoperative course may be responsible for the development of local recurrence. The local recurrence rate for myxoid liposarcoma following limb sparing therapy is approximately 9% [9].

Myxoid liposarcoma is a rare hand pathology but is important to consider when risk factors such as genetic contributions, metabolic imbalances, and trauma may be present [2]. Adjuvant radiation treatment is important for attenuating recurrent disease.

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