An uncommon case of metastatic myxofibrosarcoma presenting with elbow and hip masses

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Abstract

Myxofibrosarcoma frequently recurs locally but rarely metastasizes. Herein, we describe an elderly woman who had myxofibrosarcoma of the right elbow that went untreated during the COVID-19 pandemic. She subsequently presented with two large tumors ulcerating through the skin of her right elbow and left hip.

Keywords: cancer, dermatology, medical, metastasis, myxofibrosarcoma

Introduction

Myxofibrosarcoma (MFS) is an uncommon soft tissue tumor that typically presents in the proximal extremities as a painless mass [1]. It has a high local recurrence rate but seldom metastasizes [2]. We present a patient with advanced metastatic myxofibrosarcoma.

Case Synopsis

An 80-year-old woman presented to a mobile clinic with a chief complaint of large fungating masses of her right elbow and left hip. She had recently immigrated to the U.S. and the right elbow mass first presented in 2019 while she was living in Mexico. She underwent operations for the mass in 2019 and 2020. She also received systemic therapy prior to the COVID-19 pandemic but was unable to further describe what the therapy was. Unfortunately, she did not receive treatment during the pandemic and experienced growth of the tumor, development of a second mass, and weight loss of about 20 pounds over a one-year period. Furthermore, the left hip mass presented in January 2021 and began bleeding in May 2021, which prompted her presentation to clinic.

On examination she was tachycardic. A 12cm fungating mass was located on her right elbow, extending distally along the lateral forearm. It was nontender to palpation and bled easily on manipulation (**Figure 1**). The hip lesion had similar characteristics and was approximately 6cm in diameter (**Figure 2**).

She was promptly sent to the emergency department, where CT imaging of the chest, abdomen, and pelvis revealed the 6.2x2.7cm soft tissue mass of the left hip as well as possible para-

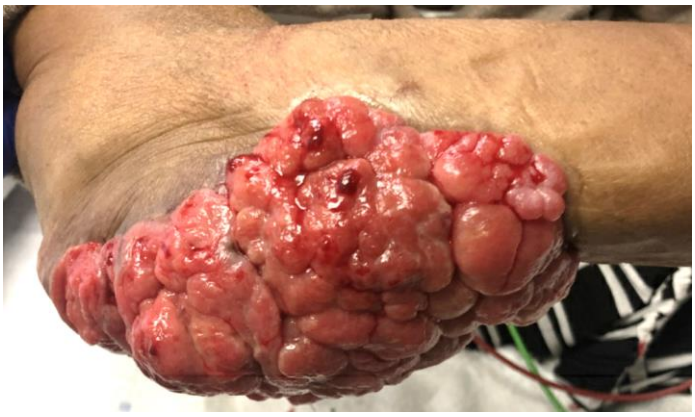

Figure 1. *A 12cm, nontender, fungating mass extended from the right elbow distally along the lateral forearm.*

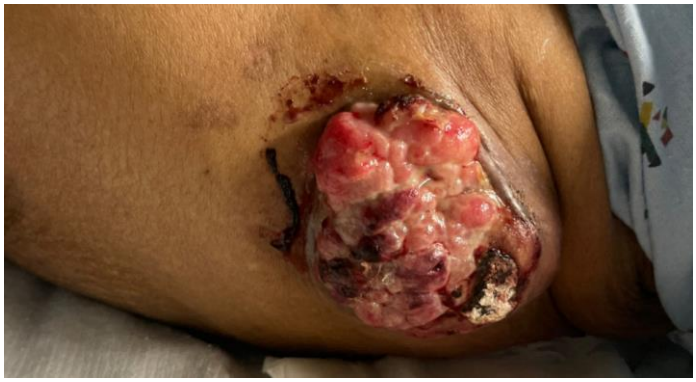

Figure 2. *A 6cm exophytic lesions was located on the left hip with similar characteristics to the elbow lesion.*

aortic, right axillary, and pulmonary metastatic lesions. Pulmonary imaging was obfuscated by concurrent COVID-19 infection. Subsequent 6mm punch biopsies of both lesions were consistent with high grade myxofibrosarcoma, showing pleomorphic spindle cells in a myxoid background with curvilinear vasculature as well as large cells with bizarre nuclei and extensive areas of necrosis (**Figure 3**). Both biopsy specimens were similar, except that the cells from the left hip were more mitotically active. Immunohistochemical staining was negative for cytokeratin AE1/3, CK903, low-molecular cytokeratins, S100, CD34, SMA, and desmin. The patient was urgently referred to the hematology/oncology department for further workup and treatment.

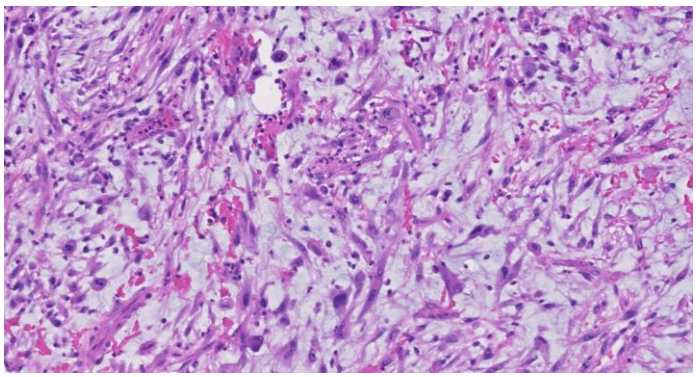

Figure 3. *Punch biopsy of the right elbow revealed pleomorphic spindle cells in a myxoid background with curvilinear vasculature. Large cells with bizarre nuclei were present as well as extensive areas of necrosis. Pathology of the lesion from the left hip was similar, with slightly more mitotic activity. H&E, 20×.*

Case Discussion

Myxofibrosarcoma is an uncommon soft tissue sarcoma, representing roughly 5% of soft tissue sarcomas [3]. Fibroblasts are the cell of origin and primary lesions have been described throughout the body [4]. Local recurrence is common, but metastatic disease usually does not occur [2]. When metastases are present, the lungs tend to be the most common site [2]. Like many sarcomas, hematogenous spread of MFS is the most common mechanism of metastasis when it does occur [5]. Our patient indicated that her arm lesion presented first and we propose that the mechanism of spread to the hip was hematogenous given the distance of the metastasis, which was not located in the same areas of lymphatic drainage as the first tumor.

Treatment for myxofibrosarcoma is often multi-faceted since lesions are highly recurrent after excision [2]. Chemotherapy regimens typically include alkylating agents, although recent advances in immunotherapy show promise [6]. Specifically, checkpoint inhibitors may be beneficial in patients whose tumors have acquired mutations from alkylating therapy [6]. Although the treatment approach is not yet fully defined for our patient, we anticipate surgical intervention followed by immunochemotherapy. Checkpoint inhibitor immunotherapy could be a consideration since our patient was previously treated with a systemic therapy.

Prognosis of myxofibrosarcoma is based on many factors including tumor spread, genetic markers, surgical margin, and tumor size, with the most important prognostic factor being the presence or absence of metastatic disease [7-9]. Positive surgical margin and CD44 expression are associated with the likelihood of metastasis [8]. Positive surgical margin and tumor size greater than 5cm are associated with local recurrence [8]. When metastases are present, the overall survival rate for soft tissue sarcomas drops to 20-30% [9].

Conclusion

Myxofibrosarcoma is uncommon and metastatic spread of MFS is particularly unusual. When present, hematogenous spread may lead to metastases in a variety of locations. Biopsy for grading and imaging for staging are important steps in determining the appropriate treatment strategy for patients who present with advanced MFS.

Potential conflicts of interest

The authors declare no conflicts of interest.

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