Exploration of Extraskeletal Myxoid Chondrosarcoma Within the Lower Extremity

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Statement of Purpose & Literature Review

Extraskeletal myxoid chondrosarcoma (EMC) is an extremely rare malignancy that possess unique morphologic, histologic, and immunohistochemical features. The aim of this case study is to discuss and review the management of EMC within the lower extremity.

EMC has been estimated to comprise 2.5% of soft tissue sarcomas and is characterized in most cases by a chromosomal translocation resulting in the EWSR1-CHN protein. The fusion gene products are generally responsible for alterations in cellular growth and differentiation. EMC is slow growing with 10-year survival rates ranging from 65% to 78%. To date, wide local excision is the only viable treatment option that offers patients a potential cure. Radiation in doses of up to 70 Gy has been reported to achieve substantial reduction in tumor size in individual patients, but this modality has not been associated with better outcomes in larger comparative studies.

Case Presentation

A 67 year old female with no significant past medical history presented to the podiatric office with concerns of a painful bump on the top of her left foot. According to the patient, the soft tissue mass had been present for a few months and had been increasing in size. Initial radiographs of the left foot were obtained which revealed no gross bony abnormalities. The MRI report revealed a prominent lobulated soft tissue mass in relation to the first metatarsal and first interspace.

Case Presentation

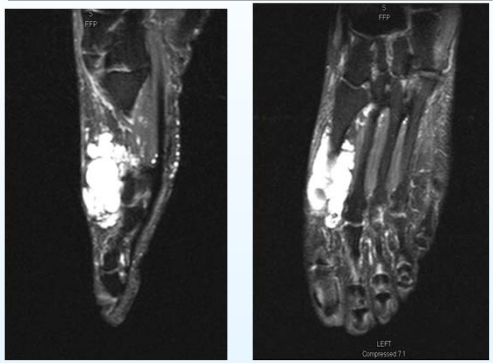

Figure 1. T2 Weighted Sagittal & Coronal MRI Images

The patient was taken to the operating room for excision of the mass. The specimen was evaluated by the pathologist and was also sent out for further evaluation. The pathology report revealed an extraskeletal myxoid chondrosarcoma, FNClCC Grade I with positive margins. The mass measured 4.9 x 3.6 x 1.0 cm and weighed 11 g. After these findings were reported, the patient underwent a PET CT whole body attenuation scan which resulted in a mild increased soft tissue density over the left foot between the hallux and the second digit. Upon reviewing the reports, the patient was then referred to an orthopedic oncologist.

The specialist took the patient back to the operating room for a second, more definitive procedure. An open Lisfranc amputation was performed along with the application of a bilayer bioengineered alternative tissue graft. Negative pressure therapy was also initiated over the graft site to assist with the wound healing process.

Discussion

Currently there are very limited case studies and research published on EMC, especially in the podiatric literature. The myxoid chondrosarcoma is a rare tumor, and according to the literature, only 2 % occur outside of the skeleton. An accurate diagnostic and therapeutic algorithm allows a precise preparation for surgery. Wide excision with negative margins, appears to be the appropriate initial approach for those diagnosed with this malignancy.

Even with adequate surgical margins, recurrence is reported to occur in about 25% of patients. Although the reports revealed the tumor to be located within the first interspace, the decision to perform a lisfranc amputation ensured that the resection was performed proximal enough. Despite a few reports of chemotherapeutic response, to our knowledge, there are no successful chemotherapeutic agents that effectively fight against EMC.

The use of radiotherapy for EMC is controversial, with some studies showing that these tumors are resistant to radiotherapy while other studies maintain that this is a radioresponsive tumor and radiotherapy should be considered. To date, the patient requires neither further surgery nor chemotherapy. At the six month follow-up period, the patient remained well healed from surgery and is cancer free.

Extraskeletal myxoid chondrosarcoma is a rare form of sarcoma which requires immediate attention and aggressive therapy. Without a surgical cure, patients will likely face poor outcomes. It is important as podiatric surgeons that we obtain proper imaging, perform biopsies and/or surgical resections of concerning lesions, and consult oncology whenever necessary.

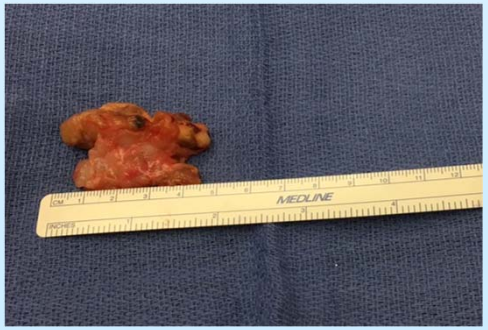

Figure 2. Intra-operative pathology specimen

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