Ewing sarcoma (ES) is a rare, malignant primary bone tumor commonly affecting children and young adults. The ES family of tumors consists of skeletal/extraskeletal Ewing sarcoma, neuroepithelioma, primitive neuroectodermal tumor, and Askin tumors. Despite being the second most common malignant bone tumor, ES rarely appears in extraskeletal locations. Extraskeletal ES is an exceedingly rare sarcoma, which typically occur in the paraspinal muscles, chest wall, gastrointestinal tract, and lower extremities. Compared to ES of the bone, extraskeletal ES is difficult to diagnose given it’s clinical presentation and various differential diagnoses relating to non specific imaging, histology and immunoprofile. The 10 year probability of survival rate is 91% which is attributed to localized disease and surgical resection. There is only 1 other published case study about extraskeletal ES in the foot or ankle. This case study aims to highlight a rare case report of extraskeletal ES progression in the foot with a functional foot salvage amputation procedure.

Case Report

10/2017: Twenty-two-year-old Caucasian female with a family history of neurofibromatosis, presented with a 3 month history of atraumatic painful right plantar lateral soft tissue mass. Initial MRI revealed 2.6 cm mass, presumed as neurofibromatosis. Change in shoewear and offloading strategies alleviated pain.

05/2018: She represented with a recurrent enlarging and painful mass. Repeat foot MRI demonstrated increase mass size with new bone involvement.

06/2018: She underwent ultrasound guided biopsy which demonstrated small round blue cell malignancy with positive FISH results. Musculoskeletal Surgical Oncology diagnosed Extraskeletal Ewing sarcoma. Regional Sarcoma Tumor Board recommended staging sarcoma with metastatic work up including PET/CT scans with initiation of neoadjuvant chemotherapy, subsequent surgical excision and adjuvant chemotherapy.

07/2018: Sarcoma staging: Bone, AJCC 7th Edition. Clinical stage T1, N0, M0. Neoadjuvant chemotherapy was initiated: vincristine, doxorubicin, cyclophosphamide, ifosfamide, etoposide

08/2018: After 4 weeks of neoadjuvant chemotherapy, the clinical exam revealed tumor size regression and new mass mobility, presumed improvement with chemotherapy. However repeat MRI demonstrated tumor size progression and worsening involvement with fifth metatarsal.

09/2018: Right complete fifth ray amputation with wide en masse resection with a peroneus brevis tendonized to cuboid with 4.75 mm biotendon screw was performed. A healthy appearing margin was visualized. The wound was primarily closed.

10/2018: Final pathology results showed narrow negative margins with a necrosis percentage of 30%, with ideal percentage of 95%. Tumor board recommended resuming and completion of initial chemotherapy with follow up PET scan.

01/2019: Follow up right foot MRI with no recurrence. Follow chest CT with no interval changes.

10/2019: One year post surgical follow up, she was ambulatory and foot MRI demonstrates no local recurrence.

Histopathology

A Rare Case of Extraskeletal Ewing Sarcoma of the Foot
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References