A Rare Case of Extraskeletal Ewing Sarcoma of the Foot

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Statement of Purpose

We present a rare case report of extraskeletal Ewing sarcoma in the foot which was treated with neoadjuvant chemotherapy and a wide resection including right fifth ray amputation with peroneus brevis tenodesed to cuboid. No local recurrence or metastases with surveillance imaging in a 14 month follow-up.

Literature Review

Ewing sarcoma (ES) is a rare, malignant primary bone tumor commonly affecting children and young adults.[1-4] The Ewing sarcoma family of tumors consists of skeletal/extraskeletal Ewing sarcoma, neuroepithelioma, primitive neuroectodermal tumor, and Askin tumors.[5,6] 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.[7] 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.

Histopathology

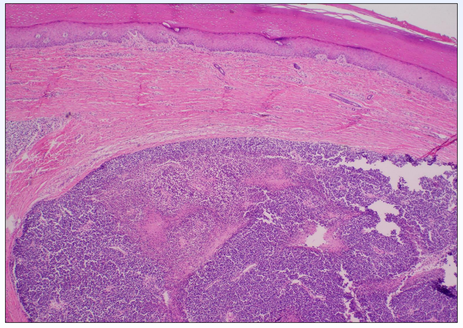

Figure 1. Biopsy of mass with small round blue cell tumor underneath the skin with necrosis. 40x magnification with Hematoxylin and Eosin stain

Clinical Photos and Imaging

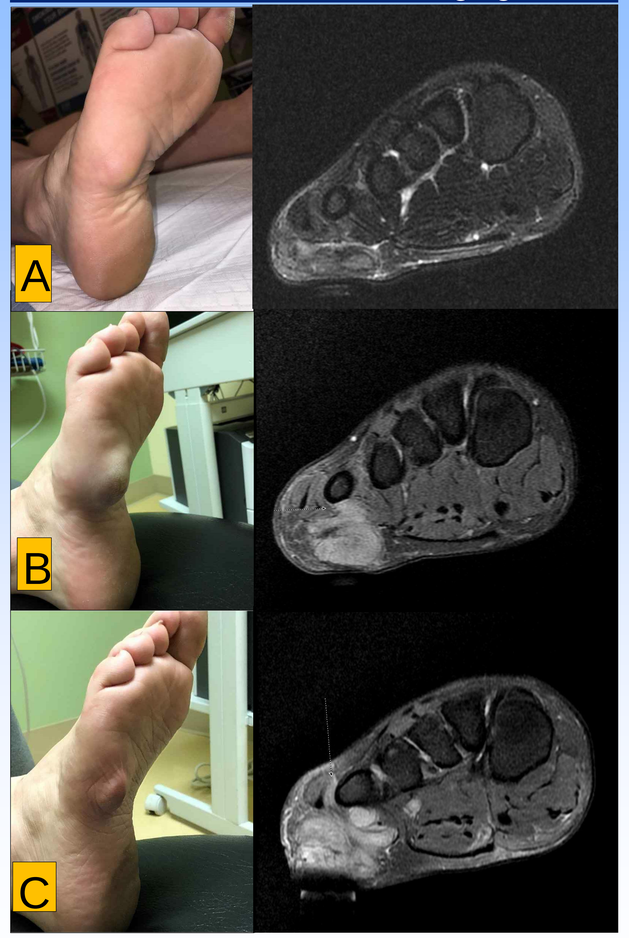

Figure 2. A) 10/2017 Initial presentation of soft tissue mass with MRI of 2.6cm mass within subcutaneous adipose layer
B) 05/2018 Tumor enlargement with MRI demonstrating new bone involvement.
C) 08/2018 After 1 month of neoadjuvant chemotherapy, the tumor decreased in clinical size but MRI demonstrated worsening tumor progression and invasion.

Case Report

10/2017: Twenty-two-year-old Caucasian female with a family history of neurofibromatosis, presented with a five month history of atraumatic painful right plantar lateral soft tissue mass. Initial MRI revealed 2.6 cm mass, presumed as neurofibromatosis. Change in shoegear 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 neovaduvant 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 tenodesed to cuboid with 4.75 mm biotenodesis 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.

Intra-operative Photos

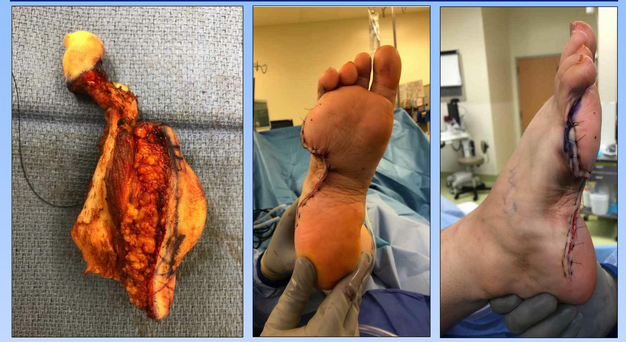

Figure 3. Functional foot sparing procedure of an en masse excision of 4.0 x 4.0 x 3.0 cm extraskeletal Ewing sarcoma of 5th ray with peroneal brevis tenodesed into cuboid with primary closure.

Discussion

Extraskeletal ES is a rare sarcoma in the foot, that is difficult to diagnosis due to early nonspecific features. Early diagnosis and treatment is necessary to prevent metastases and improve prognosis. Applebaum et al. found that regional lymph node involvement with extraskeletal ES was 12.4% compared to 3.2% for primary bone tumors.[8] Treatment of extraskeletal ES often includes chemotherapy followed by wide surgical excision with or without radiotherapy.[9,10]

This case report demonstrates the difficulty of extraskeletal ES diagnosis and tumor rapid progression despite neoadjuvant chemotherapy. The clinical fluctuant appearance was likely due to associated inflammation which highlights the importance of serial imaging to monitor tumor progression. Given her age and local disease, a functional foot salvage amputation procedure with negative margins was performed. At her one year post surgical follow up, she was ambulatory with no evidence of local recurrence. Her imaging surveillance plan is every 3 months for two years and then every 6 months for 3-5 years.

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