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Masquerading Malignancy: A Pedal Synovial Sarcoma

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Statement of Purpose

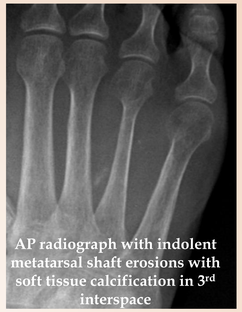
- Often presenting with benign characteristics, synovial sarcomas (SS) can cause significant morbidity and mortality if not identified early and treated aggressively. Primary synovial sarcomas of the lower extremities are rare, however the trained foot and ankle surgeon should practice a high degree of suspicion when presented with a constellation of several of the associated features.
- Hereby presented is a case of a rare primary malignant process arising within the forefoot. It particularly emphasizes the importance of an interdisciplinary approach to efficacious and proper management of an infrequently-encountered pathology.
- Multiple non-podiatric specialties within UF Health Jacksonville were closely involved in our patient's care, including General/Oncologic Surgery, Radiology, Pathology, and Radiation Oncology. Each service provided individualized care to our patient, resulting in a promising final prognosis.

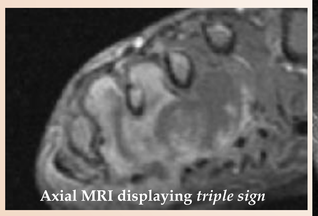
Literature Review

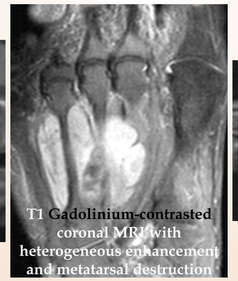
- Accounting for 8-10% of all primary malignant soft-tissue tumors, synovial sarcoma (SS) is the most common malignant soft tissue sarcoma of the foot. Over two-thirds of SS arise from the lower extremities. Often regarded as a misnomer, synovial sarcoma does not directly arise from synovial tissue; it is more commonly found adjacent to tendon sheathes and joints. Having a higher male preponderance, roughly 30% of SS emerge within the first two or three decades of life. Classic SS is described as "biphasic" – having two predominant cell types – fibrous and epithelial cells. Lung and lymph tissue are the most common sites for metastasis. The clinical examination is frequently positive for a slow-growing, occasionally painful mass of the lower extremity.[1-4]
- Radiographic features include calcification of tumor in 25% of cases and osseous involvement in 5-20%. Indolent, nonaggressive osseous erosions are appreciated in 15% of SS. CT scan can be useful in patients who cannot undergo MRI, as calcification and bone involvement can be detected.[5] However, MRI is considered standard imaging modality, due to paramount contrast resolution. MRI is utilized not only for preoperative planning, but for staging and diagnosis. T1/T2-weighted images, SS typically appears as heterogeneous multilobulated mass with high signal intensity. The *triple sign* is the combination of intermingling areas of low, intermediate, and high signal intensity on long repetition images; representing calcified/fibrotic collagenized regions, solid cellular elements, and hemorrhage/necrosis, respectively.[6]
- SS is recognized genetically by its distinguishing balanced translocation t (X; 18) (p11.2;q11.2). One of three SSX genes from chromosome X are fused with the SYT gene on chromosome 18, creating an SYT-SSX fusion gene. Variants of this particular translocation (X;18) are commonly encountered in the majority of SS and are unique to this tumor.[7]
- Wide surgical resection is the current mainstay of treatment of SS; studies have shown worse survival rates for patients with margins of less than 2 mm.[8] Limb amputations are generally reserved for patients with large lesions or deficient soft tissue envelope; limb salvage surgery is attempted in most patients.[4,9] Adjuvant radiotherapy is frequently utilized in treatment of other soft tissue sarcomas, and is indicated for patients with tumors greater than 5cm in size.[2] A recent study showed adjuvant radiation therapy had no statistically significant promotion of overall survival; however it did cause clinically significant edema, functional limb deficits, and more wound complications.[10]
- Progress with limb salvage surgery and concomitant radiotherapy has led to increases in local control and long-term survival. For nonmetastatic surgically-resected SS, the current 5-year overall and metastatic-free survival rate is 71% and 51% respectively. The projected 10-year survival rate is 20-65%.[11] Another recent study displayed that tumors more than 3 cm in total dimension were associated with a poorer overall survival. Patients treated with adjuvant radiation were at heightened chance for postoperative complications such as surgical site infections.[12]

Case Study

- ❖ A case study is presented of a 47-year-old female who presented to UF Health in Jacksonville with a chief complaint of a painful mass to the bottom of her foot. It had been slowly growing in size over the past two years and has become noticeably more painful, affecting her ambulation.
- ❖ On physical examination, a roughly 4cm x 4cm nonmobile soft tissue mass was appreciated on the plantar aspect of the patient's right foot; it was very tender to palpation, sans erythema or drainage.
- ❖ Radiographs of her foot were obtained, showing mild over-tubulation of 4th metatarsal shaft. Follow-up contrasted MRI was performed which revealed a 3x4x5cm multi-lobulated enhancing mass, interdigitating between the third and fourth metatarsal spaces; exhibiting cystic components. There was indolent erosion of the fourth metatarsal and a small, dystrophic calcification in the intermetatarsal space. The lesion insinuated within several tendons, suggesting an extra-abdominal desmoid tumor, or more concerning - synovial sarcoma.

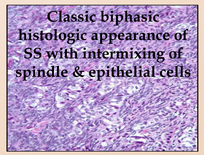

AP radiograph with indolent metatarsal shaft erosions with soft tissue calcification in 3rd interspace


Axial MRI displaying *triple sign*


T1 Gadolinium-contrasted coronal MRI with heterogeneous enhancement and metatarsal destruction


Gd-contrasted sagittal T1 MRI with tumor insinuating between flexor tendons

- ❖ Our patient was referred to the head of the Oncologic Surgery department, who scheduled our patient for immediate biopsy. Frozen sectioning showed biphasic sarcoma with spindle and epithelial cell components most consistent with synovial sarcoma. TLE-1 testing at an outside lab confirmed the diagnosis of SS, as the nuclear staining was positive in the epithelial tumor cells and less frequently in the spindle tumor cells.
- ❖ Following definitive diagnosis, a pretreatment PET scan was obtained, showing local SS only in the foot – no distant metastases were detected.
- ❖ Six weeks after initial diagnosis, local surgical resection of the tumor was performed in a joint case with oncologic surgery and podiatry. Fortunately for our patient, the location of the tumor was confined solely to her distal plantar foot. Intraoperatively, there was confirmed to be extensive metatarsal insinuation; to provide a functional amputation level and leave our patient with an ambulatory foot, it was decided to perform an amputation at the level of the midtarsal joint. Frozen sectioning revealed successful excision of entire tumor with negative margins; the closest margin was 0.3cm from the proximal aspect.


Classic biphasic histologic appearance of SS with intermixing of spindle & epithelial cells

Case Study cont.

- ❖ The patient was monitored closely in the postoperative period and suffered wound healing complications. Once tobacco cessation was victoriously achieved, healing progressed quickly in the weeks following.
- ❖ Radiation Oncology recommended against postoperative radiotherapy due to the lesion's local site and negative surgical margins; advising close wound observation and ongoing metastatic imaging surveillance.
- ❖ An ensuing PET scan was performed three months postoperatively, resulting with negative residual tumor without suggestions of metastases.
- ❖ Our patient is now a cancer survivor of over one year. She is fully-healed and ambulating without issue. She will undergo regular imaging surveys in her many promising years to come.


Initial surgical approach from medial site of foot


Interim Lisfranc amputation displaying proximal aspect of lobulated plantar tumor


Postoperative Day 21

Discussion

This case features a malignant pathology uncommonly encountered in routine podiatric practice. We attempted to emphasize the importance of recognizing a seemingly benign presentation of synovial sarcoma in the foot. Diagnostic vigilance and comprehensive workup is warranted for any long-standing or painless mass of the lower extremity. Our case illustrated that with timely diagnosis, treatment via multiple specialties, and close follow-up care, patients with this rare tumor can literally walk away with an encouraging prognosis.

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