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Benefits Associated With Early Diagnosis and Treatment of Soft Tissue Sarcomas of the Foot and Ankle

Drew Murray, BS [1], Jacob Topfer, BS [2], Alexander P. Decilveo, BA [1], James C. Wittig, MD [3]

[1] Medical Student, Robert Wood Johnson Medical School, New Brunswick, NJ
[2] Research Assistant, Department of Orthopedic Oncology, Hackensack University Medical Center, Hackensack, NJ
[3] Surgeon, Department of Orthopedic Oncology, Hackensack University Medical Center, Hackensack, NJ

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ABSTRACT

Soft tissue sarcomas arising in the foot and ankle are often misdiagnosed, resulting in excision without adequate preoperative staging or surgical margins. The goal of the present study was to review a case series of soft tissue sarcomas of the foot and ankle with attention directed at unplanned excisions of sarcomas. An unplanned excision means that a patient either underwent an inadequate preoperative workup or the preoperative workup indicated a benign entity, resulting in surgical resection. We retrospectively analyzed the medical records of 10 patients with sarcomas of the foot and ankle treated at our institution. All soft tissue sarcomas were excised with the widest margin possible without any major bone or neurovascular resection to allow for preservation of the foot. All patients were followed up for a minimum of 8 months to assess pain, function, and complications. The patients were followed up for an average of 22.4 (range 8 to 44) months. Of the 10 patients, 6 had undergone preoperative magnetic resonance imaging without contrast and the sarcomas were read as benign or cystic masses. Two patients had undergone preoperative magnetic resonance imaging with contrast, and these scans were also misread. Despite having undergone previous unplanned surgery, none of these sarcomas had recurred after repeat resection with a wider margin at a mean follow-up of 22.4 (range 8 to 44) months. At the last follow-up point, 8 patients were alive without evidence of disease. Minor complications included lymph edema in 1, stress fracture in 1, and wound infection in 1 that resolved. Preoperative MRI with contrast is recommended before resecting any soft tissue masses of the foot and ankle. In the event of an unplanned excision of a soft tissue sarcoma, the patient should be referred to an orthopedic oncologist for definitive surgery to optimize the oncologic and functional results. In the present retrospective analysis, previous intervention did not seem to affect the prognosis, including local recurrence, distant metastasis, disease-free interval, and functional outcomes.

Soft tissue sarcomas represent some of the most uncommon malignancies, constituting only 1% of cancer diagnoses (1). Fewer than 5% of soft tissue sarcomas occur in the foot and ankle (2). The 5- and 10-year survival rates range from 67% to 80% and 62.9% to 69%, respectively (2–5) and vary according to the histologic classification of the sarcoma. Overall, these sarcomas have a recurrence rate as great as 60% when treated only by local excision (6), with nearly 40% metastasizing to the lung (7). These numbers can be potentially explained by the unusually high occurrence of otherwise rare forms of soft tissue sarcomas with aggressive behavior (8) and the difficulty in achieving adequate surgical margins in the foot and ankle owing to their close proximity to bones, tendons, and neurovascular structures (3,9). Several studies (1,2,8–11) have found that the most common type of soft tissue sarcoma in the foot and ankle is synovial sarcoma (rate of metastases of 50% to 70%) (4), followed by clear cell sarcoma (10% to 14% rate of metastases) (5). Although less common, rhabdomyosarcoma, leiomyosarcoma, liposarcoma, fibrosarcoma, and malignant fibrous histiocytoma also occur in the foot and ankle (9–12).

In the setting of limb-sparing surgery, many studies have recommended total excision according to the preoperative radiologic imaging findings (2,8,9,13–15). Adjuvant radiotherapy is often indicated to eradicate potential residual microscopic disease (16,17). However, the use of adjuvant chemotherapy for soft tissue sarcomas of the foot and ankle remains controversial (5,7,18).

Despite the increasing body of data on soft tissue sarcomas, as many as 40% of cases are still misdiagnosed, and this percentage could be even greater in the foot and ankle (2,9,13). The confusion surrounding the diagnosis might result from the painless presentation of the tumor

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Address correspondence to: James C. Wittig, MD, Hackensack University Medical Center, Hackensack, NJ 07601.
E-mail address: drjameswittig@gmail.com (J. C. Wittig).

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(11,16), the relative infrequency of soft tissue sarcomas compared with benign lesions (9,12,19), and their nonspecific radiologic findings (16). These characteristics have resulted in as many as 34% to 67% of patients undergoing an unplanned marginal or piecemeal excision (1,9,10,14). An unplanned excision can result from a patient undergoing an inadequate preoperative workup, thus resulting in the "unplanned" surgical resection of, perhaps, a benign entity.

The published data are controversial regarding whether unplanned excisions adversely affect the prognosis and outcome. Several studies have shown that unplanned excisions have a negative effect on the local recurrence rates (2,6,15,16), oncologic outcomes, necessity for amputation (9), and contamination of other tissues (8,14). Thirty-five to fifty percent of patients who undergo unplanned excisions will have residual sarcoma in the tumor bed (16,20). Despite this, Helsin et al (6) and Gustafson et al (21) found that local recurrence did not affect the time to the development of distant metastases, suggesting that these patients already had microscopic disease at presentation. Nishimura et al (2) and Temple et al (22) found no difference in tumor-related deaths for patients who had undergone an unplanned excision. In a study of 1092 patients diagnosed with primary extremity soft tissue sarcoma, Lewis et al (19) found that repeat resection after unplanned excision improved survival across all grades compared with the statistics for patients who had undergone definitive surgery initially for their neoplasm. Some studies have suggested that the discrepancy might be attributable to the extent of the initial excision (20) or inherent bias in the planned excision group, which might have had larger tumors, a significant prognostic factor (2).

The goal of the present study was to review our case series of soft tissue sarcomas arising in the foot and ankle with regard to the radiologic and clinical presentation, pathologic analysis, previous preoperative imaging findings, and inaccurate diagnoses. Special attention was given to interventions performed before consultation at our facility, including aspirations and unplanned excision.

Patients and Methods

The goal of the present study was to retrospectively review the data from patients presenting to our office from 2008 to 2015 for foot and ankle sarcomas. A total of 91 patients had presented with a soft tissue tumor, 19 of whom were diagnosed with a soft tissue sarcoma. Of these 19 patients, 10 were treated at our institution. These 10 patients were the focus of the present study.

The patient data were analyzed for the following variables: tumor size, tumor grade, tumor location, pathologic analysis, previous unplanned excision or intervention, surgical margin status or residual sarcoma, initial diagnosis, preoperative symptoms, occurrence of distant metastases, local recurrence, and the use of adjuvant radiotherapy or chemotherapy. For the purposes of the present study, the patients were grouped into those who had undergone previous interventions outside of our institution and those who had not. We assessed function, pain, ambulatory capacity, postoperative complications, disease-free interval, local recurrence, and systemic recurrence. The institutional review board of our institution approved the present study.

Before definitive surgery, all the patients underwent magnetic resonance imaging (MRI) with gadolinium and staging studies. Limb-sparing excision was performed on all patients, using as wide a margin as possible. No patient required adjacent bony resection. Radiotherapy was administered on a case-by-case basis according to the pathologic analysis, including the size and grade of the mass, and the presence of tissues potentially contaminated by previous interventions. The patients were followed up with computed tomography and MRI to assess for local recurrence and distant metastases. Chemotherapy was considered on a case-by-case basis with consultations with the appropriate specialists.

Results

The details of the 10 patients treated at our institution for soft tissue sarcoma of the foot and ankle are listed in Table 1. The symptoms at presentation were nonspecific. Of the 10 patients, 6 (60%) presented with a painless mass noticed on inspection or palpation, and 4 (40%) complained of swelling, stiffness, or limited range of motion. The most common preliminary diagnosis was a ganglion cyst (Fig. 1; n = 4 [40%]) followed by schwannoma (n = 2 [20%]). Two patients were given a differential diagnosis that included a potential malignancy. Only 1 patient was diagnosed accurately from the preoperative imaging findings (Table 2). Of the 10 patients, 4 (40%) had undergone MRI with contrast to aid in the diagnostic process, one half of which were misread as showing schwannoma. Eight (80%) patients were thought to have benign masses and had undergone previous intervention before their presentation at our institution. Of these 8 patients, 2 had undergone preoperative MRI with contrast.

Data related to the treatment and tumor characteristics are listed in Table 2. The most common histologic diagnosis was synovial sarcoma (Fig. 2; n = 5 [50%]), followed by clear cell sarcoma (n = 2 [20%]). Dermatofibrosarcoma protuberans, low-grade myxoid sarcoma, and alveolar rhabdomyosarcoma occurred in 1 (10%) patient each. Chemotherapy was administered postoperatively to 3 (30%) patients, and 1 (10%) had undergone chemotherapy before surgery. Six (60%) patients received radiotherapy postoperatively. Three (30%) of these patients received both chemotherapy and radiotherapy postoperatively. These 3 patients had presented with high-grade synovial sarcoma (n = 2) or alveolar rhabdomyosarcoma (n = 1). The tumor sizes were 3.7 cm, 2.5 cm, and 1.3 cm.

The mean time from the initial presentation to definitive surgery for all patients was 23.25 (range 0.5 to 96) months. The mean time from presentation to definitive surgery was 4.28 (range 0.5 to 8) months in the group receiving the initial intervention at our institution and 28 (range 3 to 96) months in the group that had undergone previous interventions. If the 1 (10%) patient with an unusually long delay in treatment were treated as an outlier and removed from the calculation, the average time from presentation to definitive surgery in the previous intervention group was 18.28 (range 3 to 36) months.

Data pertaining to the measured outcomes of our patients are presented in Table 3. The patients were followed up for a mean of 22.4 (range 8 to 44) months. Four (40%) of the patients presented with

Table 1
Patient demographics, previous treatment, and grouping

Patient No.	Age (y)	Sex	Chief Complaint	Initial Imaging Study	Initial Diagnosis	Grouping
1	23	M	Injury, pain, swelling	MRI without contrast	Ganglion cyst	Previous treatment
2	29	M	Painless mass	MRI with contrast	Mesenchymal tumor	Previous treatment
3	28	F	Difficulty walking	MRI with contrast	Cyst/schwannoma	Previous treatment
4	46	F	Painless mass	MRI without contrast	Neuroma/schwannoma	Previous treatment
5	19	F	Painless mass	MRI without contrast	Ganglion cyst	Previous treatment
6	35	F	Painless mass	MRI without contrast	Ganglion cyst	Previous treatment
7	49	M	Painless mass	No MRI	None given	Previous treatment
8	66	F	Painless mass	No MRI	Soft tissue mass	Previous treatment
9	2	M	Swelling	MRI with contrast	Rhabdomyosarcoma	No previous treatment
10	35	F	Swelling, pain, limited ROM	MRI with contrast	Inconclusive, potential malignancy	No previous treatment

Abbreviations: F, female; M, male; MRI, magnetic resonance imaging; ROM, range of motion.