SOFT TISSUE SARCOMAS

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Introduction

Soft tissue sarcomas occurring in the foot present a unique diagnostic and therapeutic challenge. Frequently, the patient presenting with such a mass will be concerned as to its potential malignancy. Due to the numerous variations in clinical appearance and behavior, the podiatrist must possess an appreciation of basic principles involved in their management. With this knowledge, one can confidently approach the tumor with a proper plan of treatment. This paper describes a stepwise approach to the assessment of the soft tissue sarcoma. This approach can be similarly used for virtually all soft tissue tumors. The approach includes:

1. patient history,
2. physical examination,
3. accessory studies,
4. biopsy, and
5. definitive treatment plan.

The understanding of this approach will enable the physician to play a vital part in the management of patients with soft tissue sarcomas.

Distribution/Etiology

Soft tissue sarcomas are relatively rare neoplasms. However, they are most frequently seen in the extremities and will be encountered by the podiatric physician. Etiologic factors are not well understood, but are believed to be related to the activation of viral oncogenes. Chemical exposure, radiation exposure, and trauma have all been implicated as possible factors initiating the viral induction of tumor (1).

Classification

The majority of soft tissue sarcomas are thought to arise from the primitive mesoderm. However, the ectoderm does give rise to neurofibrosarcomas. Thus, sarcomas may be classified based upon their cell of origin (2) (Figure 1).

Clinical Presentation

The clinical presentation of soft tissue sarcomas can vary tremendously. The usual presentation is that of a nonspecific solid mass in the subcutaneous or deeper tissues. It is generally brought to the patient's attention by its physical size, discomfort, or paresthesias.

The patient is usually asymptomatic and first seeks medical attention when a recent change in size of a mass is noted or when the size of the mass limits shoegear. The patient will frequently present with only the innocent complaint of "swelling" in the area of the foot and/or ankle. This can be very misleading and the skilled examiner must be alert to the possibility of an underlying neoplasm.

Germ cell layer	Normal cell	Sarcoma
Ectoderm	Nerve cell sheath	Neurofibrosarcoma
		Alveolar soft-part sarcoma(?)
Mesoderm	Adipose cell	Liposarcoma
	Fibroblast	Fibrosarcoma
		Dermatofibrosarcoma problems
		Malignant fibrous histiocytoma(?)
	Smooth muscle	Leiomyosarcoma
	Skeletal muscle	Rhabdomyosarcoma
	Tendosynovial cell	Synovial sarcoma
		Epitheloid sarcoma(?)
		Malignant giant cell tumor
	Blood vessel	Angiosarcoma
		Hemangiopericytoma
		Lymphangiosarcoma
		Kaposi's sarcoma
	Miscellaneous	Malignant mesenchymoma
		Malignant granular cell tumor

Fig. 1. Cell of origin of soft tissue sarcomas.

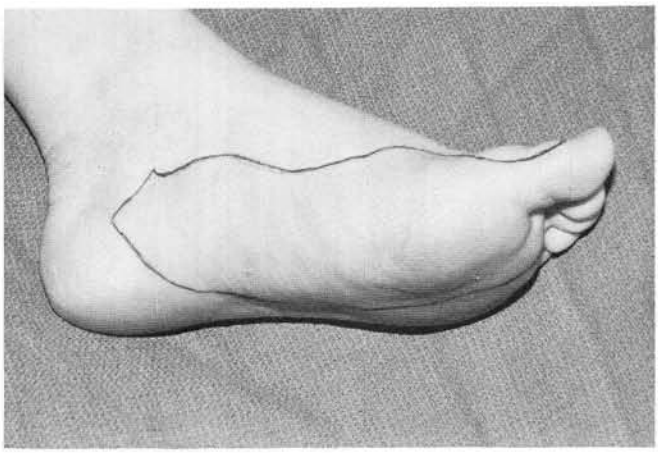

Fig. 2. Patient with plantar soft tissue mass creating exogeneous compression on the medial plantar nerve. The patient was given a skin scribe and "mapped out" the area of paresthesia.

Discomfort or paresthesias generally result from exogenous pressure of the mass upon peripheral nerve(s). Sensory abnormalities predominate over motor abnormalities and are typically well localized over the distribution of the compressed nerve(s). Pain is usually sharp or burning in nature and frequently increases with activity (3). A thorough understanding of the peripheral nerve anatomy and nerve compression are paramount to proper clinical examination (4).

Clinical Evaluation

The initial step in the evaluation of a soft tissue sarcoma is to obtain a thorough history of the lesion. The standard NLDOCAT—nature, location, duration, onset, course, aggravating factors, and treatment should always be determined. Soft tissue sarcomas are generally slow growing tumors and rate of growth will only occasionally be helpful in assessing the aggressiveness of the lesion and its potential for malignancy.

On physical examination, the mass should be accurately palpated and measured. The depth of the mass and its fixation to bone, tendon or other surrounding structures should be determined. This can generally be done with active and passive movement by the patient and by direct palpation by the examiner. Tumors above the deep fascia will generally be mobile, while those below the deep fascia will be fixed. Palpation should also include evaluation of the texture and density of the soft tissue mass.

Careful examination should also be performed for any possible neurovascular involvement. Pulses should be compared to the contralateral limb and the proximity of the tumor to major vessels assessed. The limb should be evaluated for any lymphatic or venous obstruction as well. Neurologically, palpation and percussion of the mass and surrounding nerves should be performed to assess possible involvement. In some cases the lesion may actually be endogenous to the nerve (e.g., malignant Schwannoma) rather than exogenous. Occasionally, with sensory involvement, the patient can point to areas of proximal or distal paresthesia and a specific nerve can be "mapped out" in the process (Fig. 2). Active or passive manipulation of the involved areas may exacerbate symptomatology and make the neural pathology easier to demarcate.

Although lymph node involvement is unusual, a complete physical examination should include palpation of both popliteal and inguinal lymph nodes. The areas should be soft and nontender to palpation. If the lymph nodes are easily palpated yet soft and easily movable, possible reactive lymphadenitis may be present. Harder, fixed nodes suggest possible metastatic sarcoma (5).

Accessory Studies

Once a complete history and physical examination has been performed, additional tests can be undertaken to further assess the mass. Radiographs, laboratory studies, and advanced imaging techniques can be performed if necessary.

Standard pedal radiographs are of limited value in evaluating the soft tissue tumor. The radiographs in such instances should always be taken with soft tissue attenuation. Standard radiographs with soft tissue attenuation and xeroradiographs offer similar visualization of the soft tissue mass. The margins of the mass may be determined in some instances. Evaluation should also be undertaken for any osseous invasion which may be demonstrated by cortical erosions. Finally, the mass should be radiographically inspected for calcifications. Calcifications occur due to metastatic, metabolic, and dystrophic causes. Their identification in a soft tissue mass can be highly suggestive of malignancy (6,7).

Chest radiographs should be performed and examined for possible metastatic spread. The first site of sarcoma metastasis is usually pulmonary, and about 15% of patients presenting with a primary soft tissue sarcoma will have pulmonary metastases. About two-thirds of these metastases are detectable on conventional chest radiographs and the remainder are visualized on chest tomography (8).

Laboratory studies are usually of little value in evaluating the possible soft tissue sarcoma. Alterations of the complete blood count, and blood chemistry profile may be noted once there is significant systemic involvement and metastasis.

More advanced imaging techniques may also be performed. Computerized axial tomography (CT) and nuclear magnetic resonance (NMR) also known as magnetic resonance imaging (MRI) are most helpful. Both of these techniques allow cross-sectional visualization of the anatomical part (Figs. 3, 4). Further, they both allow some assessment of the boundaries and possible tissue composition of a soft tissue mass. This can be particularly helpful in planning biopsies or surgical excision. MRI measures the water content of various body tissues, and since tumors differ from surrounding tissues primarily in water content it is generally preferable for imaging the soft tissue mass. CT-scan and MRI may also be helpful in detecting recurrence after excision of the mass or in determining remission in nonoperative cases.

Nuclear medicine studies may also prove beneficial. Enneking and associates (9) reported 92% accuracy in detecting the presence or absence of contiguous bone involvement by soft tissue sarcomas utilizing technitium-99m bone scans. However, bone scans are of no use in the identification of distant metastases, and therefore, their value is limited to confirmation of any bone involvement, or lack thereof, seen with other imaging techniques. Similarly, gallium-67 scans are of little value in assessing the soft tissue sarcoma. Bitran and associates (10) found that distant

metastases were detectable in some histological types of sarcoma (malignant schwannoma and undifferentiated sarcoma) but were frequently associated with false-negatives in most types of sarcoma.

In some instances, other tests may be desirable. For example, if the mass is pulsatile or near a major vessel, an angiogram may be ordered prior to biopsy or surgical excision. This will enable the surgeon to properly plan treatment.

Biopsy Techniques

There are four types of biopsy procedures that may be employed to establish the diagnosis of soft tissue sarcomas: fine needle aspiration, needle biopsy, incisional biopsy, and excisional biopsy. These are essential to make the histological diagnosis and establish the appropriate treatment plan.

Fine needle aspirations have been shown to be useful in the diagnosis of many carcinomas, but generally should not be used for the definitive diagnosis of soft tissue tumors. Aspiration may demonstrate certain cytological characteristics, but exact histological classification and the degree of differentiation are virtually impossible to determine from fine needle aspiration samples (5).

Needle biopsies are cores of tissue which can be obtained with various types of needles. The technique is advantageous over fine needle aspiration as it provides a tissue diagnosis and not just a cytological diagnosis. By obtaining several tissue cores a histological assessment of the tumor can be made.

Excisional biopsy should generally not be performed when a soft tissue tumor is suspected of being malignant. Excisional biopsy will compromise the tissue planes and surrounding structures making subsequent definitive procedures more difficult. Since most soft tissue sarcomas present with a pseudocapsule, simple excision of the mass will certainly leave microscopic tumor behind. Smaller tumors and more superficial tumors may be amenable to excisional biopsy in certain instances. Incisions should be placed in line with underlying structures (muscle, tendon, bone, etc.) should these structures need to be excised.

Incisional biopsy is usually the procedure of choice for assessing soft tissue tumors. The biopsy can be performed by placing an incision over the mid-portion of the mass and carried deep to the tumor. In this way, contamination of adjacent tissue planes is minimized. Again, specimens are obtained for tissue examination and histological diagnosis. Frozen sections can be obtained, if available, but a treatment plan should NOT be made based solely upon their report. Final histological sections and diagnosis should be awaited before the treatment plan is finalized (5).

Treatment Plan

The definitive treatment plan will depend on certain prognostic criteria including the site of the tumor, tumor size, degree of differentiation, histological type, and age. Involvement of an oncologist will certainly become necessary at this point if one has not been consulted earlier.

The site of the tumor is generally considered to be an important factor affecting the clinical course and prognosis. The distal extremity can be treated much more easily than proximal areas and is associated with a better prognosis in most cases.

Tumor size is also important. Sarcomas smaller than 5 cm have a far superior prognosis compared with those

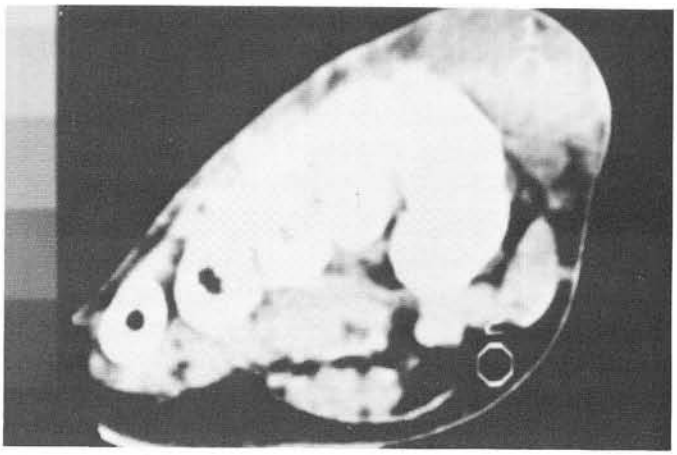

Fig. 3. Frontal plane CT-scan of forefoot demonstrating large soft tissue mass over dorsomedial aspect. Proximal portions of metatarsals are also noted.

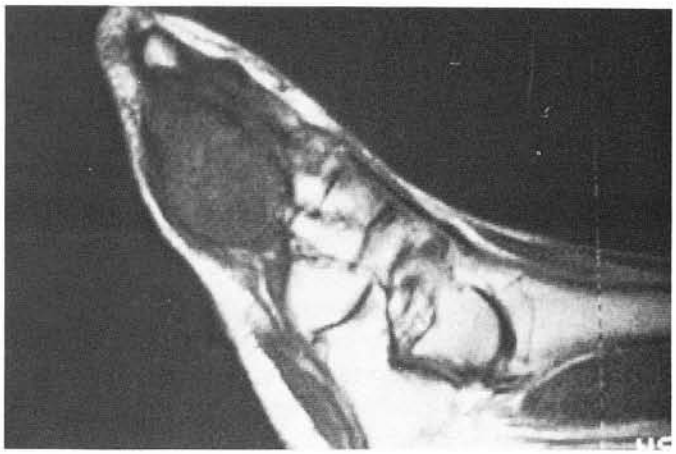

Fig. 4. Sagittal plane MRI scan demonstrating a soft tissue mass in plantar forefoot area.

Figure 5A: Schema for Staging Soft-Tissue Sarcomas by TNMG

T	Primary tumor T_1 Tumor less than 5 cm T_2 Tumor 5 cm or greater T_3 Tumor that grossly invades bone, major vessel, or major nerve
N	Regional lymph nodes N_0 No histologically verified metatastasis to regional lymph nodes N_1 Histologically verified regional lymph node metastasis
M	Distant metastasis M_0 No distant metastasis M_1 Distant metastasis
G	Histologic grade of malignancy G_1 Low G_2 Moderate G_3 High

Figure 5B: Stage Grouping for Soft-Tissue Sarcomas

Stage I	
Stage Ia $G_1T_1N_0M_0$	Grade 1 tumor less than 5 cm in diameter with no regional lymph node or distant metastases
Stage Ib $G_1T_2N_0M_0$	Grade 1 tumor 5 cm or greater in diameter with no regional lymph node or distant metastases
Stage II	
Stage IIa $G_2T_1N_0M_0$	Grade 2 tumor less than 5 cm in diameter with no regional lymph node or distant metastases
Stage IIb $G_2T_2N_0M_0$	Grade 2 tumor 5 cm or greater in diameter with no regional lymph node or distant metastases
Stage III	
Stage IIIa $G_3T_1N_0M_0$	Grade 3 tumor less than 5 cm in diameter with no regional lymph node or distant metastases
Stage IIIb $G_3T_2N_0M_0$	Grade 3 tumor 5 cm or greater in diameter with no regional lymph node or distant metastases
Stage IIIc Any $GT_{1-2}N_1M_0$	Tumor of any grade or size (no invasion) with regional lymph node but no distant metastases
Stage IV	
Stage IVa Any $GT_3N_{0-1}M_0$	Tumor of any grade that grossly invades bone, major vessel, or major nerve with or without regional lymph node metastases but without distant metastases
Stage IVb Any $GTNM_1$	Tumor with distant metastases

Fig. 5. Staging system for soft tissue sarcomas using TNMG system. (From Russell WO, Cohen J, Enzinger F, Hajdu SI, Heise H, Martin RG, Meissner W, Miller WT, Schmitz RL, Suit HD A clinical and pathological staging system for soft tissue sarcomas. *Cancer* 40:1562-1570, 1977.)

larger than 5 cm (11). Larger tumors are more difficult to adequately resect and usually have greater involvement of surrounding structures.

The most important prognostic variable is probably the degree of differentiation of the sarcoma. Patients with well differentiated tumors do relatively well, while those with poorly differentiated tumors do considerably worse (11,12).

Histological evaluation is also a prognostic factor. For years, clinicians and pathologists based their treatment on histological classification alone. However, differentiation is much more important. A well-differentiated fibrosarcoma will have virtually the same prognosis as a well-differentiated tumor of another histological type (5).

	No. of cases	Percent surviving* 1 year	2 years	5 years	10 years
All stages	1215	74%	58%	41%	30%
Complete staging information	702	73	55	40	30
Stage I	177	91	84	75	63
IA	55	98	93	(83)	(75)
IB	122	88	80	72	(58)
Stage II	86	87	74	(55)	(40)
IIA	29	(90)	(79)	(59)	(41)
IIB	57	86	(72)	(53)	(40)
Stage III	329	71	47	29	19
IIIA	69	84	(61)	42	27
IIIB	211	68	46	26	16
IIIC	49	(65)	(34)	(23)	(23)
Stage IV	110	37	19	7	3

*( ) = standard errors between 5% and 10%.

Fig. 6. Survival by stage of soft tissue sarcomas. (From Russell WO, Cohen J, Enzinger F, Hajdu SI, Heise H, Martin RG, Meissner W, Miller WT, Schmitz RL, Suit HD A clinical and pathological staging system for soft tissue sarcomas. *Cancer* 40:1562-1570, 1977.)

Likewise, age fails to correlate well with prognosis. Children with some sarcomas do better than adults, but most sarcomas do similarly in any age group.

These factors have led to the development of a tumor staging system for sarcomas by the American Joint Committee for Cancer Staging and End Results Reporting. The system known as the TNMG system can give survival rates based upon primary tumor size and tissue involvement (T), regional lymph node involvement (N), presence of distal metastases (M), and tumor histological grade (G) (13) (Figs. 5, 6).

Based upon the tumor's stage, appropriate definitive treatment can be planned. Surgical resection, amputation, radiation therapy, and chemotherapy all can be utilized alone or in combination.

The "key" principle in surgical treatment is that resection must achieve adequate margins to prevent local recurrence. If adequate local resection can be obtained, then amputation can be avoided (11). Local resection is generally combined with radiation therapy and/or chemotherapy. These combinations vary from tumor to tumor and are very controversial.

As stated earlier, regional lymph node involvement is infrequent. Therefore, lymph node dissection is generally not indicated. However, if the lymph nodes are hard and fixed or if there is evidence of distal clinical lymphadenopathy then the lymph nodes may be resected.

The most common failures following sarcoma treatment are local recurrences and distant metastasis. The clinical presentation of local recurrence is almost always the appearance of one or more nodules at the previous site of operative resection.

Summary

Although relatively rare, soft tissue sarcomas are more prevalent in the extremities. The podiatrist should be alert to the possibility that a soft tissue mass may indeed be a soft tissue sarcoma. Proper evaluation including a stepwise assessment and evaluation will ensure proper diagnosis and treatment avoiding unnecessary surgery, amputation, and mortality.

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