Soft-Tissue Tumors: Diagnosis, Evaluation, and Management

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Abstract

Benign soft-tissue neoplasms and tumorlike conditions of the musculoskeletal system are common. Sarcomas are less frequent, with only 5,000 new cases diagnosed each year in the United States. After plain radiographs of the affected area have been obtained, magnetic resonance (MR) imaging (both T1- and T2-weighted sequences) is the best imaging modality for detecting and characterizing the lesion. Although MR imaging is not specific in determining whether lesions are benign or malignant, it can be useful in evaluating other characteristics, such as size, pattern of growth, integrity of natural boundaries, and homogeneity. Biopsy must be done carefully, so as not to adversely affect the outcome. Technical considerations include proper location and orientation of the biopsy incision, meticulous hemostasis, and frozen-section analysis to ensure that diagnostic material has been obtained. Effective treatment requires close coordination between the surgeon, the radiation oncologist, the pathologist, the plastic surgeon, and the diagnostic radiologist. Limb-salvage surgery has resulted in a local control rate greater than 90%. High-grade tumors that are larger than 5 cm in diameter have the worst prognosis. The role of chemotherapy remains controversial and unresolved.

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Benign soft-tissue neoplasms and tumorlike conditions of the musculoskeletal system are common and include entities such as lipomas, hemangiomas, and giant cell tumors of the tendon sheath. Malignant lesions, such as soft-tissue sarcomas, are less frequent, with only 5,000 new cases each year in the United States.

There are many different causes of soft-tissue masses (Table 1). The principal types are (1) soft-tissue tumors and tumorlike conditions, (2) bone tumors that have penetrated the bone compartment and formed a soft-tissue mass, and (3) surface tumors of bone that have arisen from the cortex and periosteal tissues and grown into the soft-tissue compartment.

The purpose of this review is to discuss the diagnosis, evaluation, and management of masses arising in the soft tissues. The clinician must maintain an appropriate index of suspicion to make an early diagnosis of malignant neoplasm while being careful not to expend valuable resources on lesions that are neither aggressive nor malignant. Effective management depends on a knowledge of the classification and staging of soft-tissue tumors and consistent use of strategies for evaluation, biopsy, and treatment of both benign and malignant neoplasms.

Diagnosis and Evaluation

Clinical Presentation

Patients with a soft-tissue tumor generally present to their physician complaining of a lump, bump, or growth. Pain may be an accompanying symptom.

Obtaining a thorough history is an important first step in management. The following questions are important guides to establishing a differential diagnosis:

How long has the mass been present?

Masses that have been present for long periods of time are most likely benign. Examples include lipomas and hemangiomas. A new mass that has arisen over a short period must raise the index of suspicion of malignancy. However, some malignant neoplasms (e.g., synovial sarcomas) may be present for a number of years, and their chronic nature may be misleading to the clinician.

Is the mass enlarging in size?

An increase in the size of a mass indicates an active process. Malignant neoplasms tend to grow progressively. However, lesions that are not enlarging may still be malignant. Patients often have difficulty assessing the true growth pattern, as masses

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Table 1
Functional Classification of Soft-Tissue Masses

Tumors and tumorlike conditions arising in the soft tissues
Benign neoplasms
Lipomas
Hemangiomas
Fibromatosis
Malignant neoplasms
Sarcomas
Metastatic carcinomas
Tumorlike conditions
Heterotopic ossification
Tumoral calcinosis
Intramedullary bone tumors
Benign neoplasms (giant cell tumor)
Malignant neoplasms
Osteosarcoma
Ewing's sarcoma
Lymphoma
Myeloma
Tumorlike conditions (aneurysmal bone cyst)
Surface bone tumors
Benign neoplasms
Osteochondroma
Periosteal chondroma
Malignant neoplasms
Parosteal osteosarcoma
Periosteal osteosarcoma

in certain locations may not be noticed until they are of substantial size.

Is the mass causing pain?

Sarcomas often cause pain secondary to inflammation in the reactive zone of the tumor. Lesions that invade the periosteum may also cause pain. Abscesses are often painful. Sarcomas may undergo necrosis and hemorrhage within their substance, causing severe acute pain accompanied by a marked increase in size; thus, they may simulate an abscess or muscle trauma.

Is there any history of penetrating or nonpenetrating trauma?

A history of penetrating trauma suggests the presence of a foreign body, an infection, or a pseudoaneurysm. Nonpenetrating trauma can result in heterotopic bone formation. Antecedent trauma has been associated with the development of desmoid tumors (extra-abdominal fibromatosis).[1]

Is there a history of cancer?

Malignant neoplasms, such as breast and lung carcinomas, melanomas, and lymphomas, may metastasize to the soft tissues.

Is there a history of systemic signs and symptoms?

Systemic symptoms such as fever, chills, and malaise may be secondary to an abscess. Malignant neoplasms, such as lymphomas, Ewing's sarcoma, and extramedullary plasmacytoma, may also result in systemic symptoms. Angiosarcomas may cause microangiopathic hemolytic anemia (Kasabach-Merritt syndrome).

Is there a family history of soft-tissue masses?

Several conditions (e.g., neurofibromatosis, lipomas, and hemangiomas) have a pattern of familial inheritance (Table 2).

Physical Examination

Careful physical examination is important, as there may be several findings that suggest the possibility of a malignant neoplasm. Lesions that are large (greater than 5 cm), firm, deep-seated, and fixed to underlying tissues suggest a malignant process. Moderate tenderness also is compatible with a malignant process, as there is often an active inflammatory process within the reactive zone of the tumor. Small superficial and mobile lesions are more likely to be benign.

Several tumors have distinct features on physical examination. Extra-abdominal fibromatosis (desmoid) tumors frequently have a rocklike consistency. Epithelioid sarcoma often presents as a small, superficial nodule, which may ulcerate. Clear cell sarcoma also presents as a small nodule along a tendon sheath. When a mass is located in the region of a major blood vessel, the clinician should palpate the mass to detect pulsations and should listen for a bruit to exclude a pseudoaneurysm or an arteriovenous malformation.

One must carefully examine the entire extremity in which there is a soft-tissue mass. Malignant neoplasms may have satellite lesions in the vicinity of the predominant lesion. Regional and other lymph-node sites (cervical, supraclavicular, axillary, and inguinal) must also be examined. Malignant neoplasms that are more likely to metastasize to lymph nodes include synovial sarcomas, rhabdomyosarcomas, epithelioid sarcomas, and clear cell sarcomas. The clinician should examine the abdomen to detect hepatomegaly or splenomegaly.

Classification and Staging Systems

Soft-tissue tumors are most commonly classified according to the direction of cellular differentiation. There are over 200 types of benign lesions and 70 types of malignant lesions. The more common lesions that orthopaedic surgeons encounter[1] are outlined in Table 3.

Benign lesions can be classified into three categories.[2] Stage 1 lesions are latent or inactive. Stage 2 lesions are active and growing or causing symptoms. Stage 3 lesions are aggressive and are characterized by their large size and penetration of anatomic boundaries.

Malignant soft-tissue tumors have a centripetal pattern of growth (Fig. 1), expanding and penetrating natural barriers such as muscle, fascia, and periosteum. Surrounding the tumor is an interface between the tumor and

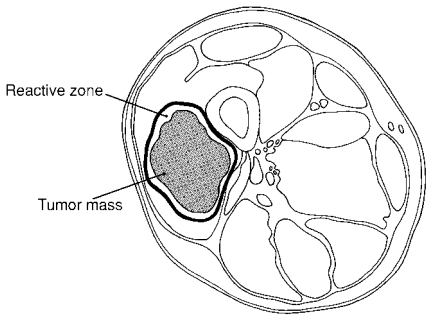


Fig. 1 Diagram of a malignant soft-tissue mass in the vastus lateralis depicts reactive zone surrounding the periphery of the lesion. The reactive zone contains edema fluid, inflammatory cells, fibrous tissue, and satellites of tumor cells.

normal tissues termed the "reactive zone," which contains edema fluid, inflammatory cells, fibrous tissue, and tumor-cell satellites.

Malignant lesions are often graded on the basis of morphologic characteristics within a given histologic entity. The surgical staging system developed by the Musculoskeletal Tumor Society is based on the grade of the lesion, local extension (intracompartmental or extracompartmental), and the presence or absence of metastases (Table 4).[3] An alternative staging system proposed by the American Joint Committee is also based on the grade, local extension, size, and presence or absence of regional or distant metastases (TNM system).

The most common malignant lesions can be categorized in a functional classification system (Table 5) as graded sarcomas, nongraded sarcomas, and small cell neoplasms (H. M. Reiman, MD, personal communication, June 1994). Graded sarcomas range from well-differentiated tumors to high-grade anaplastic tumors. Nongraded tumors tend to behave aggressively. Small cell neoplasms are responsive to both external-beam irradiation and chemotherapy.

Surgical procedures can also be classified according to the system of the Musculoskeletal Tumor Society (Fig. 2).[3] When the tumor has been entered but not entirely removed, its margin is termed "intralesional." If the reactive zone has been entered, the procedure is called a "marginal" resection. A "wide" margin is achieved when the entire lesion has been removed with a cuff of normal tissue around it. When the entire compartment containing the tumor has been removed, the resection is classified as radical.

Radiologic and Laboratory Studies

Once a thorough history has been obtained and a careful physical examination has been per-

Table 2
Soft-Tissue Tumors and Tumorlike Conditions With a Pattern of Familial Inheritance*

Type of Neoplasm	Pattern
Fibrous	
Palmar, plantar, and penile fibromatosis	Occasionally in several generations of one family and in twins
Fatty	
Lipoma	About 5% familial
Angiolipoma	About 5% familial
Fibrohistiocytic	
Xanthoma tuberosum	Occurs in familial hyperlipidemia
Tendinous xanthoma	Occurs in familial hyperlipidemia and in cerebrotendinous xanthomatosis inherited as an autosomal-recessive trait
Muscular	
Cutaneous leiomyoma	Occasional familial cases with a pattern suggesting autosomal-dominant mode of inheritance
Vascular	
Glomus	Occasional familial cases following an autosomal-dominant mode of inheritance
Osler-Weber-Rendu syndrome (hereditary hemorrhagic telangiectasia)	Autosomal-dominant inheritance
Blue rubber-bleb nevi (cavernous hemangiomas of the skin and gastrointestinal tract)	Some cases follow autosomal-dominant mode of inheritance
Neural or neuroectodermal	
Neurofibromatosis (von Recklinghausen's disease)	Autosomal-dominant inheritance with a high rate of spontaneous mutation
Neuroblastoma	Rare familial cases
Miscellaneous	
Fibrodysplasia (myositis) ossificans progressiva	Occasional familial cases
Tumoral calcinosis	Occasional familial cases

* Adapted with permission from Enzinger FM, Weiss SW: *Soft Tissue Tumors.* Philadelphia: CV Mosby, 1983, p 2.

Table 3
Histologic Classification of Common Soft-Tissue Tumors*

- Tumors and tumorlike lesions of fibrous tissue
  - Benign
    - Fibroma
    - Nodular fasciitis
    - Proliferative fasciitis
  - Fibromatoses
    - Superficial fibromatoses
      - Palmar and plantar fibromatosis
      - Knuckle pads
    - Deep fibromatoses (extra-abdominal fibromatoses)
  - Malignant
    - Adult fibrosarcoma
    - Postradiation fibrosarcoma
- Fibrohistiocytic tumors
  - Benign
    - Fibrous histiocytoma
    - Atypical fibroxanthoma
  - Intermediate (dermatofibrosarcoma protuberans)
  - Malignant (malignant fibrous histiocytoma)
    - Storiform-pleomorphic
    - Myxoid (myxofibrosarcoma)
    - Giant cell (malignant giant cell tumor of soft parts)
    - Inflammatory (malignant xanthogranuloma, xanthosarcoma)
    - Angiomatoid
- Tumors and tumorlike conditions of adipose tissue
  - Benign
    - Lipoma (cutaneous, deep, and multiple)
    - Angiolipoma
    - Spindle cell and pleomorphic lipoma
    - Lipoblastoma and lipoblastomatosis
    - Intramuscular and intermuscular lipoma
    - Hibernoma
  - Malignant
    - Liposarcoma
      - Well-differentiated (lipomalike, sclerosing, inflammatory)
      - Myxoid
      - Round cell (poorly differentiated myxoid)
      - Pleomorphic
      - Dedifferentiated
- Tumors of muscle tissue
  - Smooth muscle
    - Benign
      - Leiomyoma (cutaneous and deep)
      - Angiomyoma (vascular leiomyoma)
    - Malignant (leiomyosarcoma)
  - Striated muscle
    - Benign (adult rhabdomyoma)
    - Malignant (rhabdomyosarcoma [predominantly embryonal (including botryoid), alveolar, pleomorphic, and mixed])
- Tumors and tumorlike conditions of blood vessels
  - Benign
    - Hemangioma
    - Deep hemangioma (intramuscular, synovial, perineural)
    - Glomus tumor
  - Intermediate (hemangioendothelioma)
  - Malignant
    - Hemangiosarcoma
    - Malignant hemangiopericytoma
- Tumors of lymph vessels
  - Benign (lymphangioma)
    - Cavernous
    - Cystic (cystic hygroma)
  - Malignant
    - Lymphangiosarcoma
    - Postmastectomy lymphangiosarcoma
- Tumors and tumorlike lesions of synovial tissue
  - Benign
    - Giant cell tumor of tendon sheath
      - Localized (nodular tenosynovitis)
      - Diffuse (florid synovitis)
  - Malignant
    - Synovial sarcoma (malignant synovioma), predominantly biphasic (fibrous or epithelial) or monophasic (fibrous or epithelial)
    - Malignant giant cell tumor of tendon sheath
- Tumors and tumorlike lesions of peripheral nerves
  - Benign
    - Traumatic neuroma
    - Morton's neuroma
    - Neurilemoma (benign schwannoma)
    - Neurofibroma, solitary
    - Neurofibromatosis (von Recklinghausen's disease)
      - Localized
      - Plexiform
      - Diffuse
  - Malignant
    - Malignant schwannoma
    - Peripheral tumors of primitive neuroectodermal tissues
- Tumors and tumorlike lesions of cartilage and bone-forming tissues
  - Benign
    - Panniculitis ossificans
    - Myositis ossificans
    - Fibrodysplasia (myositis) ossificans progressiva
    - Extraskeletal chondroma
    - Extraskeletal osteoma
  - Malignant
    - Extraskeletal chondrosarcoma
      - Well-differentiated
      - Myxoid (chordoid sarcoma)
      - Mesenchymal
    - Extraskeletal osteosarcoma
- Tumors and tumorlike lesions of pluripotential mesenchyme
  - Benign mesenchymoma
  - Malignant mesenchymoma
- Tumors and tumorlike conditions of disputed or uncertain histogenesis
  - Benign
    - Tumoral calcinosis
    - Myxoma (cutaneous and intramuscular)
  - Malignant
    - Alveolar soft-part sarcoma
    - Epithelioid sarcoma
    - Clear cell sarcoma of tendons and aponeuroses
    - Extraskeletal Ewing's sarcoma
- Unclassified soft-tissue tumors and tumorlike lesions

*Adapted with permission from Enzinger FM, Weiss SW: *Soft Tissue Tumors.* Philadelphia, CV Mosby: 1983, pp 6–7.

Table 4
Surgical Staging System of the Musculoskeletal Tumor Society

Stage IA	Low-grade, intracompartmental
Stage IB	Low-grade, extracompartmental
Stage IIA	High-grade, intracompartmental
Stage IIB	High-grade, extracompartmental
Stage III	Any evidence of metastases

formed, plain orthogonal radiographs in two planes should obtained. Radiographs are helpful in establishing whether the soft-tissue mass is secondary to (1) a tumor arising from the bone, (2) a tumor arising on the surface of the bone, or (3) a tumor or tumorlike lesion arising primarily in the soft tissues.

When the clinician determines that the lesion is arising in the soft tissues, the radiograph should be carefully inspected with the following questions in mind: Is there evidence that the mass is eroding or destroying the underlying bone? Is there evidence of periosteal reaction? Is there evidence of mineralization within the soft-tissue lesion?

Table 5
Functional Classification of Malignant Soft-Tissue Sarcomas

Graded sarcomas
- Malignant fibrous histiocytoma
- Liposarcoma
- Leiomyosarcoma
- Neurofibrosarcoma

Nongraded sarcomas
- Synovial cell sarcoma
- Epithelioid sarcoma
- Clear cell sarcoma
- Alveolar soft-parts sarcoma
- Mesenchymal chondrosarcoma

Small cell neoplasms
- Rhabdomyosarcoma
- Soft-tissue Ewing's sarcoma
- Neuroblastoma
- Undifferentiated small cell sarcoma

Mineralization can occur within a soft-tissue lesion in several instances (Table 6), the most common of which is heterotopic ossification secondary to trauma (myositis ossificans). As the lesion matures, the mineralization usually appears at the periphery of the lesion, while the center does not mineralize. Hemangiomas will often have distinctive intralesional small phleboliths. Soft-tissue chondromas often will have stippled foci of mineralization.

Some malignant lesions may also demonstrate intralesional mineralization. One third to one half of synovial sarcomas are characterized by multiple small and spotty radiopacities caused by focal calcification and, less frequently, bone formation.[1] Well-differentiated liposarcomas occasionally have foci of calcification and ossification (Fig. 3). Extraskeletal myxoid chondrosarcoma and extraskeletal mesenchymal chondrosarcoma may show areas of calcification. Extraskeletal osteosarcomas will often show extensive bone formation within a soft-tissue mass.

Magnetic resonance (MR) imaging has become the most useful modality for the definition of soft-tissue masses.[4,5] The MR image provides excellent definition of normal muscle, fascial boundaries, and the tumor mass. Multiplanar (transverse, sagittal, and coronal) images can be obtained. Intravenous contrast agents are not necessary to evaluate neurovascular structures. It is important to remember that both T1- and T2-weighted sequences are essential to detect and characterize soft-tissue lesions.

Although the MR image can detect soft-tissue masses with a very high sensitivity, it is not possible to accurately predict the histology or whether a lesion is benign or malignant.[6-9] The two exceptions to this general rule are lipomas and hemangiomas. Lipomas often are very homogeneous and have signal characteristics that exactly match those of the surrounding fat, thus establishing the diagnosis. Hemangiomas contain numerous blood vessels and present with a recognizable pattern. Although accurate prediction of malignancy is not possible, an index of suspicion can be based on margination, homogeneity, effect on natural barriers, growth rate, matrix mineralization, and effect on adjacent soft tissues and bone.[10]

The reactive zone is less well defined and appears as a less dense (fuzzy) area between the main tumor mass and the normal muscle (Fig. 4). One can also determine the relationship between the tumor mass and the adjacent vascular structures, nerves, and periosteum.

Computed tomographic (CT) scans are useful in selected cases to identify patterns of mineralization within the soft tissues and erosion or destruction of underlying bone. Contrast-material-enhanced CT scans may be utilized to better delineate the anatomic features of soft-tissue masses.

A chest radiograph should also be obtained, because sarcomas most commonly metastasize to the lungs. Pulmonary metastases are usually asymptomatic initially. A CT study is useful in detecting occult pulmonary metastases when a malignant tumor is suspected.

Screening laboratory tests include complete blood cell count with differential, erythrocyte sedimentation rate, serum electrolytes, and chemistry panels including serum calcium and phosphate.

Biopsy

When the etiology of a soft-tissue mass is not apparent (e.g., lipoma),

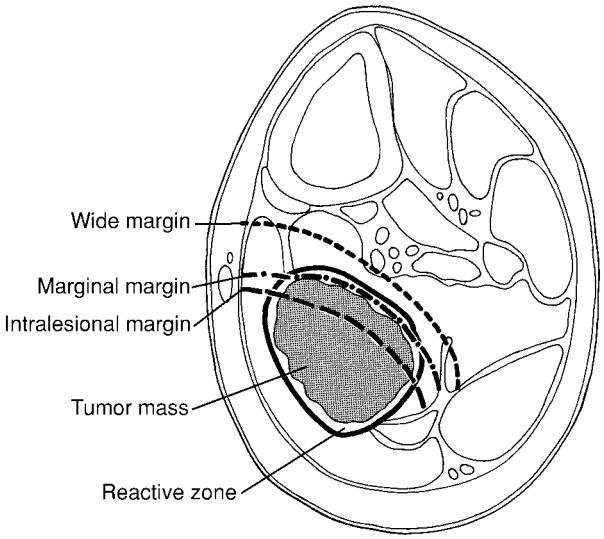


Fig. 2 Diagram of types of surgical margins. An intralesional line of resection enters the substance of the tumor. A marginal line of resection travels through the reactive zone of the tumor. A wide surgical margin removes the tumor with a cuff of normal tissue.

biopsy is often necessary. Biopsy is an important step in management; however, when done improperly, it can result in disastrous complications. There are three types of biopsy: needle biopsy, open incisional biopsy, and open excisional biopsy.

Needle biopsy (fine-needle aspirate or core) has the advantage of low morbidity with only a small skin incision. Unfortunately, the amount of tissue retrieved is small, and not all pathologists are comfortable interpreting such a small tissue sample. In addition, because the sample is so small, the pathologist may be unable to study the lesion with special stains, cytogenetic techniques, or electron microscopy. The fine-needle technique is often made more difficult by tissue heterogeneity and necrosis.

Open incisional biopsy is commonly employed, but several principles must be closely followed. The skin incision must be oriented so that the biopsy tract can be completely excised if the lesion is subsequently found to be malignant (Fig. 5). It is axiomatic that transverse and oblique incisions should be avoided. After outlining the biopsy incision, the surgeon should draw the incision that would be employed in the definitive surgery; in that way, if the lesion proves to be malignant, the orientation of the biopsy incision will allow later complete excision of the biopsy tract. Raising large flaps is to be avoided, and maintaining meticulous hemostasis is essential. Intermuscular planes and neurovascular bundles should also be avoided; it is most desirable to perform the biopsy through muscle when feasible.

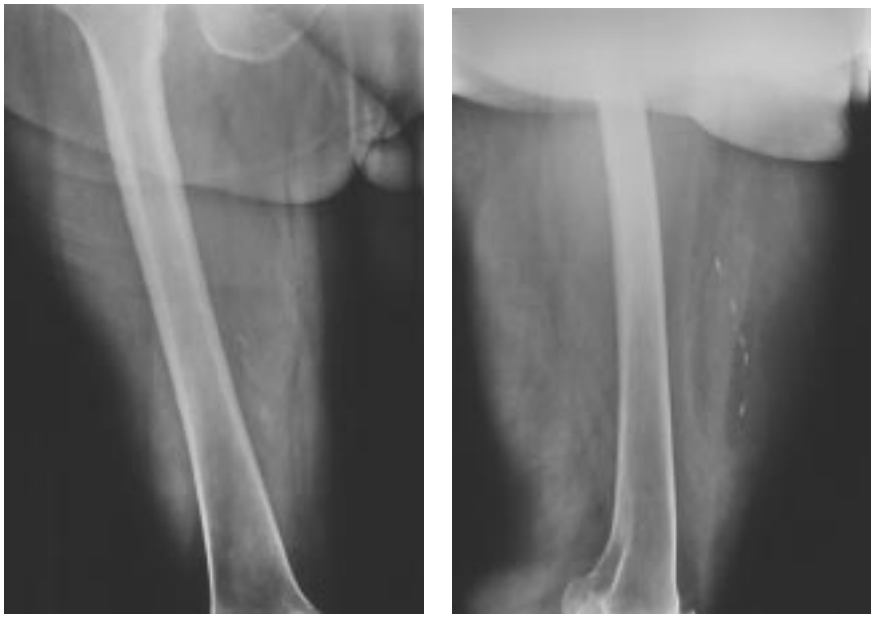
Frozen-section analysis should be performed to ensure that adequate diagnostic material has been obtained. If only the periphery of the lesion is sampled, the specimen may contain only reactive or inflammatory tissue.

A generous biopsy specimen should be obtained, taking care not to create excessive bleeding in an inaccessible hole. Many malignant tumors have large, friable vessels that tend to bleed excessively. If a tourniquet is used, it should be deflated to ensure adequate hemostasis prior to wound closure. If a drain is employed, it should be brought out at the corner of the wound in line with the incision (separated by about 5 to 10 mm). The muscle should be closed tightly. Sutures used to close the skin should be placed close to the incision (within 5 mm). A compression

Table 6
Disorders Associated With Extraskeletal Calcification or Ossification*

Metastatic calcification
Hypercalcemia
Milk-alkali syndrome
Hypervitaminosis D
Sarcoidosis
Hyperparathyroidism
Renal failure
Hyperphosphatemia
Tumoral calcinosis
Hypoparathyroidism
Pseudohypoparathyroidism
Cell lysis following chemotherapy for leukemia
Renal failure
Dystrophic calcification
Calcinosis (universalis or circumscripta)
Childhood dermatomyositis
Scleroderma
Systemic lupus erythematosis
Posttraumatic
Ectopic ossification
Myositis ossificans (posttraumatic)
Burns
Surgery
Neurologic injury
Muscle contusions
Fibrodysplasia (myositis) ossificans progressiva
Mineralization occurring within neoplasms
Benign
Hemangioma (small phleboliths)
Arteriovenous malformations (small phleboliths)
Malignant (synovial sarcoma)

*Adapted with permission from Favus MJ: *Primer on the Metabolic Bone Diseases and Disorders of Min -eral Metabolism*, 2nd ed. New York: Raven Press, 1993, p 386.



A B

Fig. 3 Anteroposterior **(A)** and lateral **(B)** plain radiographs demonstrate a large, low-density mass in the anterior thigh containing several foci of calcification.

dressing should be utilized to aid hemostasis. Antibiotics should be administered perioperatively and for 24 to 48 hours following surgery.

Excisional biopsy should be used only for small lesions and only when the surgeon is absolutely sure that the lesion is benign. Excisional biopsy has the disadvantage that a large wound is created. If the lesion is found to be malignant, it will be difficult to excise the entire biopsy tract.

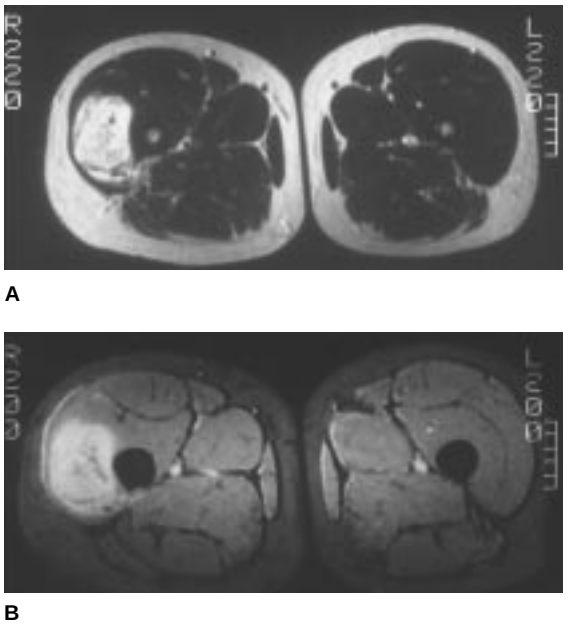
Regardless of the biopsy procedure performed, it is important to obtain complete cultures (aerobic and anaerobic bacteria, fungal, and tuberculosis), as inflammatory lesions may simulate a neoplasm.

There are many hazards associated with biopsy of soft-tissue masses, including infection, delayed wound healing, hematoma formation, and improper location or orientation of the incision. A study performed by the Musculoskeletal Tumor Society revealed that a wound complication occurred in 17% of 57 patients who underwent biopsy, and that the optimal treatment plan had to be altered in 18% of 60 such patients.[11] These problems occurred three to more than five times more frequently when the biopsy was performed at a referring institution rather than in a treating center. Simon[12] has outlined the principles of planning and biopsy technique.

Treatment

The treatment of soft-tissue masses is based on both the histologic diagnosis and the stage in the surgical staging system of the Musculoskeletal Tumor Society. Benign inactive lesions may require no treatment other than observation. Benign active lesions can often be removed with either an intralesional or a marginal line of resection. Benign aggressive lesions (e.g., desmoid tumors and large active hemangiomas) often require a wide margin with a cuff of normal tissue. Extra-abdominal fibromatosis (desmoid) tumors are difficult to treat and often require adjunctive radiation.

A multidisciplinary approach is utilized for malignant lesions, requiring the coordinated efforts of the orthopaedic oncologist, the radi-



A B

Fig. 4 Inhomogeneous mass seen in the vastus lateralis on T1-weighted **(A)** and gradient-echo **(B)** MR images suggests presence of a malignant neoplasm.

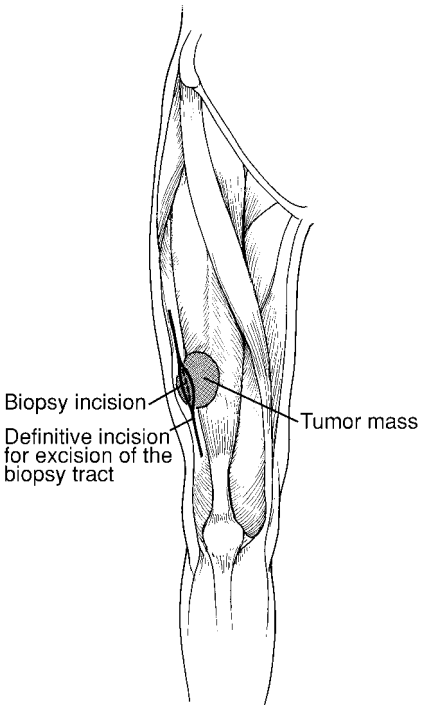


Fig. 5 Diagram of a lesion in the lateral aspect of the quadriceps mechanism. A short longitudinal incision is made over the lesion. Prior to incising the skin, a second incision line should be drawn, to demonstrate how the biopsy tract can be removed at the time of the definitive surgery.

ation oncologist, the medical oncologist, the plastic surgeon, and the thoracic surgeon.

Surgery of Malignant Lesions

When appropriate, limb salvage is the preferred technique for malignant extremity lesions. The two prerequisites for limb-salvage surgery are that (1) local control of the lesion will be at least equal to that achievable with amputation, and (2) the salvaged limb will be functional.

Preoperative planning is crucial to ensure success. The MR imaging and CT studies should be reviewed to accurately define the tumor volume in order to determine whether the lesion will be resectable with a limb-salvage procedure. The MR images are most useful in determining the size of the tumor, its boundaries and its relationships with adjacent structures (nerves, arteries, veins, fascia planes, and muscles). The CT study is most useful in determining whether there is any erosion or destruction of underlying bone. Angiography can be performed to define the vascularity of the lesion and to detect encasement of a major vessel. As the resolution of MR imaging has improved, the indications for angiography have diminished.

The surgical procedures are designed to remove the lesion with a cuff of normal tissue (wide surgical margin). If a major vessel is encased by the tumor, it may be necessary to resect and reconstruct the vessel. If cortical bone destruction is present, the involved bone must also be removed with a wide margin. If the major nerves of the limb are surrounded by tumor, amputation is probably necessary, because the limb will not be functional with a limb-salvage procedure.

The second phase of surgery is reconstruction. The surgical defect must be carefully closed to minimize the risk of fluid collections and delayed wound healing. When necessary, large defects should be closed with either local rotational muscle flaps or free microvascular tissue transfers. Split-thickness skin grafts should be utilized when there is a defect with underlying healthy muscle.

Radiation Therapy

Radiation therapy plays a major role in the treatment of soft-tissue sarcomas following limb-salvage surgery. Although surgery alone may yield good results in patients with small lesions,[13] soft-tissue sarcomas are often very large and located too close to major nerves, vessels, and bone to obtain sufficient margins. The use of adjuvant irradiation in the pre- or postoperative period allows the surgeon to conserve normal tissue without compromise of local control or ultimate survival.[14-16]

Irradiation can be delivered with the use of (1) a high-energy external beam in the pre- and/or postoperative period, (2) brachytherapy utilizing afterloading catheters placed during the operative procedure, (3) intraoperative electron therapy, or (4) a combination of these procedures. External-beam techniques are the most widely available and most commonly used. The use of high-dose postoperative irradiation (60 to 65 Gy) is associated with a decreased risk of wound complications, but generally treatment with larger fields is required because the entire surgical bed must be included. Compared with postoperative therapy, preoperative treatment often improves the resectability of lesions, allows treatment of smaller volumes, and has been associated with better local control rates for larger lesions.[14]

Brachytherapy has been used to deliver the total radiation dose[15] with excellent results. However, many lesions are not amenable to a primary en bloc resection without the sacrifice of crucial structures (e.g., vessels, nerves, tendons). There is also concern about dose homogeneity with large-volume implants. Therefore, brachytherapy and intraoperative techniques are most often used as a substitute for a portion of the external-beam treatment. These techniques allow delivery of a high dose of radiation to a well-defined area and can be done at surgery or in the immediate postoperative period rather than waiting 4 to 6 weeks for adequate wound healing before additional external-beam treatment. In the case of large or marginally resectable lesions, preoperative external-beam radiation (50 to 55 Gy) is generally used, followed by an additional 10 to 15 Gy of radiation delivered intra- or perioperatively to areas of close margins. If these tech-

niques are not feasible or available, an additional 15 Gy may be given to a boost field by means of an external beam. Local control rates with a combined-modality approach have been reported to be 90% or greater.[13,15,16] However, combined-modality treatments are not without potential complications; the complication rate may approach 30%, especially with very large lesions treated with preoperative irradiation.

In cases in which an excisional biopsy reveals a high-grade soft-tissue sarcoma and MR imaging reveals no evidence of gross residual disease, reoperation with placement of afterloading catheters and delivery of 15 to 20 Gy of radiation followed by 45 Gy of postoperative external-beam treatment may be used. Preoperative external-beam irradiation alone is an alternative in this situation.[17]

Chemotherapy

The role of adjuvant chemotherapy in the treatment of high-grade soft-tissue sarcomas (with the exception of Ewing's sarcoma and rhabdomyosarcoma) continues to be the subject of investigation. Only two prospective, randomized trials of adjuvant chemotherapy in extremity lesions have shown improvement in disease-free and overall survival.[18,19] Other trials have not shown any significant benefit.[20-22] At present, the lower rate of metastatic spread with low-grade lesions may not justify the potential risks of chemotherapy.

Preoperative intra-arterial chemotherapy with or without irradiation also has been studied in a number of institutions, but the benefit of these techniques to later survival has not yet been established in randomized trials.[23]

Adjuvant chemotherapy given preoperatively, both pre- and postoperatively, or postoperatively is being studied prospectively in a number of institutions. Effective chemotherapy agents and regimens continue to be sought as a method of improving survival, as has been documented in patients with intramedullary osteosarcoma, Ewing's tumor, and rhabdomyosarcoma.

Follow-up

Patients should be monitored closely following treatment and then at 3-month intervals for 2 years with careful physical examination to detect local recurrence. A baseline MR imaging study should be obtained 3 months after surgery; MR imaging should then be performed at 1-year intervals for 5 years thereafter.

Chest radiographs and CT scans should be obtained at 3-month intervals for 2 years and then at 6-month intervals for 6 years. At 8 years after surgery, they should be obtained once a year.

Prognosis

The prognosis for the individual patient depends on the grade and size of the tumor and the absence or presence of metastases. Large (greater than 5 cm in diameter) and high-grade lesions have a high potential for metastasis. Pulmonary metastases develop in as many as 50% of patients with high-grade lesions, and these patients subsequently die of the disease. The overall 5-year survival rate for patients with high-grade lesions but only localized disease is approximately 70% to 80%.

Patients who have pulmonary metastases at presentation or within 6 months of diagnosis have an extremely poor prognosis, with only the rare long-term survivor. Pulmonary resection of metastases is feasible when there are no extrathoracic metastases and the primary tumor is under control.[24] Patients in whom pulmonary metastases develop 1 year after tumor resection may be cured with multiple thoracotomies in about 25% of cases.

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