

Soft-Tissue Tumors About the Knee

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Abstract

Soft-tissue tumors about the knee include a wide variety of entities, ranging from synovial cysts to aggressive high-grade sarcomas. The overlap in clinical presentation of these various masses frequently results in misdiagnosis and inappropriate treatment. Unnecessary and sometimes costly arthroscopy sometimes precedes the diagnosis of soft-tissue sarcoma about the knee. A poorly planned or executed biopsy has been demonstrated to have an adverse effect on patient prognosis and may lead to unnecessary amputation. Special vigilance in evaluation is warranted when a soft-tissue mass is not in the typical position or does not have other characteristic features of a meniscal or Baker's cyst, when the size of the mass or the accompanying symptoms seem out of proportion to the injury or underlying degenerative process, and when symptoms persist beyond what is expected. When malignancy is suspected, the patient should be referred to a musculoskeletal oncologist before biopsy.

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The evaluation and treatment of soft-tissue tumors about the knee should be familiar to all who treat musculoskeletal problems of this joint, from the general orthopaedic surgeon to the sports medicine specialist and the adult reconstructive surgeon. The wide array of processes that may present as a soft-tissue mass about the knee can make diagnosis difficult. Benign soft-tissue masses, such as synovial cysts, may be related to knee injuries and internal derangement. In addition, the far more common sports- and occupation-related injuries frequently coexist and may even bring attention to the existence of an underlying neoplasm. Many patients with soft-tissue sarcomas will attribute the onset of a mass to an injury. Unfortunately, this overlap in presentations may also delay diagnosis and treatment of malignant soft-tissue tumors that masquerade as injuries and cysts.

Failure to consider the possibility of neoplasm may lead to unfortunate mistakes. Lewis and Reilly¹ underscored this important point in their review of 36 "sports tumors"—neoplasms that were misdiagnosed initially as sports-related injuries. Half of those tumors were soft-tissue neoplasms, with synovial sarcoma being the most frequent. Alarming, 70% of the patients with malignant neoplasms in the region of the knee had undergone arthroscopy or arthrography before the correct diagnosis was established.¹ Inappropriate initial management of soft-tissue sarcomas can contribute to a poor overall prognosis and unnecessary amputation.²

In this article, we will present a diagnostic approach, the differential diagnosis, and the treatment principles for soft-tissue tumors about the knee.

Diagnostic Approach

Evaluation of any patient with a soft-tissue mass should proceed in an orderly and systematic fashion. The history and the physical examination provide essential diagnostic and often important prognostic information that complements the findings from subsequent radiologic studies.

Historical Features

The age of the patient is particularly helpful in the differential diagnosis of a soft-tissue mass about the knee.^{3,4} In children and adolescents, hemangiomas and vascular malformations, deep and superficial, are common soft-tissue tumors. The only synovial process in the knee with a peak occurrence in childhood and adolescence, apart from more diffuse rheumatologic conditions, is synovial hemangioma.⁵ The most common soft-tissue sarcoma in this young age group is rhabdomyosarcoma.³

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In adults, the most common deeply seated soft-tissue mass about the knee is an intramuscular lipoma, but hemangiomas, nerve-sheath tumors, and desmoids are relatively common as well.⁴ Pigmented villonodular synovitis most commonly occurs in young adults. Synovial chondromatosis has its peak occurrence in middle-aged adults. The most common soft-tissue sarcoma about the knee in the young adult is synovial sarcoma, whereas the older adult will most commonly be affected by a malignant fibrous histiocytoma or liposarcoma.³

Symptoms

Excluding neoplastic simulators, the vast majority of soft-tissue true neoplasms, including sarcomas, will be asymptomatic other than the presence of an enlarging mass.³ The remaining 10% to 15% may be associated with pain, tenderness, and other symptoms referable to the knee joint itself, including swelling, locking, popping, and buckling. In contrast, benign synovial processes are far more likely to cause joint symptoms. Both pigmented villonodular synovitis and the rare synovial hemangioma will produce recurrent painful hemarthroses.^{5,6} Synovial chondromatosis characteristically produces loose-body symptoms in addition to joint pain and swelling.⁷ Baker's cysts and meniscal cysts are frequently also associated with joint symptoms indicative of an underlying meniscal tear or degenerative change; proximal tibiofibular cysts classically are not.⁸ Patients with hemangiomas commonly experience engorgement, which is often painful, after exercise or simple dependency of the limb.

Occasionally, a predisposing condition that places the patient at increased risk for the development of a soft-tissue neoplasm will be

present. Genetic conditions such as neurofibromatosis, polyposis coli, Li-Fraumeni syndrome, retinoblastoma, and xeroderma pigmentosa are rare causes of various benign and malignant soft-tissue neoplasms.³ Prior radiation exposure and a chronic lymphedematous extremity are accepted risk factors for the development of soft-tissue sarcomas. The presence of human immunodeficiency virus infection makes Kaposi's sarcoma more likely. Both fibrosarcomas and malignant fibrous histiocytomas have been known to occur in the scars of thermal and acid burns.³

Physical Examination

Soft-tissue masses should be evaluated by inspection, palpation, auscultation, percussion, and thorough neurovascular and lymph node assessment. Only rarely will inspection identify specific overlying skin changes, such as those observed with superficial hemangioma, lymphangiosarcoma, dermatofibrosarcoma, and Kaposi's sarcoma. Rhabdomyosarcoma may give the appearance of a muscle bulging through the skin.

Palpation will reveal the size, location, and deep or superficial position of the mass. Most soft-tissue sarcomas are discretely palpable, whereas rhabdomyosarcomas are often poorly defined on examination, much like the often palpably boggy hematoma. If the mass is larger than 5 cm or deep to the subcutaneous tissue, it is much more likely to be malignant.⁹ While the converse is also true, one should remember that a third of soft-tissue sarcomas are located in the subcutaneous tissue.¹⁰ Warmth as a manifestation of hyperemia is observed with soft-tissue sarcomas and inflammatory processes, but rarely with benign neoplasms.

Position relative to the knee joint line is important in differentiating

other masses from meniscal cysts; however, it must be remembered that benign cysts may dissect well away from the joint, and that superficial soft-tissue sarcomas may lie directly over the joint line. Popliteal masses must be carefully palpated to avoid dismissing a soft-tissue sarcoma masquerading as a Baker's cyst. The stalk of a Baker's cyst typically originates posteromedially and extends between the semimembranosus and the medial head of the gastrocnemius.

Auscultation may identify a bruit in an aneurysm or a high-flow arteriovenous malformation. Percussion may reveal a Tinel's sign suggestive of a peripheral nerve-sheath tumor or nerve involvement by the mass. Transillumination may be useful, particularly if a superficial mass is suspected of being cystic. Distal neurovascular examination is crucial in identifying involvement preoperatively, although the presence of abnormalities is unusual even with large masses and nerve-sheath tumors. A thorough lymph node examination in the proximal extremity may reveal adenopathy, which is more commonly seen with extremity lymphomas than with the rare sarcoma.

Laboratory Evaluation

Laboratory tests are entirely nonspecific relative to soft-tissue masses. The erythrocyte sedimentation rate and the white blood cell count are usually normal in the absence of a concomitant infectious or inflammatory process. The serum lactate dehydrogenase concentration is typically markedly elevated in patients with a lymphoma. A rheumatology screen may be appropriate when considering synovial processes, as may laboratory assessment of diabetic control when diabetic skeletal muscle necrosis seems likely.

Radiologic Evaluation

Radiologic studies of soft-tissue masses provide important information for determining the need for biopsy and defining the extent of the mass and its relationship to vital structures. However, these studies provide a specific diagnosis in relatively few cases.

Plain radiographs should always be obtained first. Most soft-tissue masses are depicted on radiographs as only a soft-tissue shadow. More specific positive findings include fat density, calcification, ossification, and bone involvement. Fat density reflects the tissue type but does not distinguish lipomas from lipomalike liposarcomas. Calcification may represent phleboliths of a hemangioma, loose bodies associated with synovial chondromatosis, or mineral deposition within a sarcoma. Ossification within the soft tissues about the knee is most commonly due to myositis ossificans, although soft-tissue osteosarcomas may produce a similar picture.

The bones in the knee region may be involved by synovial processes as well as benign and malignant neoplasms. Juxta-articular erosions on both sides of the joint in the presence of a suprapatellar pouch and/or Baker's cyst filled with boggy soft tissue or loose bodies should bring pigmented villonodular synovitis or synovial chondromatosis to mind. Cortical hyperostosis often accompanies deep intramuscular hemangiomas chronically juxtaposed to the bone. Visible periosteal reaction from a soft-tissue sarcoma is rare; if noted, the more sensitive technetium bone scanning should be used to further evaluate osseous involvement.

Ultrasound may be helpful in identifying purely cystic masses. A purely cystic mass about the knee that is less than 5 cm in size and is

located superficially is highly unlikely to be malignant.¹¹ Unless magnetic resonance (MR) imaging is indicated for evaluation of a potential concomitant intra-articular disorder or for preoperative planning, further imaging of such a mass is not routinely necessary.

When further imaging is necessary after plain radiography, MR imaging is the modality of choice in the vast majority of cases. The MR imaging examination of soft-tissue masses is analogous to the plain-film examination of bone lesions in that these studies yield the most diagnostic information for those conditions. However, MR imaging is often inconclusive as to a specific diagnosis.¹² The need for imaging of a soft-tissue mass instead of or in addition to the knee joint itself should be communicated to the radiologist because a routine MR study of the knee may not provide adequate information about the mass. Specific techniques not usually performed in an MR study done for meniscal or ligamentous injury, such as gadolinium enhancement and fat suppression, may be requested by the well-informed radiologist aware of the presence of a soft-tissue mass.

Unfortunately, MR imaging cannot be relied on with absolute certainty to distinguish benign from malignant soft-tissue masses.¹² Features generally considered to be associated with malignancy, such as lack of encapsulation, irregular borders, heterogeneous signal, neurovascular involvement, large size, and edema, are far less than completely predictive, as they occur in both benign and malignant soft-tissue processes.

Nuclear medicine studies play only a minor role in the evaluation of soft-tissue masses. At our institutions, a baseline total-skeleton bone scan is obtained only during baseline staging of a soft-tissue sarcoma.

Biopsy

The goal of biopsy is to obtain sufficient tissue to allow the pathologist to make a diagnosis while avoiding complications that may adversely affect subsequent surgery and prognosis. When feasible, biopsy should be performed at a tertiary institution by an experienced musculoskeletal oncologist.² Careful preoperative planning and adherence to strict biopsy principles should be the rule.^{13,14}

The options for biopsy include fine-needle aspiration, closed core-needle biopsy, and open incisional biopsy. The advantages of needle biopsies include the reduced cost when done outside the operating room and reduced morbidity to the patient.¹⁵ The main disadvantage of needle biopsies is that they provide less tissue, which causes some pathologists to be less confident in interpretation. Strict guidelines must be followed to avoid potentially disastrous complications, particularly when an open incisional biopsy is performed.

When planning the biopsy incision, one must bear in mind the incision to be subsequently used for resection. Ideally, the biopsy incision should be oriented longitudinally directly overlying the most superficial aspect of the soft-tissue mass. Approximately 0.5 to 1.0 cm³ of tissue should be obtained without contaminating noninvolved compartments. Tissue flaps should not be developed; instead, sharp dissection should be carried down directly to the mass. Tissue from the periphery should be submitted to ensure viability.

Meticulous hemostasis should be obtained. A tourniquet may be utilized, but it should be released before closure to ensure hemostasis. Drains, when used, should exit in line with, and just a few millimeters away from, the end of the inci-

sion. Frozen section is generally recommended to ensure that sufficient tissue has been obtained. Tissue should generally be submitted fresh, rather than in formalin, unless the latter is specifically requested by the pathologist. If lymphoma or another small blue round cell tumor is being considered in the diagnosis, tissue should be set aside for immunocytochemical analysis. The surgeon should ascertain from the pathologist whether additional tissue is needed for electron microscopy.

Differential Diagnosis

Soft-tissue masses about the knee may be grouped into three broad categories: neoplastic simulators, benign neoplasms, and malignant neoplasms. Neoplastic simulators include juxta-articular cysts, synovial processes, and myriad other less common diagnoses. The benign neoplasms that occur in this area are reflective of the long list of these processes that occur in nearly all extremity locations. Those more frequently seen about the knee include hemangiomas, vascular malformations, and lipomas. The malignant process most commonly thought of is the soft-tissue sarcoma, but lymphomas are not uncommon in the extremities. In rare instances, metastatic disease can involve the soft tissues.

Neoplastic Simulators

Juxta-articular cysts most commonly occur in one of three forms: Baker's (popliteal) cyst, meniscal cyst, and proximal tibiofibular synovial cyst. A characteristic historical feature of each of these processes is a waxing and waning course or even disappearance and reappearance over time. When large and superficial enough, cysts may be transilluminable.

The Baker's cyst typically occurs in the presence of intra-articular injury, synovitis, or frank arthritis. Its diagnosis in the absence of an intra-articular lesion in an adult should be considered suspect. Baker's cysts originate from the tibiofemoral portion of the joint and can typically be palpated posteromedially. The cyst most commonly presents as a painless mass. The purely cystic nature may be confirmed with ultrasound or MR imaging (Fig. 1). Less than purely cystic characteristics on ultrasound or any heterogeneity on MR imaging should cast doubt on the diagnosis.

The meniscal cyst occurs in association with a meniscal tear, originates at the tibiofemoral joint level, and may vary in prominence with changes in position of the knee. Typically, a meniscal cyst may manifest no more than a bulge at the joint level; in unusual cases, it may present as a larger process medial or lateral to the joint.

The proximal tibiofibular synovial cyst is commonly symptomatic

and may progress to peroneal nerve dysfunction, erosion of the tibia or fibula, or compartment syndrome.⁸ In one series, 50% of such cysts intimately involved the peroneal nerve.⁸ Magnetic resonance imaging is nearly diagnostic, although nerve-sheath tumors may have an identical appearance.

An intra-articular synovial process may present as a painful soft-tissue swelling or mass, as the source of mechanical symptoms or effusion in the knee, or as an incidentally noted arthroscopic finding.

Pigmented villonodular synovitis classically presents as recurrent monarticular hemarthroses of the knee in a young adult (Fig. 2).⁶ It is best described as a locally invasive benign synovial proliferation of unknown etiology, which may occur in diffuse and localized forms. In the diffuse form, plain radiographs may reveal a soft-tissue mass with juxta-articular erosions. The localized form may appear as a focal pigmented area under arthroscopic visualization.

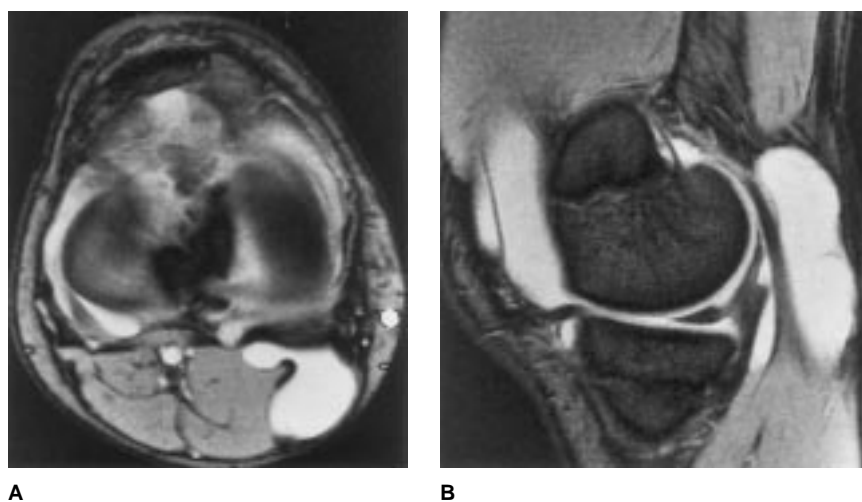


Fig. 1 A classic Baker's cyst accompanied by intra-articular effusion as demonstrated on axial (A) and sagittal (B) T2-weighted gradient-echo MR images of the knee of a 17-year-old female basketball player. Note that the purely homogeneous fluid signal within the cyst is identical in characteristics to that of the effusion.

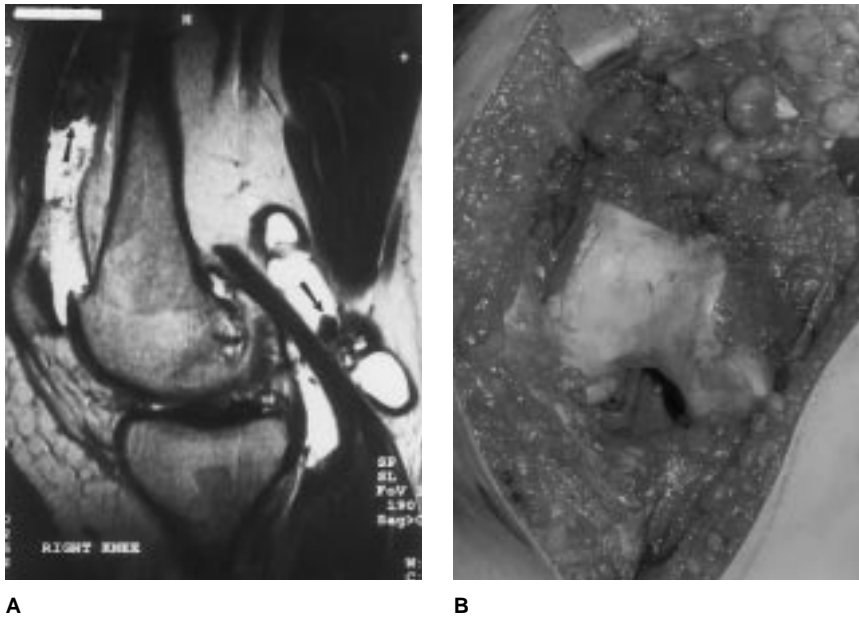


Fig. 2 A, T2-weighted sagittal MR image of a 26-year-old woman with recurrent hemarthroses and painful limited range of motion in the knee. Image shows multiple low-signal densities throughout the knee, most prominent in the suprapatellar pouch (arrow at left) and the associated popliteal cyst (arrow at right). B, At open synovectomy, the gross appearance was characteristic of pigmented villonodular synovitis.

The MR imaging appearance of low signal intensity within the solid portions on all sequences, due to signal attenuation by the hemosiderin deposition, is highly suggestive. The diagnosis is confirmed if a synovial biopsy specimen demonstrates villous synovial hypertrophy with a background of mononuclear cells, giant cells, lipid-laden macrophages, and hemosiderin deposition.⁶

Synovial chondromatosis is a metaplastic synovial proliferative process characterized by the formation of cartilaginous and secondarily mineralized or ossified loose bodies, typically in middle-aged adults.^{3,7} Eighty percent of cases involve the knee. The loose bodies often produce mechanical symptoms in addition to pain, swelling, and diminished motion. The mineralized and ossified loose bodies are apparent on plain films and

may be accompanied by juxta-articular erosions or even frank degenerative changes in the most advanced cases. Even in the absence of plain-film findings, MR imaging will demonstrate the characteristic loose bodies. Arthrography will demonstrate filling voids. Histologically, the synovium exhibits characteristic cartilage metaplasia.³

Synovial hemangioma is a rare condition most commonly seen in children and young adults, which presents with a characteristic history of recurrent, atraumatic, painful monarticular hemarthroses nearly exclusively involving the knee joint.⁵ A spongy, compressible mass overlying the knee that diminishes in size with elevation may be identified. The plain-radiographic findings are nonspecific. Arteriography was used in the past to define the extent of this

process, but MR imaging is currently the modality of choice. Both localized or pedunculated tumors and diffuse lesions have been described. The histologic picture is that of a cavernous hemangioma, although the chronic intra-articular bleeding may create a reactive synovitis with villous synovial hypertrophy and hemosiderin deposition.⁵

Synovial lipoma in the knee occurs in one of two forms: the true intra-articular lipoma and the intra-articular lipomalike lesion (lipoma arborescens), which consists predominantly of hypertrophic synovial villi distended by fat.³ The true intra-articular lipoma occurs as a solitary, slow-growing, discretely defined, purely fatty mass located typically in the infrapatellar fat pad. Symptoms attributable to these masses are often vague. The lipoma arborescens lesion is associated with rheumatoid, degenerative, and posttraumatic knee arthritis and frequently involves the suprapatellar pouch.³

Other soft-tissue neoplastic simulators are numerous and include examples from most categories of disease: metabolic, endocrine, traumatic, infectious, and autoimmune. A uniquely metabolic and endocrine tumor simulator poorly described in the orthopaedic literature, but better known to radiologists and diabetologists, is diabetic skeletal muscle necrosis (Fig. 3). This process, which nearly always involves the distal quadriceps musculature, most commonly affects patients with long-standing insulin-dependent diabetes. The patients classically present with a history of painful distal thigh swelling for 4 weeks or less. On examination, a tumoriform, exquisitely tender, palpable soft-tissue mass may be present, most commonly over the vastus medialis obliquus. Com-

puted tomography or MR imaging shows muscle swelling, rather than a true soft-tissue mass. Often, several individual muscles are involved. In almost all cases, some portion of the quadriceps musculature is affected.

Trauma-induced hematomas, particularly when chronic, may be confused with soft-tissue tumors clinically and on MR imaging. When recognized early after the inciting event, however, diagnostic evolution of MR imaging signal characteristics obviates the need for biopsy. Myositis ossificans frequently occurs in the quadriceps musculature after blunt trauma. A peripheral pattern of osteoid maturation within the mass is highly suggestive of the diagnosis in the appropriate clinical setting. In the absence of a discrete traumatic precursor, however, mineralized deep soft-tissue masses about the knee should be viewed with cau-

tion, as numerous soft-tissue sarcomas, particularly soft-tissue osteosarcoma and synovial sarcoma, may produce a similar appearance (Fig. 4).

Juxta-articular myxoma is an accumulation of mucinous material most frequently found about the knee, commonly in association with meniscal cysts and severe osteoarthritis. It may be seen incidentally during total knee arthroplasty. The origin of juxta-articular myxoma is unknown, but a traumatic source is suspected. These myxomas occur in adults as a painful and tender juxta-articular swelling or mass at or about the joint line. Histologically, the myxoma is composed of fibroblastlike cells within a myxoid matrix surrounded by a fibrous capsule. Hypercellularity and slight pleomorphism should not be mistakenly considered signs of malignancy.

Infectious tumor simulators include abscesses and lymphadenitis, which are usually identifiable on clinical examination by the cardinal signs of infection and palpable lymphadenopathy. Autoimmune causes include rheumatoid arthritis, with its rheumatoid nodules, and erythema nodosum, with its similarly subcutaneous and classically pretibial nodules. Historical features and dermal involvement are usually present to distinguish such rheumatologic conditions from superficial soft-tissue neoplasms.

Benign Neoplasms

The array of soft-tissue neoplasms that occur about the knee is as vast as that in any other region of the body. Despite recent advances in imaging techniques, a specific diagnosis is not possible in most cases without biopsy and histologic examination. Exceptions include hemangiomas, vascular



Fig. 4 Lateral radiograph of the knee of a 38-year-old man shows a mineralized soft-tissue mass in the popliteal region. The mass proved to be a mesenchymal chondrosarcoma, although similar soft-tissue mineralization may be seen with multiple soft-tissue sarcomas, particularly synovial sarcoma.

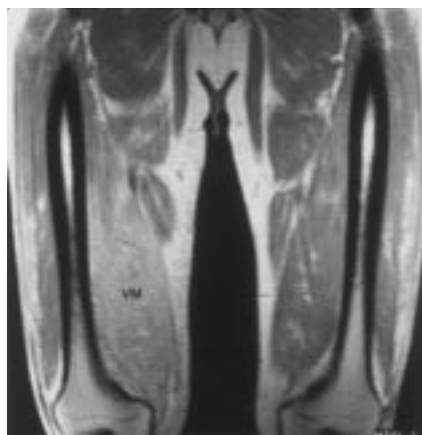


Fig. 3 A 54-year-old man with adult-onset insulin-dependent diabetes presented with a 4-week history of painful swelling in the right anteromedial thigh, which was exquisitely tender on examination. Coronal T1-weighted MR image of the thighs shows diffuse swelling of the right vastus medialis muscle (VM). Biopsy specimen showed only necrotic skeletal muscle, confirming the diagnosis of diabetic skeletal muscle necrosis.

malformations, and lipomas.¹² Extra-abdominal desmoids and benign nerve-sheath tumors may also be strongly suspected on the basis of the characteristic MR imaging findings. These entities will be discussed individually.

Hemangiomas and vascular malformations are among the most common soft-tissue tumors and are the tumors most frequently observed in infancy and childhood.^{3,4} The distal thigh is a common site of involvement. These processes have protean manifestations. The deep forms may present simply as a soft-tissue mass with normal overlying skin. The superficial forms have the telltale blue, purple, or red skin changes suggestive of their vascular origin. A palpable thrill or an audible bruit over the mass and increased warmth of the overlying skin are often diagnostic signs in patients with clinically significant shunting. Increased limb girth and venous enlargement are not uncommon, and limb hypertrophy

may be present. Plain radiographs may show phleboliths and curvilinear calcifications, and a soft-tissue shadow, as well as hypertrophy, bowing, chronic benign solid periosteal reaction, and cortical hyperostosis in the immediately adjacent bone (Fig. 5). Irregular mixed signal intensity is seen on MR imaging, reflective of the interspersed fat, fibrosis, fibrofatty septa, and numerous vessels. Histologic examination will identify the tissues of origin. As categorization of the process on the basis of vessel type and size does not change treatment, biopsy is typically unnecessary.³

Lipomas are not particularly common in the knee region, being more likely to occur in the torso and proximal extremities. However, lipomas are undoubtedly the

most frequent soft-tissue tumor; therefore, their occurrence about the knee cannot be considered unusual. Subcutaneous lipomas far outnumber deep ones overall. The thigh is the most common location for intramuscular and intermuscular lipomas. Their occurrence before age 20 is very unusual.

The soft, doughy consistency of subcutaneous lipomas is characteristic. Deep intramuscular lipomas may be palpable only when muscle contraction converts the tumor into a firm mass. After an initial growth phase, lipomas do not typically enlarge dramatically. Their size may increase with weight gain. Weight loss may also accentuate their presence, as the fatty tissue within these tumors is relatively unavailable for metabolism. Plain radiographs of deep lipomas may show a globular radiolucent mass clearly delineated from the surrounding higher-density muscle tissue shadows. Both computed tomography and MR imaging will show homogeneous fatty signal within a discretely margined lesion. Fibrous tissue septa may be present in lipomas but are conspicuously absent from liposarcomas. Any other heterogeneity in a predominantly fatty tumor should cast doubt on the benignity of the process. On histologic examination, the lipoma is seen to be composed of mature lipocytes. No lipoblasts or cells with atypical nuclei should be identified.

Approximately one eighth of extra-abdominal desmoids occur in the thigh or popliteal fossa.¹⁶ This common, locally aggressive benign fibrous-tissue neoplasm usually presents as a deep, very firm, slowly growing, painless mass in patients between skeletal maturity and age 40. Pain is usually indicative of neurologic involvement, as

can be observed with tumors in the popliteal fossa. The visualization on T1- and T2-weighted MR images of a dense, low-signal-intensity mass contiguous with the fascia is very suggestive, but only a biopsy specimen showing interlacing bundles of consistently benign-appearing fibroblasts separated by large amounts of collagen will confirm the diagnosis.^{3,16}

The most common benign peripheral nerve-sheath tumor in the knee region is the neurilemoma (benign schwannoma). Despite its neurogenic origin, pain and neurologic symptoms are uncommon until the tumor becomes quite large. Most commonly, at presentation the neurilemoma is a slowly enlarging, painless mass on the flexor surface that has been noted for a year or more. Occasionally, some waxing and waning in size may be noted, attributable to variation in the relatively frequent cystic degeneration that occurs within neurilemmomas. The mobility of these masses is dependent on the size of the nerve of origin. Mobility in all directions is typical of smaller-nerve involvement, whereas restricted movement is noted in the longitudinal axis of a larger nerve. When neurilemmomas originate on identifiable larger nerve branches, their eccentric location relative to the nerve fibers is characteristic and aids in their easy separation without nerve sacrifice.

An MR imaging study may suggest a nerve-sheath tumor, particularly when an apparently encapsulated mass is in continuity with an identifiable nerve. Cystic fluid-filled regions within larger neurilemmomas are often noted on MR images. The classic histologic finding of a neurilemoma is an alternating pattern of Antoni A (highly cellular spindle cell) areas and Antoni B (loosely arranged myxoid) areas.



Fig. 5 Vascular malformation immediately juxtaposed to the tibia resulted in characteristic cortical hyperostosis and bowing of the tibia, as shown on this lateral radiograph. Limb hypertrophy and overgrowth were apparent on clinical examination.

Most solitary neurofibromas occur in the subcutaneous tissue, although they may also arise within major nerves. Neurofibromas are no more frequent in the knee region than elsewhere. Like neurilemmomas, these tumors are most common in the young adult and are suspected on MR imaging only if there is demonstrable continuity with a peripheral nerve. Unlike neurilemmomas, neurofibromas are not encapsulated and are histologically composed of a monophasic population of irregular, spindle-shaped cells. Their complex interdigitation with the nerve fibers of major nerves renders complete extirpation without nerve sacrifice extremely difficult. However, one should always attempt to preserve neurologic function. Careful dissection will often allow removal of a neurofibroma without a major resultant neural deficit.

Malignant Soft-Tissue Neoplasms

Malignant neoplasms in the soft tissues include soft-tissue sarco-

mas, lymphomas, and metastatic tumors, in descending order of frequency. Soft-tissue sarcomas are by far the most common of these in the knee region. The most prevalent soft-tissue sarcomas overall are malignant fibrous histiocytoma, liposarcoma, rhabdomyosarcoma, and synovial sarcoma. Synovial sarcomas are particularly common in the vicinity of the knee, where approximately one third of these tumors occur (Fig. 6). Both malignant fibrous histiocytoma and liposarcoma occur most commonly in the thigh region, including the knee. Rhabdomyosarcomas are relatively unusual in the extremities and show no predilection for the thigh or knee region.

Although soft-tissue sarcomas are not reliably differentiated radiographically, the prevalence of these tumors differs notably between age groups.³ In the older adult, malignant fibrous histiocytoma and liposarcoma are the most common. The peak incidence of malignant fibrous histiocytoma

occurs in the 50- to 70-year age group. The diagnosis should be made with caution in patients younger than 20. The peak incidence of liposarcoma occurs at 40 to 60 years. Liposarcomas are nearly unheard of in infants and children. Synovial sarcoma has a peak incidence in the 15- to 40-year age group. It may be seen in newborns, but it is unusual beyond age 50. Rhabdomyosarcoma is the most common soft-tissue sarcoma in childhood and adolescence. It is quite unusual beyond age 30.³

A soft-tissue sarcoma most commonly presents as a slowly or progressively enlarging, but otherwise asymptomatic, deep-seated mass of a few to several months' duration. One third of soft-tissue sarcomas present as subcutaneous masses (Fig. 7).¹⁰ Only approximately 10% to 15% of these tumors will elicit pain, tenderness, or other local symptoms. Synovial sarcomas about the knee are particularly prone to misdiagnosis, perhaps due to the relatively more

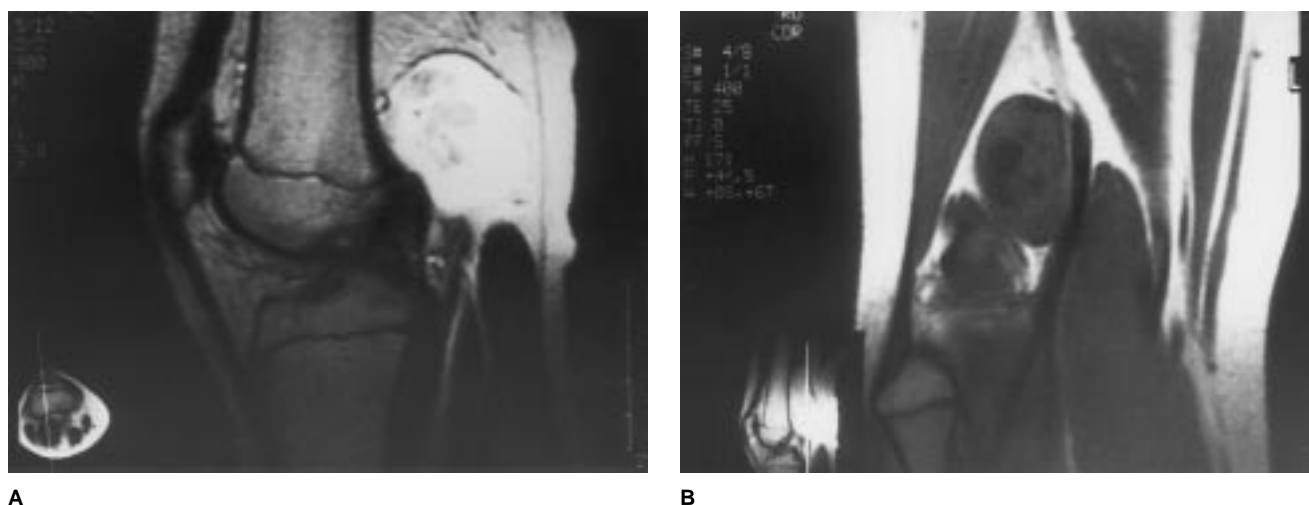


Fig. 6 Sagittal T1-weighted (A) and coronal T2-weighted (B) MR images of the knee of a 17-year-old female volleyball player. The clinical impression was a Baker's cyst, but the heterogeneity of the large, deeply seated mass was inconsistent with a synovial cyst. The mass, excised in a marginal fashion with contamination of the neurovascular bundle, was solid and proved histologically to be a synovial sarcoma. Amputation was recommended.

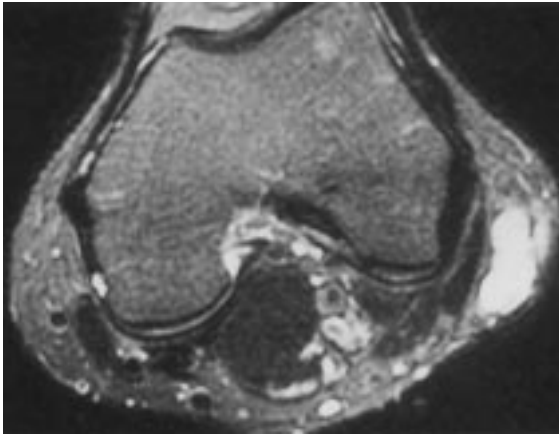


Fig. 7 Subcutaneous slowly enlarging 4-cm mass as shown on coronal T2-weighted spin-echo MR image. Despite the subcutaneous position and small size, histologically this represented a malignant peripheral nerve-sheath tumor (formerly referred to as a neurofibrosarcoma).

frequent concomitant complaints of joint-line pain, swelling, locking, popping, and buckling of the knee.

Plain radiographs of soft-tissue sarcomas are typically unrevealing except for the suggestion of a soft-tissue shadow. However, they may depict mineralization within the tumor or, rarely, secondary bone erosion. Any soft-tissue sarcoma may show mineralization, but synovial sarcomas, mesenchymal chondrosarcomas, and soft-tissue osteosarcomas are particularly likely to do so. Like synovial sarcomas, the rare mesenchymal sarcomas are most likely to occur in persons aged 15 to 40 years.³ Soft-tissue osteosarcoma rarely occurs in persons younger than 40 years old.³ Up to one third of synovial sarcomas classically show one of two patterns of mineralization: either a peripherally pronounced "peppered" calcification or, less frequently, a patchy, cloudlike pattern.³ Mesenchymal chondrosarcomas often show irregular stipplings, arcs, flecks, or streaks (Fig. 4).³ The ossification within a soft-tissue osteosarcoma may be massive and is typically more prominent centrally, in contrast to that seen with myositis ossificans.

Magnetic resonance imaging of soft-tissue sarcomas about the knee is essential for determining the extent of tumor and the relationship to neurovascular structures. A size of 5 cm or larger, lack of encapsulation, poor margination, heterogeneous signal, involvement of neurovascular structures, and soft-tissue edema are all consistent with a diagnosis of sarcoma, but these features are nonspecific.¹² Histologic types of soft-tissue sarcomas are not reliably predictable on the basis of the MR imaging appearance.¹²

The myriad types of soft-tissue sarcomas are distinguishable only on examination of histologic sections, typically supplemented by immunohistochemical staining.³ Synovial sarcomas may be biphasic (composed of epithelial and spindle cells) or monophasic and may be confused with other soft-tissue sarcomas, malignant melanomas, and even metastatic carcinomas. Malignant fibrous histiocytomas classically have a storiform arrangement of plump spindle cells, although myxoid, giant cell, and inflammatory subtypes are also observed. The necessary feature for a diagnosis of liposarcoma is the identification of lipoblasts,

which vary from large "signet ring" cells to cells with small nuclei and only a few tiny lipid droplets. Well-differentiated lipomalike liposarcomas may be extremely difficult to distinguish from simple lipomas because of the paucity of lipoblasts in the former. The most common subtypes of rhabdomyosarcoma observed in the extremity are alveolar and embryonal. Cross-striations within the rhabdomyoblasts of rhabdomyosarcomas signify their skeletal muscle origin.³

A lymphoma may present as a deep soft-tissue mass about the knee, but the primary mass is frequently accompanied by palpable or otherwise demonstrable proximal lymphadenopathy (Fig. 8). Subcutaneous or skeletal muscle nodules metastatic from various sarcomas and carcinomas have been noted, but these typically



Fig. 8 Diffuse large-cell non-Hodgkin's lymphoma presented as an enlarging soft-tissue mass (arrows) adjacent to a total knee replacement, as demonstrated on this anteroposterior plain radiograph. Adenopathy was clinically evident in the groin ipsilateral to the mass.

occur in combination with known metastatic disease to visceral organs.

Treatment

Treatment of synovial processes has increasingly involved the use of arthroscopy. The source of the effusion contributing to Baker's cysts may often be confirmed and treated arthroscopically, especially in the case of intra-articular derangement, without the need for a separate approach to excise the popliteal cyst. Because pediatric Baker's cysts are particularly prone to recurrence, excision should generally be discouraged. Meniscal cysts are also best treated arthroscopically.¹⁷ Appropriate treatment of the associated meniscal tear with decompression of the cyst will lead to resolution in many cases.

Proximal tibiofibular synovial cysts, by virtue of their origin, are not amenable to arthroscopic correction. An open surgical approach to these cysts is appropriate when the diagnosis is in question, when neurologic involvement is evident, or when symptoms can be attributed to the mass.⁸ The possible need for loupe dissection of the cyst from within the epineurium of the peroneal nerve should be anticipated.⁸ Whenever synovial cysts are excised in an open fashion, a marginal excision including only the cyst and its lining tissue is appropriate. Normal structures should not be sacrificed in an attempt to eradicate the cyst. An effort should be made to trace the stalk of the cyst back to its origin in the joint capsule.

The standard treatment of synovial chondromatosis by way of complete loose-body removal and synovectomy⁷ has been called into question by the reported success

of loose-body excision alone.¹⁸ The role of arthroscopy depends on the extent of disease, but many surgeons believe that a complete synovectomy can be accomplished as well, if not better, by arthroscopy than by an open technique.⁷ Removal of loose bodies and the synovial lining from within an associated popliteal cyst is probably best addressed in an open fashion.

Eradication of pigmented villonodular synovitis is generally considered to require complete removal of all involved areas of the joint lining. Complete anterior and posterior synovectomy is recommended for treatment of the diffuse form, although irradiation has been used in some centers for difficult cases. The preferred method of achieving a complete synovectomy is controversial.^{6,19} Clearly, partial arthroscopic synovectomy is inferior to complete arthroscopic synovectomy.¹⁹ However, there have been no controlled trials comparing the results of arthroscopic and open complete synovectomy.

For those benign neoplasms that are specifically identifiable with confidence on physical examination and can be confirmed with MR imaging, observation is frequently the best course of action. Lipomas, hemangiomas, and vascular malformations fall into this category.

For other symptomatic benign active neoplasms confirmed at biopsy, marginal excision as previously described for synovial cysts is the treatment of choice in most cases. A notable exception is the desmoid tumor, which is particularly prone to local recurrence.¹⁶ A wide excision with a cuff of surrounding normal tissue should be the goal, when possible, for desmoid tumors. Achieving such a margin is often difficult due to the nonencapsulated, infiltrative growth pattern of

this tumor. Vital structures should not be sacrificed to achieve this goal. However, radiotherapy should be considered when desmoid tumors occur in the popliteal fossa or when a recurrent tumor is not amenable to wide reexcision.¹⁶ Otherwise, radiotherapy should generally be avoided in the treatment of benign soft-tissue masses.

Both radiotherapy and surgical resection have a role in the treatment of soft-tissue sarcomas (Fig. 9, A).²⁰ Particularly for high-grade or deep-seated soft-tissue sarcomas or those juxtaposed to a neurovascular structure, adjuvant radiotherapy should accompany wide surgical excision. While some researchers suggest an advantage to preoperative radiotherapy compared with postoperative radiotherapy, the risk of wound problems appears to be higher when the surgical incision is made through previously irradiated skin.

Another approach that produces similarly excellent local control is the use of brachytherapy, either alone or in combination with external-beam radiotherapy. Brachytherapy involves the introduction of radioactive beads through intraoperatively placed loading tubes early in the postoperative period (Fig. 9, B).

With the use of these modern combined modalities, local control has been 90% to 95%.²⁰ Unfortunately, the excellent local control achieved in this fashion has not translated into improvements in overall rates of metastatic disease or survival. Multiple chemotherapy trials have failed to document greater benefit than treatment with radiotherapy and surgery alone. Chemotherapy for localized soft-tissue sarcomas deserves continued investigation but should still be considered investigational.

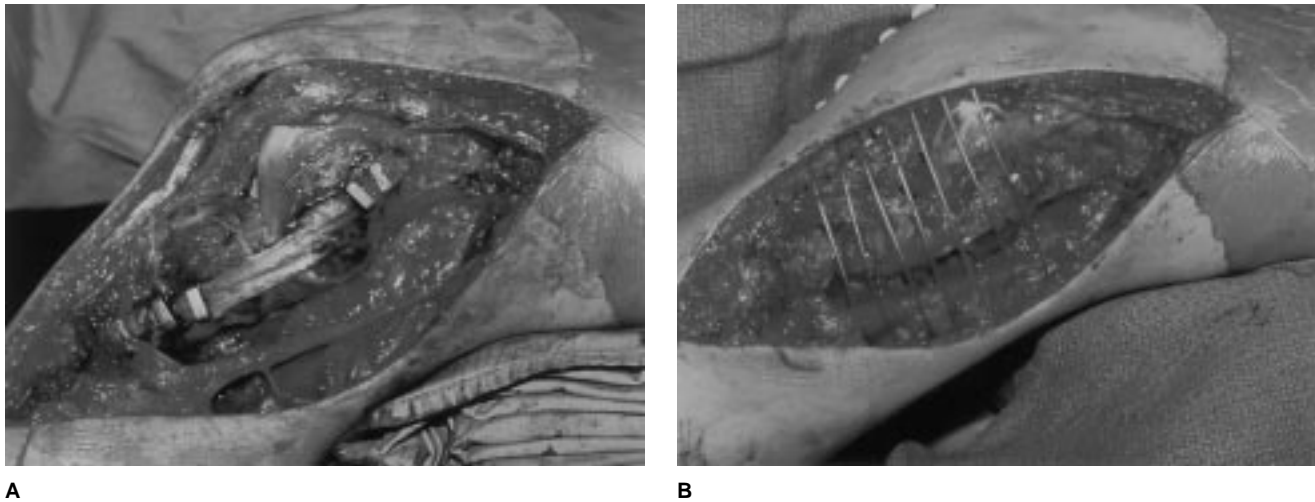


Fig. 9 A small, subcutaneous mass adjacent to the lateral joint line of this patient's left knee was considered clinically to represent a meniscal cyst. At the time of planned excision, however, the presumed cyst was found to be solid. Histologic sections showed a synovial sarcoma. The patient subsequently underwent wide reexcision of the entire prior incision and the lateral aspect of the knee joint, including the lateral distal femur, lateral proximal tibia, proximal fibula, and lateral collateral ligament. The peroneal nerve was preserved. **A**, Reconstruction consisted of a bone-patellar tendon-bone autograft to reconstitute the lateral collateral ligament. **B**, Brachytherapy tubes were placed intraoperatively, and both brachytherapy and external-beam postoperative radiotherapy were administered. Five years postoperatively, the patient was disease-free with a stable knee.

Summary

Many soft-tissue neoplasms that occur about the knee have a peak age incidence identical to that for the much more frequent sports- and occupation-related knee injuries in adolescents and young adults. In addition, the thigh and knee region is a frequent location

for other soft-tissue neoplasms that have a peak age incidence among patients more commonly evaluated for degenerative changes in the knee. Special vigilance is warranted when a soft-tissue mass is not in the typical position or does not have other characteristic features of a meniscal or Baker's cyst, when the size of the mass or the accom-

panying symptoms seem out of proportion to the injury or underlying degenerative process, and when symptoms persist beyond what is expected. When evaluation of a soft-tissue mass about the knee raises suspicion of a neoplastic process, consideration should be given to referral to a tertiary care center before biopsy.

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