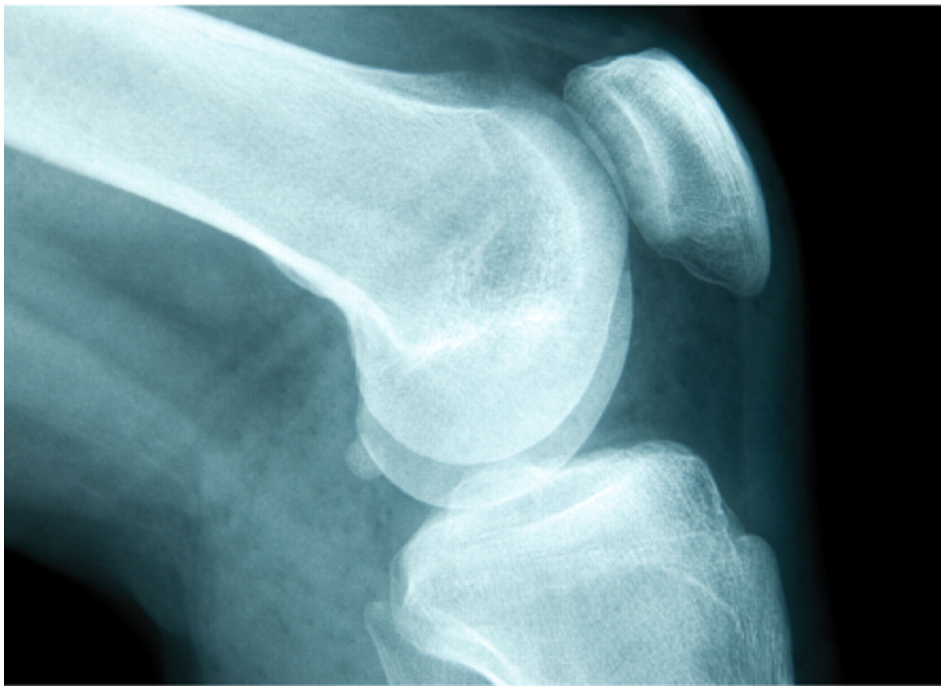
Case 8 Soft tissue swelling

26-year-old female presented with a swelling on the back of the left knee joint since the last 6 months and chronic pain in the calf and foot since the last 2 months. Pain in the calf started as dull aching pain appearing initially on walking which later on progressed and became continuous.

On careful examination the lower half of the leg and the foot showed changes of chronic limb ischemia like decreased local temperature, dryness of skin, hair loss, transverse ridges over the nails along with absent dorsalis pedis and posterior tibial artery pulse on palpation.

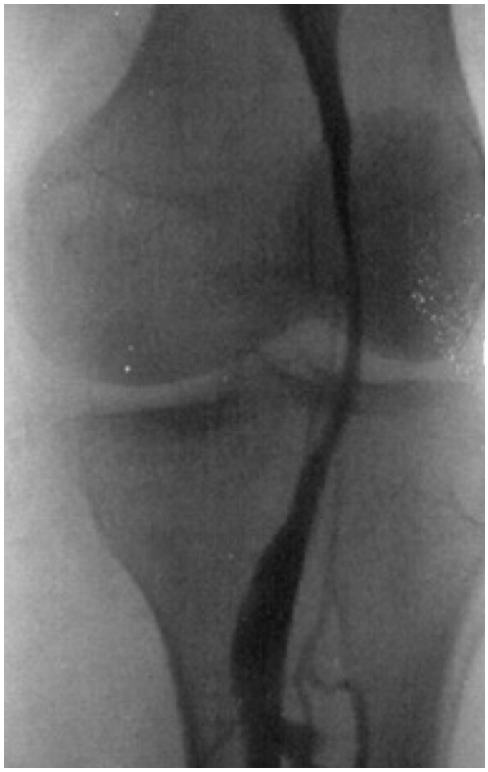
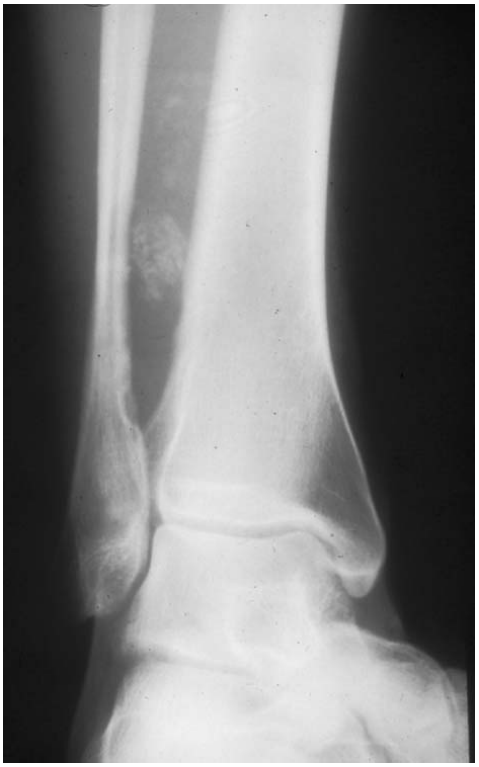
Swelling in the popliteal fossa was tender on palpation and irreducible on flexion of the knee joint. It was fixed to the underlying structures, firm in consistency and the skin over the swelling was normal. X-ray of the joint produced a soft tissue shadow on the posterior aspect of the knee joint.



Your Diagnosis

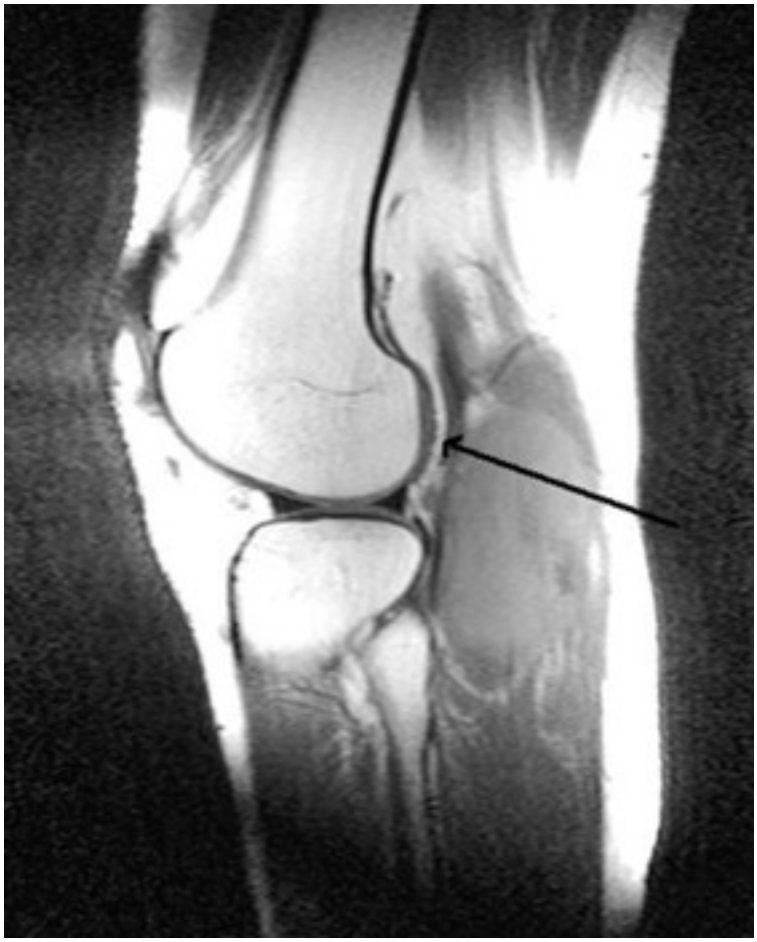
Diagnosis Synovial Sarcoma

Arteriogram: Arteriography for evaluation of peripheral vascular disease showed extraluminal obstruction of the popliteal artery in the popliteal fossa resulting in slow distal flow into the anterior and posterior tibial arteries. Pull back venogram showed extraluminal popliteal vein obstruction

Sometimes radiographs may show a cluster of small, irregular calcifications .

Coronal T1 and T2-weighted MRI confirms a well-defined mass with predominately intermediate T1- and high T2- signal characteristics.



Following gadolinium administration, axial T1-weighted MRI with fat saturation shows avid enhancement of the soft tissue components of the mass that abuts but does not invade the subjacent femur.

INCIDENCE

Soft tissue sarcomas are an infrequent group of mesenchymal tumors, with 5000 cases annually.

10% of these are synovial sarcoma, the fourth most common soft tissue sarcoma.

The majority of soft tissue sarcomas present in older adults,

usually in the sixth decade of life.
Synovial sarcomas, however, are most common in adolescents and young adults aged between 15 and 40 years, with a median age of 35 years.
Synovial sarcomas account for 1% of all childhood malignancy but 30% of soft tissue sarcomas in childhood.
Sex predominance is unclear.

CLINICAL PRESENTATION

Synovial sarcoma typically presents as a slow growing palpable soft tissue mass. Pain and tenderness at the site of the mass are frequent, but pain without a corresponding palpable lesion has been reported. Due to the frequently slow growth of the mass, the average duration of symptoms prior to diagnosis is 2 to 4 years.
Some lesions, however, present as a painless, stable mass that may be dormant for long periods before acting aggressively. This initial slow growth and long duration of symptoms simulate a benign process, which commonly leads to delay in diagnosis. Therefore, imaging studies can be a key to earlier diagnosis.

DISTRIBUTION

No evidence indicates that synovial sarcoma arises from synovial cells or within the synovial lining of a joint. In fact,

synovial sarcoma is infrequently an intra-articular lesion. Only 5% to 10% originate within a joint. These intra-articular lesions are often more aggressive. The more characteristic location is para- or juxta-articular, with the majority of tumors (40%-50%) adjacent to a joint.

Proximity to the joint has been variously reported as within 5 cm of a joint (60%-75%). 90% arise in an extremity.

More commonly seen in a lower extremity, with predominance in the popliteal fossa.

Synovial sarcoma is the most common foot and ankle soft tissue malignancy in patients aged 6 to 45 years, and is the most common lower extremity malignancy of any type in patients aged 6 to 35 years.

Of the nonextremity cases, 3% to 10% are found in the head and neck, pelvis, thorax/chest wall, and retroperitoneum, with reported incidences of 8%, 7%, and 0.3%.

HISTOPATHOLOGY

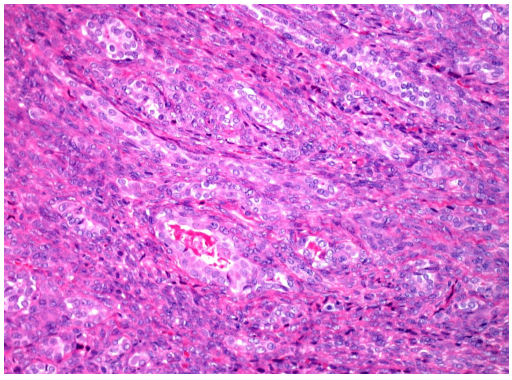
Currently the WHO puts synovial sarcoma in the category "Tumor of Uncertain Differentiation."

The histogenesis of synovial sarcoma does not indicate origin

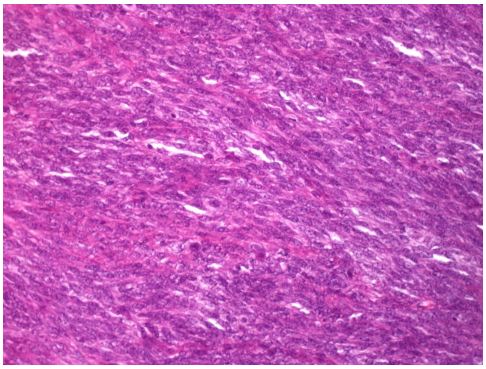
from synovial lining or cells. Although these sarcomas arise from mesenchymal tissue, the cells differentiate sufficiently to have a histological appearance resembling synovioblastic cells, giving rise to the name based on histologic appearance.

The tumors are composed of 2 morphologically distinct cell types: spindle cells and epithelioid cells. Based on the ratio of these cellular constituents, a continuum of 3 histologic tumoral subtypes has been described, including biphasic, monophasic fibrous, or monophasic epithelial.

Biphasic



Monophasic



Approximately 70% are the monophasic fibrous subtype and 30% are biphasic.

It is noteworthy that, since monophasic subtypes have entirely ovoid-spindle cell morphology, this subtype can be difficult to differentiate from fibrosarcoma.

In contrast, the biphasic form consists of 2 distinct cell types (spindle cell and epithelial) in varying proportions.

Less than 5% of all lesions are a poorly differentiated form that represents a form of tumor progression that can arise from

either monophasic or biphasic synovial sarcoma.

Most synovial cell sarcomas demonstrate immunoreactivity to cytokeratin or epithelial membrane antigen. Immunohistochemical analysis for cytokeratins, vimentin, Bcl-2, S100 protein, CD34, smooth muscle actin, and desmin can help narrow the differential diagnosis, but definitive identification may not be possible by histochemistry alone. Molecular genetics and fluorescent in-situ hybridization have proved to be valuable when histologic and immunohisto-chemical tumor analysis is inconclusive.

More than 90% of synovial sarcomas contain a unique genetic translocation,

t(X;18) (p11.2;q11.2),23 that involves the SYT gene on chromosome 18 and 1 of several homologous genes on the X chromosome (SSX1, SSX2, and SSX4).

IMAGING

Radiographs

Synovial sarcomas typically manifest as a round to ovoid juxta-articular soft tissue mass. Extremity lesions are most often periarticular or in close proximity to a bursa or tendon sheath. The most common location is within 10 cm of the knee. Up to 50% of lesions have visible dystrophic calcifications. Calcifications are more frequent than in other soft tissue sarcomas but are nonspecific and can be central or peripheral.

Calcific matrix is variable.
Some calcifications resemble the dense, poorly defined osteoid matrix of a primary bone tumor such as osteosarcoma or early myositis ossificans.
Periosteal new bone formation or bony erosion have been reported in 11% to 20% of cases.

Computed Tomography

With the availability of ultrasound and MRI, CT is seldom used as a primary diagnostic imaging test.
Areas of internal low attenuation necrosis or high attenuation hemorrhage may be seen.
Effect on adjacent bone ranges from indolent pressure erosion to, less commonly, aggressive bone destruction.

Ultrasound

Ultrasound is especially useful for interrogation of superficial lesions.
Most lesions were round and well defined with homogeneous low internal echo patterns, evoking little suspicion of their aggressive nature. More complex lesions with fluid components, hemorrhage, or necrosis in addition to abnormal vascular flow.

Magnetic Resonance Imaging

MRI is the choice of investigation.

Lesions 5 cm in size are often homogenous on all MRI sequences and simulate indolent processes. Larger lesions are usually lobular, well defined masses with dominant soft tissue components of T1 signal intensity slightly lower than muscle. In addition, classic lesions may also have components with intermediate or high T1 signal.

On T2-weighted imaging, most lesions have heterogeneous, predominantly high T2-signal intensity. Portions with high signal on both T1- and T2-weighted images due to hemorrhage .

The combination of a septated, multilobulated appearance and hemorrhage with fluid levels has been termed a bowl of grapes appearance.

DIFFERENTIAL DIAGNOSIS

1.calcifications:

chondrosarcoma, osteosarcoma,

leiomyosarcoma, rhabdomyosarcoma,

liposarcoma

2.When calcifi cation is absent,

Fibrosarcoma and PVNS

TREATMENT AND PROGNOSIS

Surgical Management

Limb-sparing surgery is always preferred when feasible, with no difference in survival compared to amputation. Wide surgical margins are essential for optimal outcomes.

Radiotherapy

If margins are negative, adjuvant radiation is recommended based on the presence of risk factors. In patients with no risk factors, the 5-year risk of local relapse is 9%, but with 2 risk factors this increases to 31%. Radiotherapy has improved local recurrence free survival without change in overall survival.

A retrospective review of 150 patients at 10 years from adjuvant radiation showed local control in 82% of cases, but 44% had distant metastases and 1% had nodal metastases at this time.

Radiation can be external beam or brachytherapy, depending on the individual case. With brachytherapy, the 4-year disease-free survival is 57%,42 and 5-year overall survival is 70%.

Chemotherapy

A large body of literature concerns neoadjuvant, adjuvant, and metastatic disease chemotherapy for soft tissue sarcoma and a smaller group of studies for synovial sarcoma. In general,

the chemotherapy for soft tissue sarcoma is based in doxorubicin and ifosfamide.

Overall survival was equivalent between the 2 groups. For tumors 5 cm, in patients 16 years and younger, 5-year overall survival with chemotherapy was 66%, and 50% without.
For patients 17 years and older, survival with chemotherapy was 58%, and 56% without.41

Novel Therapies

Trabectedin is a novel chemotherapeutic drug .

CONCLUSION

Synovial sarcoma is a rare soft tissue neoplasm of mesenchymal origin, most often juxta peri-articular in young adults, with a predilection for the extremities, particularly the knee. These tumors can mimic benign entities because of their long periods of dormancy preceding a period of rapid growth. Cytogenetics have proven pivotal for correct classifi cation of synovial sarcoma, with a t(X;18) (p11.2;q11.2) translocation that produces a SYT-SSX.

The strongest determinant of synovial sarcoma prognosis is size 5 cm, but other factors also affect prognosis. Treatment is often multimodal.