Synovial sarcoma of the knee joint compressing the popliteal artery: A case report

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Citation

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Abstract

Synovial sarcoma is a malignant mesenchymal neoplasm commonly found in middle age patients, usually in the extremities. We are reporting a case of a 26-year-old female with synovial sarcoma in the left knee joint compressing popliteal vessels with signs of chronic limb ischemia in the distal extremity. Total excision of the tumor was followed by radiotherapy. Histopathologic examination of the excised tumor revealed a monophasic spindle cell synovial sarcoma. Prompt improvement in the chronic limb ischemia in the distal limb was evident with no clinical and radiological signs of local recurrence for 12 months.

KEY MESSAGE

Synovial sarcoma of the knee involving popliteal vessels is a rare condition. The treatment is wide resection with negative margins. Involvement of the vessels needs excision of the involved part with reconstruction by reversed graft interposition. Resection should be followed by localized irradiation.

INTRODUCTION

Synovial sarcoma of the knee involving popliteal vessels is a rare condition. This case is extensively studied with arteriography, pull back venography as well as MRI. Pathological diagnosis is supported by immunohistochemistry.

CASE HISTORY

A 26-year-old female came to our office 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, even on rest, localized more in the forefoot. 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.

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 (Figure 1). Pull back venogram showed extraluminal popliteal vein obstruction (Figure 2).

Figure 1

Figure 1: Arteriography showing extraluminal obstruction

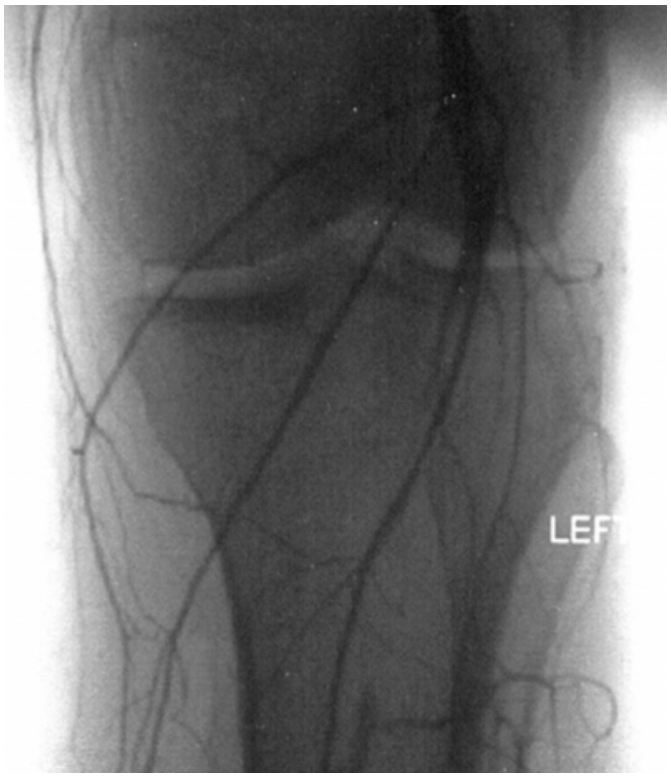
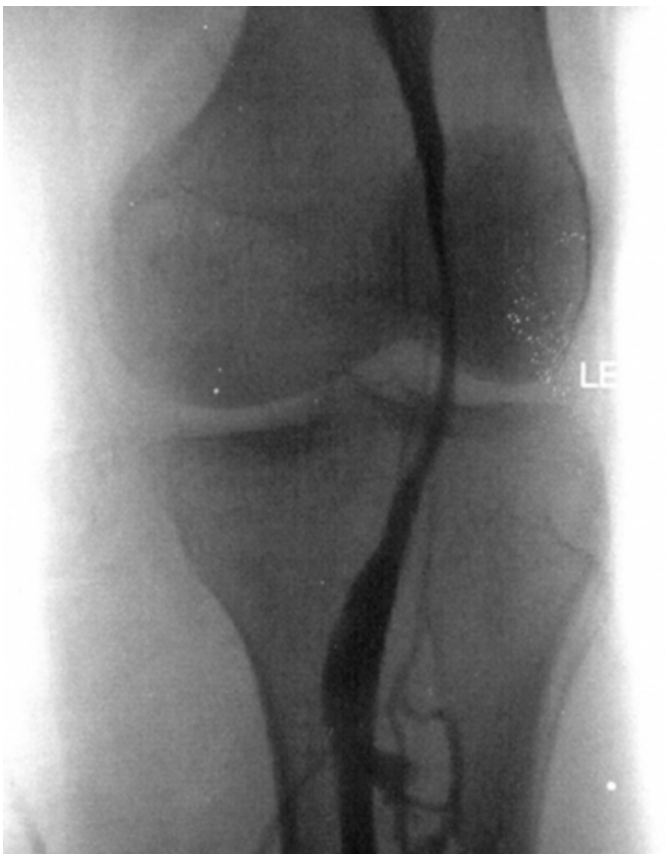


Figure 2

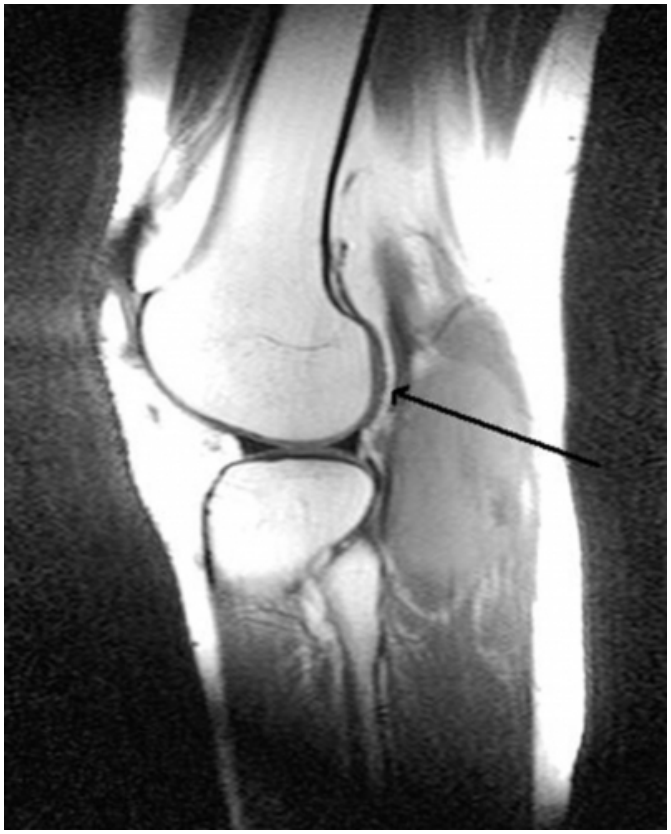
Figure 2: Pull back venogram showing extraluminal obstruction



Ultrasonography of the joint showed a lesion with solid cystic areas and punctate calcification in the popliteal fossa compressing the popliteal vessels and no communication with the joint cavity. MRI of the knee joint revealed a lesion of similar dimensions in the popliteal fossa, compressing and adherent to the popliteal vessels at some places (Figure 3). Fine needle aspiration cytology of the lesion showed spindle shaped mesenchymal cells.

Figure 3

Figure 3: MRI with arrow indicating compressed popliteal artery



Under spinal anesthesia vertical incision was done on the most prominent part of the tumor in the popliteal fossa. The tumor was reached by fine dissection (Figure 4), mobilized from the popliteal vessels and excised with intact capsule (Figure 5).

Figure 4

Figure 4: Tumor arising from the back of the knee joint

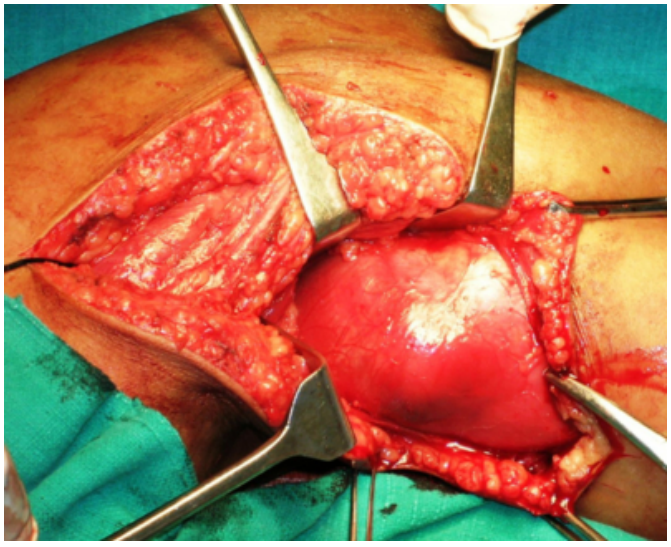
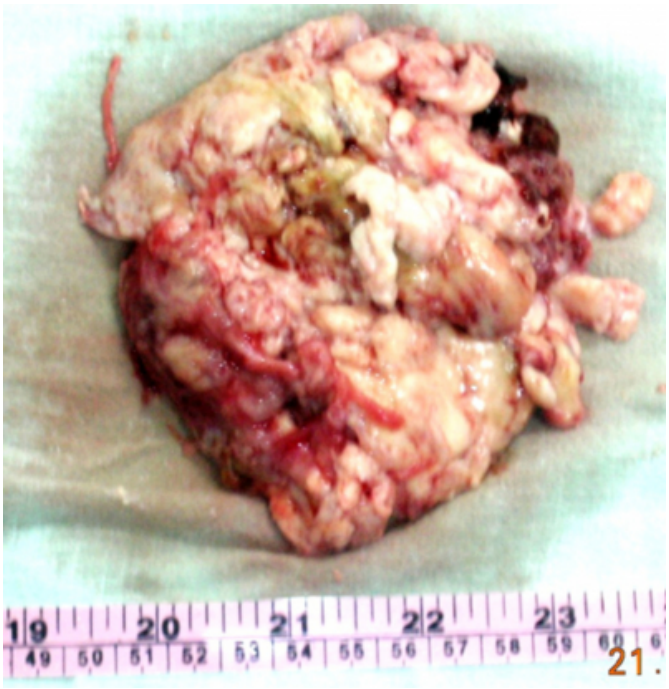


Figure 5

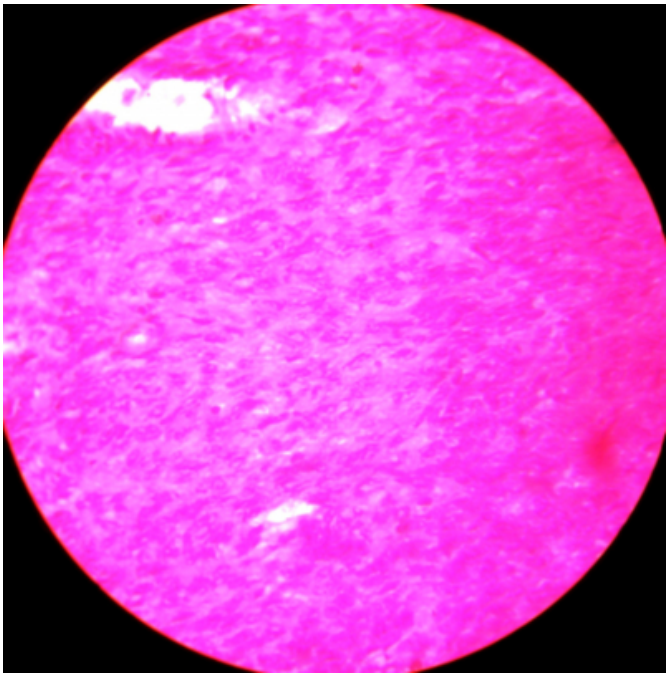
Figure 5: Excised tumor, variegated appearance



The patient was postoperatively given 50 Gray of local irradiation. Histopathology of the specimen revealed predominance of crowded spindle cells in a wavy or short fascicular pattern with few scattered epitheloid cells (Figure 6). Immunohistochemisrty of the tumour cells showed the spindle cells positive for vimentin (Dako, Kyoto, Japan).

Figure 6

Figure 6: Microscopic picture showing crowded spindle cells



There was dramatic improvement in the chronic limb ischemia of left leg and foot with immediate appearance of pulsations of the dorsalis pedis and posterior tibial arteries.

DISCUSSION

Synovial sarcoma occurs mostly in young adults, with a median age of 26.5 and with a slight male predominance [1]. About 50 percent of synovial sarcomas develop in the legs, especially the knees. The second most common location is the arms [2]. Less frequently, the disease develops in the trunk, head and neck region, or the abdomen [1, 2]. It is common for synovial cancer to recur within the first two years after treatment. Half of the cases of synovial sarcoma are notorious to spread to the lungs, lymph nodes or bone marrow [1]. The incidence of synovial sarcoma is ranging from 5% to 10% of all sarcomas. It is, however, the most common soft-tissue sarcoma in the age from 20 to 40 years. A specific cytogenetic abnormality identified is the translocation between X-chromosome and the 11th chromosome. The SYT gene at 18q11 is fused with the distal portion of one of the two duplicated genes SSX1 or SSX2 at Xp11. This SYT-SSX fusion is present in >95% of cases of synovial sarcoma [3]. The tumor frequently presents as a slowly growing mass. Pain and tenderness frequently accompany the mass and are usually responsible for bringing the patient to clinical attention. Tumors near joint spaces may cause limitation of movements if they grow sufficiently large [4].

Most synovial sarcomas are found within 5 cm of a joint but only 10% of cases are intra-articular. Tumors are usually well circumscribed, but in unusual cases, they may interdigitate between muscles and tendons or encase neurovascular structures. Invasion of adjacent bone is seen in 11-20% of patients. MRI is the imaging procedure of choice because MRI is useful for evaluating the extent of the tumor and its involvement of adjacent soft-tissue structures. For instance, MRI is helpful in the differentiation of tumor from muscle tissue and in depicting the involvement of neurovascular structures, tendons, fascia, fat and bone marrow. MRI is also helpful for the differentiation of recurrent soft-tissue tumors from postsurgical or postirradiation changes. Most tumors display heterogeneous intermediate signal-intensity on T1-weighted images and high signal intensity on T2-weighted images [5]. CT is especially useful in depicting calcifications, bone invasion, or periosteal reaction. Angiography allows the gross evaluation of tumor size, vascularity and vessel compression if present. Fine-needle aspiration is less accurate than other techniques for the diagnosis of soft-tissue tumors. Open biopsy is often difficult to perform, and the procedure is associated with an increased prevalence of complications such as poor wound healing and breakdown, bleeding, and infection. Image-guided biopsy is now commonly performed for the initial histological diagnosis of soft-tissue tumors. Synovial sarcomas cannot be distinguished from other soft-tissue tumors by percutaneous biopsy techniques. CT-guidance is optimal for the biopsy of small lesions, or lesions near neurovascular bundles or other critical structures. Fluoroscopy is a real-time imaging modality that can be used for the biopsy of larger lesions that are easily seen on 2-dimensional radiographs. Grossly, the tumor shows the cystic spaces with adjacent areas of necrosis and hemorrhage and characteristic tan-colored, fleshy areas. Histologically synovial sarcoma can be classified into 4 subtypes: classic biphasic, monophasic spindle cell, monophasic epithelioid cell, and poorly differentiated round cell. The classic biphasic type is characterized by admixtures of rather large epithelioid cells and spindle cells. Epitheloid cells have oval, vesicular nuclei, small nucleoli and abundant amounts of eosinophilic cytoplasm and are arranged in small nests. Spindle cells are characterized by hyperchromatic spindle-shaped nuclei and small amounts of cytoplasm. Monophasic spindle cell synovial sarcoma is composed entirely of the spindle cell component. The spindle cells are arranged in alternating fascicles, imparting the so-called herringbone pattern [6].

Immunohistochemistry has become a vital component in the diagnosis of synovial sarcoma. Its hallmark is the expression of both epithelial markers either cytokeratin or epithelial membrane antigen [EMA] in epitheloid cells and vimentin in spindle cells. Cytokeratin 7 and 19 seem to be specific for this tumor, and Bcl-2, CD-99, Leu-7 and S-100 can also be found [7]. Cytogenetic analysis is an important diagnostic tool. Approximately 90% of all synovial sarcomas have a characteristic X:18 translocation. This RNA fusion product can be detected by either reverse-transcriptase polymerase chain reaction or fluorescent in situ hybridization probes in formalin-fixed, paraffin-embedded tissues [8].

For monophasic synovial sarcoma, the differential diagnosis would include fibrosarcoma (an exact morphologic mimic), solitary fibrous tumor, hemangiopericytoma, malignant peripheral nerve sheath tumor (MPNST), leiomyosarcoma, and spindle cell carcinoma. The key determinant that distinguishes synovial sarcoma from all of the above (except spindle cell carcinoma) is the presence of epithelial

differentiation within the spindle cells.

Five- and 10-year survival rates average 76% and 63%, respectively. The tumor has a propensity for late recurrence and metastasis. Favorable prognostic indicators include patient age below 20 years, tumor diameter of less than 3.0cm, heavy calcification, and lack of poorly differentiated areas, a distal limb location, negative resection margins and adjuvant radiation therapy.

CONCLUSION

Compression of the popliteal vessels by synovial sarcoma of knee is an uncommon incidence. The recommended treatment is wide resection with negative margins, which often include surrounding muscle groups or total amputation. Involvement of the vessels needs excision of the involved part with reconstruction by reversed graft interposition. Resection is commonly followed by localized irradiation. Unlike in most other sarcomas, chemotherapy does play a role in patient management, as tumor cells appear to be sensitive to a rather wide range of therapeutic agents, ifosfamide in particular.

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