Leiomyosarcoma of the popliteal artery: Case report and review of the literature

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Vascular leiomyosarcomas are rare tumors, and only 21 have been described as developing in the systemic arteries. We present a case, the sixth in the literature, of a leiomyosarcoma originating in the popliteal artery of a 67-year-old woman treated with en bloc excision of the neoplasia and the artery followed by a popliteal tibioperoneal trunk bypass. The patient then was underwent adjuvant radiotherapy and chemotherapy. After 31 months of follow-up, the patient has mild claudication but does not have either recurrence or metastasis. (J Vasc Surg 2003;37:206-9.)

Vascular leiomyosarcomas are unusual tumors[1,2] that attack arteries less often than veins. The arterial ones are usually seen in the peripheral arteries of the lower legs. Only five reports of popliteal artery leiomyosarcomas have been published in the literature. The rarity of the tumor, the aggressive histologic behavior, the high rate of mortality, and the fact that it may be an unusual cause of early claudication have made diagnosis and early treatment important.

CASE REPORT

A 67-year-old woman with no history of trauma had pain in the left popliteal fossa. No fever or weight loss was associated. The physical examination showed a hard, firm, immobile, and not pulsatile mass occupying the popliteal fossa. No palpable lymphadenopathy or distal edema was found. Scars from a great saphena stripping operation were evident on the skin of the limb. Color Doppler ultrasound scan was performed and showed a solid mass bounding the popliteal artery. An angio–computed tomographic (CT) scan confirmed the report (Fig 1). Angiography showed irregular aspects of the artery's wall with small pathologic collateral vessels supplied by infragenicular and supragenicular branches. The angio–magnetic resonance (MR) scan appearance was consistent with a mass surrounding the popliteal vessels, hypervascularized with a fibrotic component originating in the vessels (Fig 2). 99m diphosphonate technetium scanning and abdominal and chest CT scans were negative for bone, abdominal, or chest metastasis. The patient underwent en bloc excision of the mass together with the popliteal artery and the first tract of the anterior tibial artery. The popliteal vein was displaced but not infiltrated by the neoplasia. The reconstruction was accomplished with a popliteal-tibioperoneal trunk bypass with contralateral inverted saphena. Revascularization of the anterior tibial artery was not deemed necessary because of the good distal outflow. This was confirmed with the postoperative Doppler scan, which showed the presence of good flow of the posterior tibial and dorsalis pedis arteries. The surgical specimen consisted of a solid mass measuring 5 × 3.5 cm that surrounded the popliteal artery and the first tract of the anterior tibial artery. An intraoperative biopsy showed mesenchimal malignant neoplasia, and the definitive histology report showed undifferentiated leiomyosarcoma with involvement of the margins. The neoplastic tissue was formed of eosinophilic spindle cells with vesicular nuclei from ovoid to cigar-shaped with many mitoses (Fig 3). The immunohistochemistry results showed neoplastic cells strongly positive for smooth muscle actin and myosin.

At discharge, Doppler continuous wave scanning revealed an ankle-brachial index of 1, and 2 weeks after surgery, the patient had good motility of the knee and a normal march tolerance. The patient then underwent chemotherapy with epirubicin (30 mg/m^2/d ×3) and ifosfamide (3 g/m^2/d), administered contemporaneously for six 3-day courses with a 6-week rest between them. The patient then underwent radiotherapy with 200 cGY, each administration repeated 30 times. After the radiotherapy, 6 months after surgery, the patient had claudication with a march tolerance of 350 to 400 m. The ankle-brachial index, which was 1 at the time of discharge, now showed a value of 0.65 on both the anterior and posterior tibial arteries. The angio-CT scan ruled out the presence of recurrences, and the angio-MR scan revealed the patency of the bypass but showed obstruction of the tibioperoneal trunk. Vascular outflow appeared to be ensured by several collateral vessels supplying the proximal tracts of the tibial arteries. By the 31st month after surgery, the patient feels good and has no sign of either local recurrence or metastatic disease. The claudication is not disabling, so the patient has refused a new operation. Another angio-MR scan showed the presence of good collateral vessels supplying the tibial arteries.

DISCUSSION

Leiomyosarcomas represent about 5% to 7% of all soft tissue sarcomas.[3-5] They originate in the smooth muscle cells, so most often they affect the gastroenteric system and the uterus, less frequently the respiratory apparatus, and rarely the limbs. The adult population is the most susceptible, particularly women.[3-5] Leiomyosarcomas can be distinguished as intraperitoneal and retroperitoneal, cutaneous and subcutaneous, and vascular neoplasia.[3,5-7]

Vascular leiomyosarcomas represent only 2% of all these neoplasia,[8] and less then 200 cases are reported in the literature.[1,2] Veins are involved five times more often than

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Competition of interest: nil.

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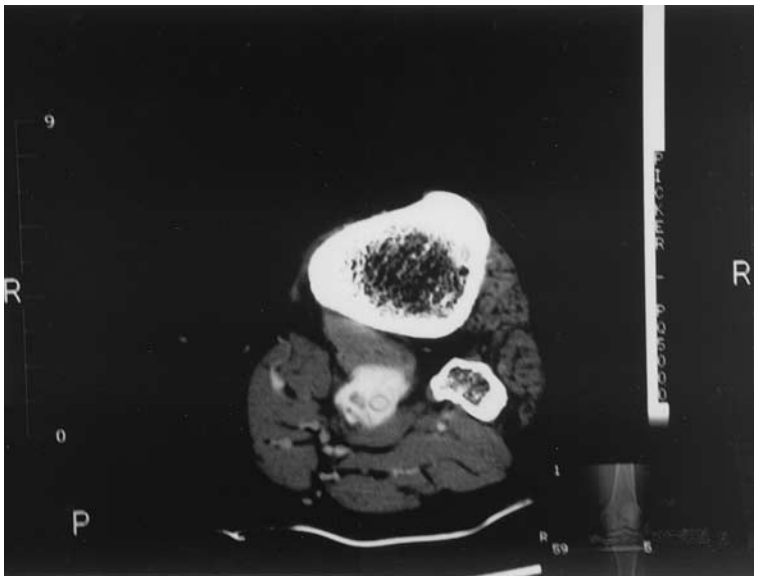

Fig 1. Angio-CT scan: solid mass bounding popliteal artery.

arteries, particularly the vena cava, which alone represents 50% of all localizations, followed by the big systemic veins and the great saphenous.[9] Arterial leiomyosarcomas are equally distributed between the pulmonary artery (19 cases, the one most often involved) and the other systemic arteries (21 published cases).[6]

The first review of the literature on leiomyosarcomas involving arteries and veins was by Kevorkian and Cento,[10] who in 1973, identified eight tumors, of a total of 86 cases, originating in the systemic arteries, three of them considered doubtful. The second review of the literature was made by Briggs and coworkers,[11] who in 1990, while observing a popliteal leiomyosarcoma, added eight more observations to the five cases described by Kevorkian and Cento,[10] for an overall total of 13: four of the femoral artery, three of the common iliac artery,[10,12,13] two of the popliteal artery,[9,11] one of the aorta,[14] one of the axillary artery,[15] one of the internal mammary artery,[9] and one of the inferior mesenteric artery.[9] The most recent review was by Giangola and coworkers[8] (1995), who described one case of subclavian leiomyosarcoma and added two other findings of popliteal leiomyosarcomas[1] and one case of superficial femoral leiomyosarcoma,[16] bringing the overall number of described cases to 17. Further descriptions of cases (not reported by Giangola et al[8]) are one by Basu and coworkers[17] (1988) with popliteal localizations, one by Malone and coworkers[18] (1996) with an aortic localization, one by Gill and workers[19] (2000) with a renal localization, one by Rhode and coworkers[2] (2001) with a splenic localization, plus the case we describe, so that now the overall number is 22 published cases of leiomyosarcomas of the systemic arteries.

Arterial leiomyosarcomas seem to have a predilection for the lower limb arteries (14 localizations on 22 cases).

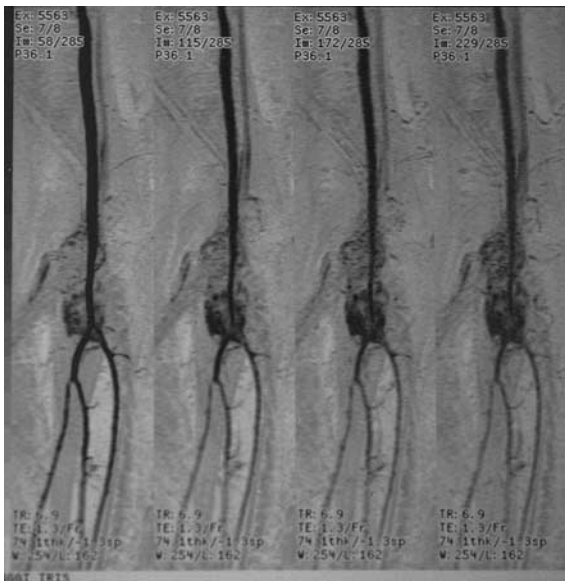

Fig 2. Angio-MR: mass surrounding popliteal vessels, hypervascularized with fibrotic component.

They show high histologic grading (II to III) and correlation between histologic and biologic behavior.[8,10,15] The most reliable prognostic indices are the mitotic index and, above all, histologic grading. Advanced age (>60 years), vascular invasion, and aneuploid DNA are related to more aggressive behavior, so that vascular leiomyosarcomas have a worse prognosis than other soft-tissue leiomyosarcomas.[8,20]

The best treatment of these neoplasia seems to be complete surgical excision or, when this is not possible, at

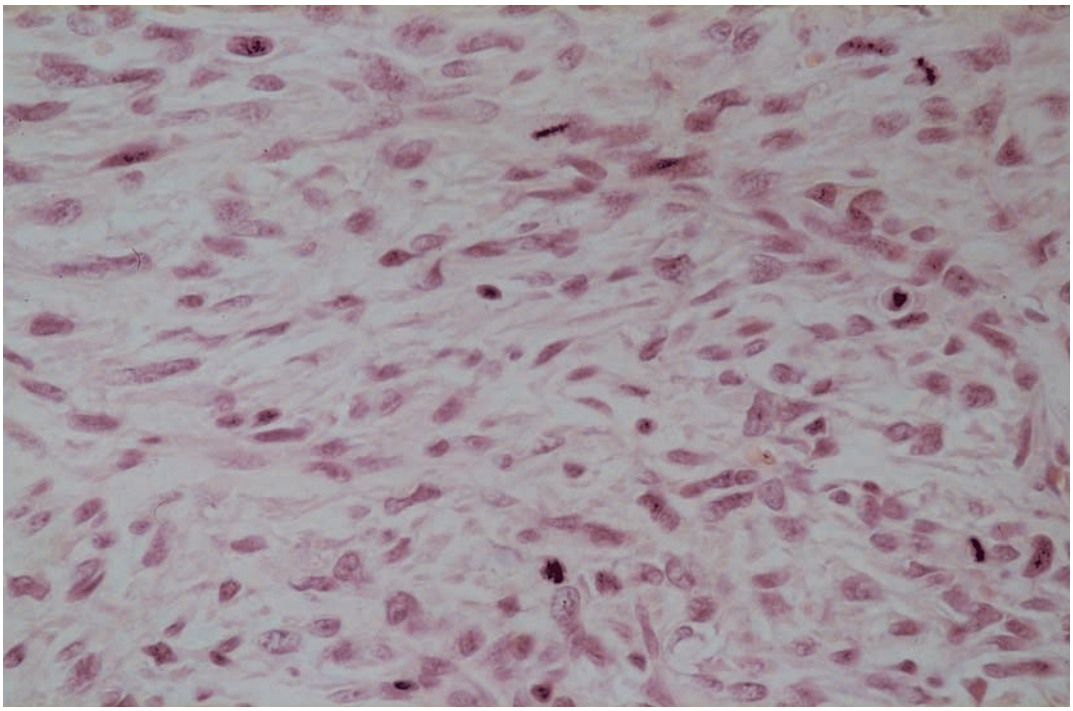

Fig 3. Histology report: neoplastic tissue formed of eosinophilic spindle cells with vesicular nuclei from ovoid to cigar-shaped with many mitoses.

Published cases of leiomyosarcoma of popliteal artery

Author	*Age (y)/ gender*	*Symptoms*	*History*	*Spread*	*Treatment*	*Results*
RL Treiman et al, 1977	77/male	Claudication	Several months	Local: mass recurrence	Excision en bloc with popliteal artery, radiotherapy	Dead after 7 months
Basu et al,[17] 1988	65/male	Claudication, swelling	8 months	Lung	Thigh amputation	Alive with metastasis after 12 months
Leeson, Malaei, and Makley,[1] 1990	65/female	Pain, swelling			Excision and radiotherapy	Alive after 5 years
Leeson, Malaei, and Makley,[1] 1990	51/male	Pain, swelling		Lung	Thigh amputation	Dead after 4 years
Briggs et al,[11] 1990	70/male	Claudication	18 months	Lung	Excision and radiotherapy; thigh amputation	Dead after 10 months
Our case, 2001	67/female	Pain, swelling	12 months		Excision en bloc with the popliteal artery, chemotherapy and radiotherapy	Alive after 31 months

least partial surgical excision accompanied by aggressive radiotherapy. Nevertheless, the 5-year survival rate is poor and is comparable with the survival rate for other soft tissue leiomyosarcomas with vascular invasion (36%).[20]

An interesting role is played by the leiomyosarcomas of the popliteal artery because, although rare, they are the most frequent among those of the peripheral arteries. They may appear with pain in the popliteal fossa and popliteal swelling but also with claudication only[11] (Table).

The diagnosis is usually late, and our patient, too, was seen 1 year after the onset of pain. Although in the past diagnosis was made with radiograph, angiography, and biopsy, ultrasound scan, color Doppler scan, and especially CT and MR scanning can now be used. In our case, angio-CT and angio-MR scans gave us important data in the preoperative studies on the nature of the lesion and the vascular supply of the lower limb, and in the postoperative follow-up, they enabled us to rule out recurrences and to

evidence the situation of the arterial tree after vascular reconstruction.

The management of the published popliteal leiomyosarcomas consisted of amputation in three cases (thigh) and conservative surgery in two cases followed by radiotherapy (Table). Popliteal leiomyosarcomas show a severe prognosis and a high mortality rate. Indeed, three of the five patients reported in the literature died of the disease, one displayed pulmonary metastasis 1 year after surgery as of the date of the report (1988), and only one patient was alive 5 years after the diagnosis (1990). Survival of the deceased patients was between 7 months and 4 years. Tumor spread was observed to be local in one case and to the lungs in the other three (Table).

In our case, after the patient refused amputation, we performed a neoplasia excision with vascular reconstruction followed by chemotherapy and aggressive radiotherapy because of the presence of infiltrating margins, which is now the recommended management comprising a conservative surgical treatment together with adjuvant therapies. Indeed, in the others leg sarcomas, aggressive radiotherapy had the same results in patients with infiltrating and free margins (overall survival rate of 60% to 70% after 5 years).[1]

In conclusion, leiomyosarcomas of the popliteal artery are rare neoplasias belonging to the group of soft tissue sarcomas of the limbs of vascular origin, which should be considered by vascular surgeons both for their gravity and because they can be unusual causes of early claudication. The management of these tumors is today oriented towards preoperative staging and conservative surgical treatment with adjuvant therapy rather than limb amputation. Indeed, although it reduces local recurrences, the demolitive procedure has not been shown to provide a lower rate of metastasis and higher overall survival.

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