Limb Saving Procedure in Metastatic Leiomyosarcoma of the Femoral Vein: Case Report and Literature Review

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Abstract Primary venous leiomyosarcoma is very rare and first description was made by Perl in 1871. Most cases are from venous system and half of them are reported to originate in inferior vena cava. We report the case of a 77-year-old Caucasian man with a leiomyosarcoma of the femoral vein. A leg preservation surgery was decided. Eight months later, the patient died of tumor progression. A PubMed search using the terms “leiomyosarcoma femoral vein” and “leiomyosarcoma vein” was performed. All cases of composite iliac-femoral leiomyosarcoma were excluded, and as far as possible, only well-documented cases were included. Median age was 55 years, seven men and six women, tumor resection was performed by six authors, six others performed a vascular resection, and one preferred for a thigh excision. Prognosis remains poor if metastasis is present, and in these cases, a conservative surgery is recommended to preserve patients’ quality of life.

Keywords Leiomyosarcoma · Femoral vein · Femoral leiomyosarcoma · Surgery

Introduction

Leiomyosarcoma accounts for 5 to 10 % of all soft tissue sarcomas occurring mostly in adults, although it may develop in young adults even in children [1]. Primary venous leiomyosarcoma is very rare and first description was made by Perl in 1871 [2]. To date, about 400 cases have been reported in literature. Most cases are from venous system and half of them are reported to origin in inferior vena cava. Sex prevalence depends on the tumour location with a female predominance for retroperitoneal and vena cava leiomyosarcoma [3] but no difference if tumors localisation is in the lower limb [4].

Case Report

We report the case of a 77-year-old Caucasian man admitted 1 year ago in our department for phlebitis. Past medical history included only hypothyroidism and hypertension controlled with medical therapy. At physical examination, a painful mass in right thigh was discovered. Doppler ultrasound showed a thrombus in the femoral vein and a 7×13 cm mass in the adductor canal (Fig. 1a). Femoral artery appeared dislocated and lumen was reduced at the computed tomography scan. A leg perfusion with tumor necrosis factor was not feasible due to the vein occlusion and an amputation was excluded due to the presence of pulmonary micro-metastasis. A leg preservation surgery was decided according with the patient. First of all, an artery covered stent was placed into the femoral artery by-passing the tumor compression. Surgery started with a safety arterial dissection and the

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Fig. 1 **a** Mass presence in right leg. **b** H-caldesmon immunostain. In the center of the photo an atypical mitotic figure. **c–d** Hematoxylin and eosin stain magnification $\times 10$. The leiomyosarcoma arising from the vascular wall

isolation and mobilization of the femoral nerve for his complete preservation. Ligation of the femoral vein behind saphenous vein origin and thrombectomy was then performed. The last step was en bloc removal of the sartorius muscle and piece of the vastus medialis and adductor longus. The tumor was composed of spindle-shape neoplastic cells intensely positive for smooth muscle actin, for desmin and h-caldesmon and negative for cytokeratin and epithelial membrane antigen. The mitotic count was ten mitotic figures for ten high power field. Foci of coagulative necrosis were found (Fig. 1b, c, d).

After surgery, no pain, no claudication, and no motor deficit was observed. Patient began chemotherapy with epirubicin (80 mg/m^2 every 21 days) 1 month after surgery. Eight months later, the patient died for tumor progression.

Discussion

Although leiomyosarcoma of soft tissue share common histological characteristics, it is useful to divide them in four main

Table 1 Review of femoral vein leiomyosarcoma cases from 1993 to 2013

Case number	Author	Publication year	Patient age	Gender	Origin of tumor	Surgery	Follow up months	References number
1	Roy	1993	39	Male	Femoral vein	Vascular resection	6	18
2	Porcellini	2003	31	Female	Artery femoral	Vascular resection	30	19
3	Gow	2005	46	Male	Superficial FV	Thigh excision	5	20
4	Bashir	2005	50	Female	Femoral vein	Vascular resection	24	21
5	Condilis	2005	75	Male	Femoral vein	Vascular resection	12	22
6	Subramaniam	2007	29	Female	Femoral vein	Vascular resection	Unknown	23
7	Shindo	2009	77	Male	Femoral vein	Vascular resection	2 died	24
8	Paraskovas	2009	55	Female	Superficial FV	Vascular resection	12	25
9	Propper	2009	84	Female	Bifurcation FV	Mass resection	6	26
10	Yfadopoulos	2011	69	Male	Superficial FV	Mass resection	60	27
11	Gil-Sales	2012	69	Male	Femoral vein	Mass resection	12	28
12	Perisano	2013	47	Male	Common FV	Mass resection		29
13	Perisano	2013	44	Female	Superficial FV	Mass resection	12	29
14	Present case		77	Male	Femoral vein	Vascular resection	8	

F femoral, V vein

groups. Intraabdominal leiomyosarcomas are the most common group, accounting for 40–45 % of cases, and develop in omentum, mesentery, and retroperitoneum usually in middle age and old women. As they tend to reach a very large size, a complete excision is often impossible and the overall 5-year survival rate is 20–30 % with metastasis mainly to the lungs and liver. Subcutaneous and deep soft tissue leiomyosarcomas of the limbs accounts for 30–35 % of cases affecting the same age group but with a slight male predominance. About a half of them develops metastasis and the overall 5-year survival rate is 60–65 %. Cutaneous leiomyosarcoma account for about 15–20 % of cases affecting mainly young adults and showing a male predominance. They are often painful, interesting the limbs, the lower leg being the site of predilection. The local recurrence is very common and rarely metastasize. Vascular leiomyosarcoma accounts for 5 % of cases arising usually from the inferior vena cava or from large veins of the lower limb. Leiomyosarcoma is preponderant in female if the vena cava is the tumor localisation. The overall 5-year survival is about 20 % with cases arising from smaller and distal veins appearing to have a better prognosis. Review of literature is resumed in Table 1. The histological features of leiomyosarcoma vary according to the degree of differentiation but are usually characterized by intersecting fascicles of brightly eosinophilic spindle cells with vesicular, cigar-shaped nuclei.

The tumor cells usually display strong positivity for smooth muscle actin, desmin, and h-caldesmon but some of them may be, at least focally, positive for cytokeratin and epithelial membrane antigen [5]. Positive surgical margins were predictive of worse survival in the series reported by Hines et al. [6]. Varela-Duran et al. [7] attempted to make a correlation between their microscopic features and biologic behavior. Pathology findings relating to tumor grade, mitotic rate, and tumor seemed to be closely related to the clinical outcome of these tumors. But the value of histologic prognostic indicators remains controversial. Venous leiomyosarcomas are highly aggressive; they mainly metastasize to the lung and liver. Postoperative therapy and the management of recurrence are difficult because of the absence of evidence of their effectiveness in patients. Radiation has been used in both the neoadjuvant and adjuvant settings, and some authors believe

that it may help with the local control of disease. For localized, resectable, soft-tissue sarcoma, adjuvant chemotherapy with doxorubicin or a combination of doxorubicin and ifosfamide has been shown to prolong the time to recurrence and the overall survival [8]. The surgical resection remains the key treatment.

Conclusion

Leiomyosarcoma of the femoral vein is very rare, often misdiagnosed because of symptoms mimicking thrombosis and claudication. Prognosis remains poor if metastasis is present, and in these cases, a conservative surgery is recommended to preserve patients' quality of life. It should be recommended to refer such cases to multidisciplinary group to give the best therapeutic option to the patient.

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