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Leiomyosarcoma of the great saphenous vein: a case report and review of the literature

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Abstract

Introduction: Leiomyosarcoma of the venous system is rare, even more so in the greater saphenous vein. In the 85 years since van Ree described the first case in 1919 only 25 cases have been reported in the world.

Methods: We describe a case of an 85-year-old woman who was successfully treated by excision of the tumour. We also reviewed pertinent literature with regard to age, gender spread, tumour size, survival, occurrence of metastases and therapy.

Results: The median age was 54 years (range 2–85 years), with a 3:2 female to male ratio. The median size of the tumours was 4.1 cm (range 2–12 cm) and metastases occurred in seven of the 25 cases. If any form of adjuvant therapy is used it is usually radiotherapy. Chemotherapy seems to be reserved for cases where metastasis occurs. Average survival was 4 years (range 1 month to 17 years). Currently the best treatment seems to be wide excision of the tumour, with selective vascular reconstruction combined with adjuvant radiotherapy.

Key words: *leiomyosarcoma, great saphenous vein*

Introduction

Primary malignant venous tumours are very rare. Leiomyosarcoma of the saphenous vein even more so, constituting about one in every million malignant tumours. In the 85 years since Van Ree reported the first case in 1919, only 24 cases have been reported in the international literature. We describe an additional one, and pertinent literature is reviewed.

Case

An 85-year-old woman was admitted to our hospital with loss of weight and general discomfort. She also complained of a progressive and painful swelling in the right groin, which she had noticed 3 weeks before. She had no relevant medical history.

On physical examination the woman was seen to be cachectic. A solid swelling with a diameter of 3 cm was observed in the right groin inferior to the inguinal ligament. The swelling was mobile in relation to the skin but fixed to a deeper structure. It was thought to be an enlarged lymph node. Further physical examination showed no other abnormalities.

Additional ultrasound and MRI of the right groin showed a tumour 2.5 cm in diameter in close proximity of the femoral vein (without obstructing it) and the cortex of the pubic bone just below the skin. Ultrasound guided biopsy showed leiomyosarcoma grade II. Preoperative workup showed no evidence of metastases.

During surgery we observed that the tumour originated from the great saphenous vein close to its insertion in the common femoral vein (Fig. 1) The tumour was excised with a wide margin.

Pathological examination of the excised tumour showed a leiomyosarcoma grade II with a diameter of 3.6 cm radically excised and four lympnodes with reactive changes but no signs of metastasis.

Postoperative course was complicated by a wound infection, which was contained with a wound debridement and antibiotics. There were no further complications and the patient made a slow but full recovery. After 27 days she was discharged from the hospital.

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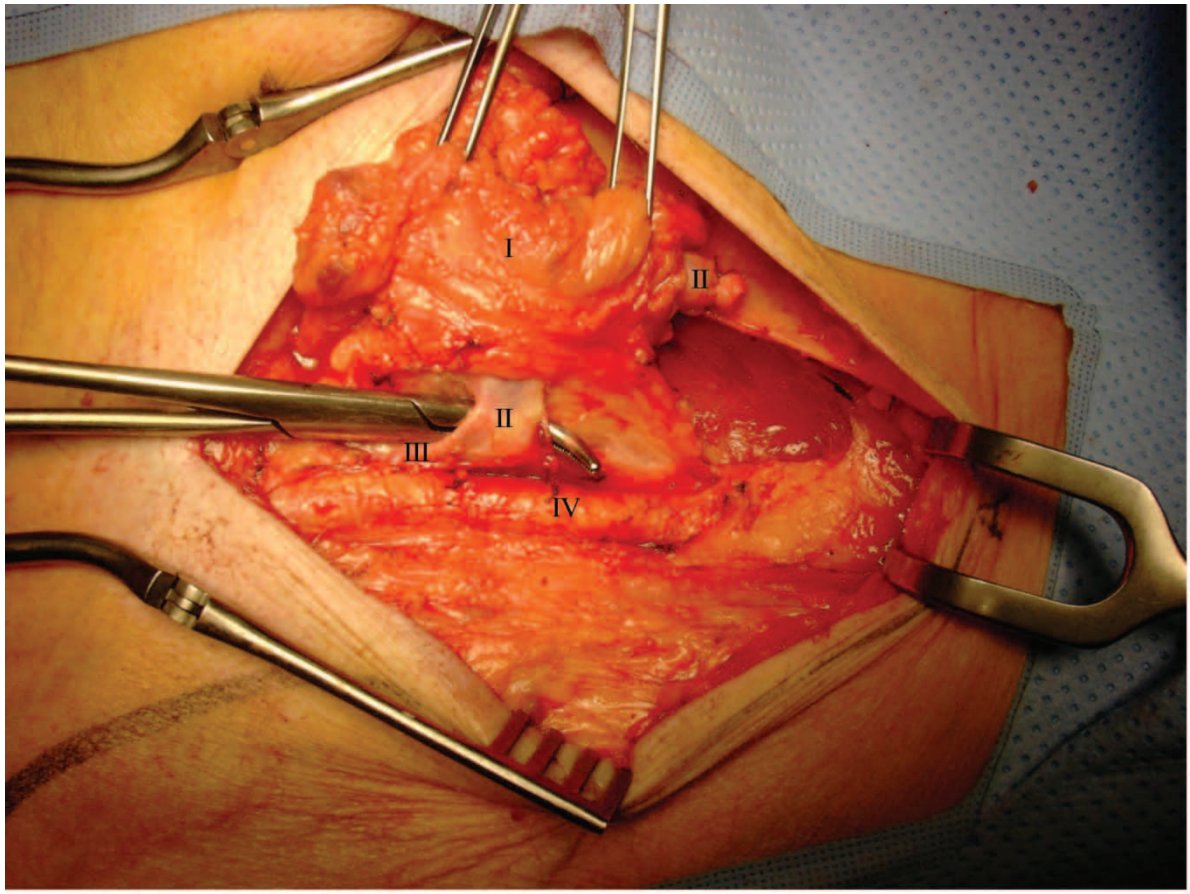

Fig. 1. *Leiomyosarcoma of the greater saphenous vein. (I) Leiomyosarcoma; (II) greater saphenous vein; (III) common femoral vein; (IV) common femoral artery. The head of the patient is on the left hand side.*

She refused adjuvant radiotherapy. At her last outpatient visit, 2 months after the surgery, the wound had fully healed, and she suffered no effects from surgery.

Discussion

Malignant tumours that arise from venous walls are very uncommon, even more so in the lower extremity.[1,3] Primary venous leiomyosarcomas constitute less than one in every 100,000 malignant tumours, and about 10% of these originate from the great saphenous vein, with the inferior caval vein being the predominant location. This case is the second reported in the Dutch literature, and only the 25th in the international literature. The international literature consists mostly of case reports. There are only a few reviews and series.

Reix *et al.*[1] describe a series of seven patients with a venous tumour, six with a leimyosarcoma and one with hemangioendothelioma. Four of them originated from the superficial femoral vein, two from the common femoral vein and only one originated from the great saphenous vein. Median survival in this group was 31 months. Most of them died from metastasis. Local recurrence was never observed. Of the patients with leiomyosarcoma, four received adjuvant radiotherapy (60 Gy in all cases), one refused; and in one case was not indicated because the tumour was small and wide, and wide excision was performed. Chemotherapy was only offered in case of metastasis. Prognosis was poor due to early occurrence of metastasis, occurring in six of the seven cases.

In the same institute, a study was conducted to assess the best imaging technique for diagnosing venous tumours. It was concluded that echo duplex and MRI were best suited for diagnosis of venous tumours.[2] This was confirmed by Dzsinich *et al.*[3]

The latter author described a series of 13 patients with a primary venous leiomyosarcoma.[3] Of these 13, only two had a tumour arising from the greater saphenous vein. Most occurred in the inferior caval vein. At the time of their publication they state that only 197 cases of primary venous leiomyosarcoma had been reported in the period of over 100 years since Perl[4] reported the first in 1871. Of these, only 19 originated from the greater saphenous vein.

Humphrey *et al.*[5] described a case and reviewed the literature. At the time of their publication in 1987, only 15 cases of a leiomyosarcoma originating from the great saphenous vein had been reported,

Table 1. *Reported cases of leiomyosarcoma of the saphenous vein*

Case	Author	Year reported	Age	Sex	Size (cm)	Metastasis	Local recurrence	Adjuvant RT/Chemo	Secondary RT/Chemo	Follow-up	Outcome
1	Van Ree	1919	42	F	*	No	No	No	No	15 months	Alive, no evidence of disease
2	Smout	1960	76	F	4	No	No	Yes, 6000 Rad	No	8 years	Death, no evidence of disease
3	Dorfman	1963	56	M	3	No	No	No	No	1 year	Alive, no evidence of disease
4	Cristiansen	1964	68	F	4.5	No	No	No	No	2 months	Alive, no evidence of disease
5	Allison	1965	3,5	F	3.5	No	No	No	No	6 months	Alive, no evidence of disease
6	Leu	1969	40	M	*	No	No	No	No	2.5 years	Alive, no evidence of disease
7	Szaz	1969	68	M	5.5	Liver, cheek	Yes	No	No	4 years	Death, liver failure
8	Hughes	1973	53	F	2.5	No	No	No	No	6 months	Alive, no evidence of disease
9	Jenstrom	1975	64	M	12	No	No	No	No	14 months	Alive, no evidence of disease
10	Gross	1975	46	M	5	Thyroid, subcutaneous	No	No	Yes, RT 6500 Rad	3 years	Alive, no evidence of disease
11	Stringer	1977	39	M	*	No	No	No	No	8 years	Alive, no evidence of disease
12	Stringer	1977	36	F	*	Lung, scalp, chest, bone, heart	No	No	Yes chemo and RT; 4750 Rad	11 years	Death, pulmonary metastasis
13	Fischer	1982	66	F	2	No	No	No	No	4 years	Alive, no evidence of disease
14	Berlin	1984	60	M	3	Lung, liver	No	No	No	1 month	Death, pulmonary embolism
15	Humphrey	1987	45	M	2.5	No	No	Yes, 6800 Rad	No	3 years	Alive, no evidence of disease
16	Welk	1991	35	F	5	No	No	Yes, 64 Gy	No	11 months	Alive, no evidence of disease
17	Song	1991	54	F		No	No	*	*	*	Alive, no evidence of disease
18	Dzsinich	1992	70	F	*	No	No	*	*	17 years	Alive, no evidence of disease
19	Dzsinich	1992	54	F	*	Lung	No	*	*	9 months	Death, pulmonary metastasis
20	Saglik	1992	61	F	6	Lung	Yes (twice)	No	Yes, chemo	5,5 years	Death, pulmonary metastasis
21	Stellard	1992	64	F	7	No	No	No	No	*	Alive, no evidence of disease
22	Bryard	1993	2	F	2.5	No	Yes (twice)	No	Yes, chemo	12 years	Alive, no evidence of disease
23	Stambuk	1993	48	M	4.1	No	No	Yes, 50 Gy	No	1 year	Alive, no evidence of disease
24	Reix	1998	64	M	5	Skin, lung, brain	No	Refused RT	Yes, chemo	6 years	Death, pulmonary metastasis
25	Van Marle	2004	85	F	3.6	No	No	Refused RT	No	3 months	Alive, no evidence of disease

*No data available.

theirs included. Since then another nine have been published,[1,3,6–11] making this case the 25th case in 85 years since Van Lee[12] described the first case of leiomyosarcoma originating from the greater saphenous vein in 1919. These 25 cases are presented in Table 1. The median age was 54 years (range 2–85 years). Most cases occur in the fourth to sixth decade.

In previous reports describing leiomyosarcomas of the venous system in the lower extremities[1,5] no gender predisposition was observed, which was somewhat surprising taking into account that a 4:1 female to male ratio is observed when the leimyosarcoma is located in the inferior caval vein. With 25 cases reviewed we observe a 3:2 female to male ratio.

The median size of the tumours was 4.1 cm, with a range from 2 to 12 cm. This is much smaller compared with leiomyosarcoma of the inferior caval vein, with a median size greater than 10 cm.[3]

Metastases occurred in seven of the 25 cases, with the lungs being the predominant site, and were the main cause of death. Because leiomyosarcomas are slow-growing tumours they often stay undetected for a long time, which is probably one of the causes of the large proportion of patients with metastases.

If any form of adjuvant therapy is given it is usually radiotherapy. The level of evidence to support this is low because of the rarity of the disease. However Mutter *et al.*[13] state that radiotherapy, 55–70 Gy, should follow excision of primary leiomyosarcoma if it occurs in the femoral vein. We offered it to our patient but she refused to undergo radiotherapy. Chemotherapy is mostly reserved to treat metastasis, with a wide variety of regimes being used, but doxorubicin and methotrexate are most commonly used. Initially the metastases respond well to the therapy, but ultimately the patient dies.[1,9]

When reviewing the literature, survival is found to be limited, with median survival ranging from 2.5 to about 4 years.[1,3,5] With 25 cases reviewed we find an average survival of 4 years, which could be much better if corrected for the short follow-up periods in some reports. The longest disease-free survival reported is 17 years.[5]

In conclusion, it can be stated that leiomyosarcoma of the saphenous vein is a very rare tumour. Because of its location in a vein, and because of its slow-growing tendency, and therefore its high rate of metastasis, survival is limited. If metastases occur, the lungs are the predominant site. In terms of diagnosis, MRI and Doppler echography seem to be the imaging techniques of choice. Because of the rarity of the disease there can be no conclusions drawn with regard to the necessity of adjuvant therapy. Radiotherapy is, however, frequently offered and seems beneficial.

Wide local excision with selective venous reconstruction, and adjuvant local radiotherapy, probably offers the only hope for prolonged survival.

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