CASE REPORT

Leiomyosarcoma of great saphenous vein localised to the calf

Wah Wah Lin,¹ Serkan Erkan,² Richard Carey Smith,³ Kishore Sieunarine⁴

¹Department of Vascular Surgery, Fiona Stanley Hospital, Murdoch, Western Australia, Australia

²Fiona Stanley Hospital, Murdoch, Western Australia, Australia

³Department of Orthopaedic Surgery, Sir Charles Gairdner Hospital, Nedlands, Western Australia, Australia

⁴Royal Perth Hospital, Perth, Western Australia, Australia

Correspondence to

Kishore Sieunarine,
Kishore@westcoastvascular.com.au

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SUMMARY

We present the case of a 76-year-old man with a 2-month history of mildly tender swelling in the right calf for which he had an initial excision and then a wide local excision followed by a split skin graft because the initial histopathology confirmed that there was a Grade I leiomyosarcoma of great saphenous vein. A simple lump in the lower limb could be a malignant vascular tumour and should always be considered in the list of differential diagnosis of a lump in the lower limb.

BACKGROUND

Vascular leiomyosarcoma is a rare tumour, more frequently arising from veins rather than arteries. The commonest site for venous leiomyosarcoma is inferior vena cava followed by renal vein, femoral vein and great saphenous vein. Leiomyosarcoma of great saphenous vein affects men and women equally with the peak incidence found in the 55–61 age range.¹ Early diagnosis is vital for survival, and the exclusion of distant metastasis is crucial for long-term prognosis. A complete surgical excision is the definitive treatment with or without adjuvant radiotherapy or chemotherapy in the case of distant metastatic spread.

CASE PRESENTATION

A man aged 75 years was referred to a tertiary centre vascular clinic with a 2-month history of a slightly painful lump on the medial aspect of right lower leg. He did not have any other associated symptoms such as pressure, pulsation and neurological symptoms. There was no history of previous injury to the right leg. The only medical history was hypertension, gastric reflux, the angiokeratoma of the scrotum, right total knee replacement and laminectomy for lumbar spondylosis. He was on regular medication of felodipine, ramipril, esomeprazole and atorvastatin. He was an ex-smoker with a 20-pack-a-year smoking habit and also a social drinker.

INVESTIGATIONS

A Doppler ultrasound revealed a well-defined hypoechoic, solid appearing lesion surrounding the great saphenous vein just below the knee (figure 1), measuring 20 mm in length and 9 mm in diameter. There was no involvement of the surrounding soft tissues and underlying muscles. The findings suggested a possibility of a mesenchymal tumour from the great saphenous vein or a thrombosed segment of varicose great saphenous vein.

TREATMENT

An initial localised excision of the lump revealed on histopathology confirmed the suspicion of a Grade I leiomyosarcoma. As a result, a wide local excision (WLE) was performed with an split skin graft (SSG) of the underlying defect. The post-operative course was uneventful. The specimen showed positive desmin and actin, negative Melan A, HMB45, AE1/3 and p63. The tumour differentiation score was 1, with the mitoses score and necrosis score standing at 1 and 0, respectively. The final surgical margin was clear, and there was no evidence of distant metastasis on the positron emission tomography (PET) scan.

OUTCOME AND FOLLOW-UP

The patient was fine with no local recurrence or distant metastases at the 6-month follow-up after the surgical resection.

DISCUSSION

Vascular leiomyosarcoma is a rare mesenchymal tumour arising from smooth muscle cells, and it is the most frequent type of primary malignant vascular tumour² making up <2% of all leiomyosarcomas in which veins are involved five times more often than arteries;³ most commonly the inferior vena cava followed by renal vein, femoral vein and great saphenous vein. With reference to the literature, the vascular leiomyosarcoma of extremities has been reported in cephalic vein, femoral vein, superficial femoral vein, popliteal vein, great saphenous vein and dorsal vein of the foot. The great saphenous vein is the most frequent site of origin in the lower extremities.

Its rarity in the lower extremities is shown by the fact that there are only 33 cases reported during the span of nearly 100 years from 1919 to 2015 in the literature on the leiomyosarcoma of great saphenous vein. According to the review by Marle *et al*⁴ in 2004 of 25 cases and the subsequent

Figure 1 Hypoechoic lesion at the right calf.

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8 new cases reported in the literature, the tumour occurs in a wide age range from 2 to 85 years whereas the peak incidence is shown in the 55–61 age range¹ with the median tumour size of 4.1 cm in the 2–12 cm range.⁵

The patients of low-grade saphenous leiomyosarcomas have a 90% chance of surviving for 5 years with a wide resection while those of high-grade saphenous leiomyosarcomas have an 80% chance of surviving for 5 years after a surgical resection.⁶

Histologically, leiomyosarcomas have a typical pattern of interlacing, sweeping bundles of spindle-shaped cells, with elongated occasionally torquated, blunt-ended nuclei.³

It usually presents itself as a lower extremity mass, and it is often mistaken for a deep or superficial vein thrombosis or lymphedema. In our case, it was initially thought to be a thrombosed varicose vein. A high index of suspicion is important for an early diagnosis. A Doppler ultrasound facilitates the diagnosis, and MRI is necessary to discern the extent of local

involvement and differentiation from other soft tissue sarcoma. A PET scan is also necessary to exclude the distant metastasis.

A WLE is the treatment of choice while radiotherapy is an adjuvant therapy depending on the tumour grading. Chemotherapy is mainly for distant metastasis or systemic therapy. Local recurrence is rare, and the lungs are the most common site for a distant metastasis. In the literature, no leiomyosarcoma of great saphenous vein was found to have a distant metastasis at the time of diagnosis.⁷ As a result, a better prognosis should be expected.

Prognosis is generally poor for patients with distant metastasis. An early diagnosis therefore is important for survival while adjuvant radiotherapy is helpful for a high-grade tumour.

Contributors All authors contributed to the case report. KS involved in planning of the case report, initial surgery, follow-up the patient, supervision and revision of the case report. RCS involved in later surgery and followed the patient up for the leiomyosarcoma. SE and WWL involved in literature search and writing the case report.

Competing interests None declared.

Patient consent Obtained.

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Learning points

- It is important to be aware that a lower extremity mass can be a vascular leiomyosarcoma, and therefore, a high level of suspicion is indicated, which is to be confirmed by an early diagnosis.
- In our case, a wide local excision was performed without adjuvant radiotherapy or chemotherapy because there was no distant metastasis.
- MRI and a PET scan are required to exclude the local extension of disease, recurrence and distant metastasis.
- Surgical excision is the mainstay treatment; followed by radiotherapy and/or chemotherapy for distant metastasis.
- Prognosis is generally poor for patients with distant metastasis.

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