CASE REPORT

Leiomyosarcoma presenting as 'idiopathic' unilateral lower limb lymphoedema

Haji Khairul Abd Kadir,¹ Jagjeeth Naik,² Coonoor R Chandrasekar¹

¹Trauma and Orthopaedics Department, Royal Liverpool and Broadgreen University Hospitals NHS Trust, Liverpool, UK

²Vascular Surgery Department, Royal Liverpool and Broadgreen University Hospitals NHS Trust, Liverpool, UK

Correspondence to
Mr Haji Khairul Abd Kadir,
haji.kadir@doctors.org.uk

Accepted 9 October 2017

SUMMARY

A 74-year-old woman presented with an 8-year history of unilateral swelling of her right lower limb, which was thought to be 'idiopathic' lymphoedema until she noticed a painless swelling in her right groin. Physical examination showed a painless, non-pulsatile, deep-seated mass in her right proximal thigh with significant lymphoedema distally. MRI scan showed a large mass at her right inguinal region, involving the common femoral vein. Histological examination on complete excision revealed leiomyosarcoma. It is important to exclude proximal obstructive lesion before making a diagnosis of unilateral idiopathic lower limb lymphoedema.

BACKGROUND

Leiomyosarcoma accounts for 5% to 10% of all soft-tissue sarcomas mainly presenting in adults.¹ This malignant lesion arises from the smooth muscle cells mostly found in the uterus and the gastrointestinal tract.² Primary venous leiomyosarcomas are even rarer, representing less than 2% of all leiomyosarcomas, and majority of them occur in large veins, specifically the inferior vena cava.³ It has been initially reported by Perl in 1871, and to date, about 400 cases of vascular leiomyosarcoma have been recorded in the literature.⁴ We report a case of a primary leiomyosarcoma of the common femoral vein. Due to its rare incidence and lack of familiarity, diagnosis tends to be delayed.

CASE PRESENTATION

A 74-year-old woman was referred to our hospital with an 8-year history of gradual onset of worsening unilateral right lower limb swelling with lymphoedema. She denied any inciting events to this problem. She had been investigated by her general practitioner and treated for a subclinical deep vein thrombosis and cellulitis. She started noticing a painless mass over her right groin in the last 6 months that was gradually increasing in size. She denied any systemic symptoms or weight loss.

On physical examination, there was a painless mass measuring 5×9 cm in her right proximal thigh. It was firm, non-pulsatile with a well-defined margin and a smooth surface. It was arising from deeper structures underneath the fascia and not fixed to the skin. There was a significant lymphoedema involving the whole of the right lower limb and no enlarged lymph nodes were found.

INVESTIGATIONS

Ultrasound and MRI scans demonstrated a large (5.5×5×9.5 cm) mass in the intermuscular compartment of the right inguinal region abutting the common femoral vein and artery (figures 1 and 2). A tumour nodule has breached the common femoral vein wall. She was further investigated with an ultrasound-guided core biopsy, which showed leiomyosarcoma, while CT thorax, abdomen and pelvis did not reveal any convincing evidence of metastasis.

TREATMENT

The patient underwent a planned excision of the leiomyosarcoma under general anaesthesia with a team of sarcoma and vascular surgeons. This was performed via a 15 cm longitudinal incision. Once the common femoral artery was safely dissected and protected (figure 3), the lesion was fully excised along with the common femoral vein and a cuff of normal tissues surrounding the lesion (figure 4). The common femoral vein was not reconstructed during the procedure.

Histopathological analysis showed a thinly encapsulated, malignant, smooth muscle neoplasm comprising intersecting fascicles of myoid fibres with 'cigar-shaped' hyperchromatic nuclei and abundant eosinophilic cytoplasm. There were large muscular vessels at the tumour periphery from which it had arisen. This confirmed grade 2 leiomyosarcoma on the modified Trojani system.

Figure 1 T1W MRI showing the tumour arising from the common femoral vein.

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To cite: Abd Kadir HK, Naik J, Chandrasekar CR. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-219898

Figure 2 T1W fat suppression with contrast MRI showing the tumour arising from the common femoral vein.

OUTCOME AND FOLLOW-UP

Her postoperative period was uneventful. Her lower limb oedema had slightly worsened, but it was well controlled with compression stockings and postural measures. She had postoperative radiotherapy treatment and her lymphoedema had improved after a year following her surgery. A follow-up MRI scan performed at 1 year showed no evidence of local recurrence.

DISCUSSION

Vascular leiomyosarcoma is rare and constitutes 5% of all leiomyosarcoma cases usually originating from the smooth muscle cells of the inferior vena cava or from large veins of the lower limb.⁴ Leiomyosarcoma occurs more in female patients if it originates from the vena cava, but it does not show such preference in the lower limb.⁵ Since 1993, fewer than 20 cases of this condition have been reported that involved the lower limb.^{4 6 7}

Patients may present with significant venous stasis, lymphoedema, deep vein thrombosis, ill-defined pain and palpable mass with or without embolism. It has clinical features that may be confused with deep venous thrombosis or infective process in lower limbs, thereby leading to

Figure 3 Intraoperative photograph showing the tumour arising from the common femoral vein.

Figure 4 Macroscopic appearance of the tumour with the common femoral vein.

a delay in diagnosis and appropriate treatment. However, it is important to investigate and identify proximal obstructive lesions before making a diagnosis of 'idiopathic' lymphoedema.

Complete removal of the tumour should be a priority as positive surgical margins were predictive of inferior survival rate as reported by Hines *et al.*⁸ Radiation, both in neoadjuvant and adjuvant settings, has been used to help with the local control of disease. Surgical resection still continues to be the key management plan for this condition.⁹

Venous leiomyosarcomas are particularly aggressive leading to early metastasis to the lung and liver. Bode *et al* estimated that half of the patients would have already developed metastases at diagnosis, hence, adjuvant or palliative chemotherapy needs to be prudently considered in these cases. However, due to its rarity, its usefulness has not yet been demonstrated.⁶ Some authors suggested the use of chemotherapy in recurrent lesion or in large, high-grade sarcomas.²

Louail *et al* reported on six patients with peripheral venous leiomyosarcomas, four of whom died as a result of diffuse metastases with a median follow-up of 36.4 months. The median survival was 31 months and the local recurrence appeared in 80% of cases in the first 2 years.¹⁰ The average 5-year survival rate is 20%–30% with cases having more desirable prognosis if it arises from smaller and distal veins.⁴

Recent research from a global cohort of 1162 sarcomas has suggested that more than half of sarcomas possess abnormal monogenic and polygenic variation in their genes. Therefore, family members with a significant history of high-risk genetic variants would benefit from regular surveillance and prevention approaches.¹¹

Learning points

- ▶ Vascular leiomyosarcoma is a rare, malignant soft-tissue tumour arising from the smooth muscle cells.
- ▶ In the primary care setting, proximal obstructive lesions should be ruled out before making a diagnosis of an 'idiopathic' unilateral lower limb lymphoedema.
- ▶ Complete surgical excision and postoperative radiotherapy is the standard treatment.

Contributors HKAK has been involved in the care of the patient during the perioperative period, data collection and manuscript write-up. CRC is the orthopaedic consultant who has been involved in the overall care of the patient including surgical intervention and part of the write-up of this manuscript. JN is the vascular consultant who was involved in the surgical intervention.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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