Primary leiomyosarcoma of the great saphenous vein: A case report

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ABSTRACT

Introduction and importance: Vascular smooth muscle sarcomas are rare neoplasms that comprise less than 2% of all leiomyosarcomas. These malignancies usually originate in the inferior vena cava, with a limited number of cases affecting the great saphenous vein. Due to the limited reports on these sarcomas, epidemiologic data remains insufficient.

Case presentation: We report the case of a 67-year-old Hispanic female that presented with an asymptomatic growing mass in her right thigh. She was managed with an En bloc resection under the impression of a smooth muscle vascular sarcoma. The diagnosis was confirmed after histopathologic evaluation.

Clinical discussion: Vascular leiomyosarcomas remain a rare and challenging diagnosis. They usually present as a slowly growing mass that is initially asymptomatic. High clinical suspicion and a comprehensive radiologic evaluation, including magnetic resonance imaging, are crucial. Histopathologic evaluation is essential for diagnostic confirmation. Surgical excision remains the treatment of choice, with radiation therapy mostly considered for local disease control. Postsurgical surveillance is necessary every three months to monitor for signs of recurrence.

Conclusion: Physicians should remain aware of the nonspecific presentation of leiomyosarcomas and the importance of a comprehensive diagnostic approach. Early diagnosis and adequate management are fundamental elements in the treatment of these aggressive tumors.

1. Introduction and importance

Smooth muscle sarcomas (SMS) arising from a vessel wall are extremely rare [1–3]. They represent less than 2% of all leiomyosarcomas and occur five times more frequently in veins than arteries [1,4,5]. These neoplasms are most commonly associated with the inferior vena cava (IVC), followed by a few cases arising from the great saphenous vein (GSV) [4,6]. They present as a slowly growing painless mass [1,4,6]. Due to their insidious and asymptomatic presentation, their diagnosis and treatment are frequently delayed resulting in poor prognosis, with distant metastases found in 10 to 25% of patients at the time of diagnosis [5]. We present our experience with the clinical presentation, diagnostic approach, and surgical management of a patient with a GSV leiomyosarcoma with no signs of recurrence 24 months after treatment.

This case is reported in line with the updated consensus-based surgical case report (SCARE) guidelines [7].

2. Case presentation

2.1. Clinical presentation

A 67-year-old Hispanic female with a past medical history of hypertension, diabetes mellitus type 2, and obstructive sleep apnea presented to our musculoskeletal oncology clinic with a complaint of a right thigh mass. She noticed a small non-tender lump in her inner thigh five months prior to our evaluation. She did not recall any preceding injury or trauma but was concerned that it had grown steadily. She denied

Abbreviations: CD-US, Color Doppler Ultrasound; CT, Computed Tomography; GSV, Great Saphenous Vein; IVC, Inferior Vena Cava; MRI, Magnetic Resonance Imaging; SMA, Smooth Muscle Actin; SMS, Smooth Muscle Sarcoma; VL, Vascular Leiomyosarcoma.

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She also denied allergies, previous surgeries, or malignancy. Still, her family history was pertinent for ovarian cancer in her paternal grandmother and lung cancer in her maternal grandfather. Social history was negative for smoking, alcohol abuse, or drug use. Physical examination was remarkable for a non-tender mobile mass on the medial aspect of the right thigh. The lesion was superficially mobile but appeared to be fixed to the deeper structures. Her neurovascular evaluation showed preserved sensation and palpable arterial pulses within the affected extremity.

2.2. Diagnostic assessment

A right thigh color doppler ultrasound (CD-US) was performed, which showed an endoluminal lesion within the GSV (Fig. 1). The contrast-enhanced magnetic resonance imaging (MRI) confirmed an enhancing solid soft tissue mass arising from the GSV (Fig. 2). Based on the patient’s history of a growing mass, and after careful evaluation of the radiologic studies, the patient was scheduled for surgery under the diagnostic impression of a vascular SMS. Additional imaging studies for tumor staging were ordered, and these were negative for distant lesions.

2.3. Surgery and histopathologic evaluation

A wide excision was performed by the senior author at a major oncologic hospital. At surgery, a 4 cm soft tissue mass was encountered in association with the GSV. For vascular delimitation, the GSV was identified and ligated at the proximal and distal aspects. An En bloc excision was performed. On gross examination, a well-delimited roundish mass was identified, measuring 4.0 × 2.5 cm. A sample was sent to a major national referral center for diagnostic confirmation. Their report described a circumscribed spindle cell proliferation with variable cellularity with frequent hypercellular areas. High mitotic activity (> 20/10HPF) and focal tumor necrosis were also noted (Fig. 3). Immunohistochemical studies were positive for desmin and smooth muscle actin (SMA). The tissue analysis was consistent with a high-grade leiomyosarcoma involving the GSV. Moreover, the report suggested that the sarcoma occupied the resection margins. Consequently, a tumor bed excision was scheduled one month after the first intervention. During surgery, a large seroma was encountered, but no signs of recurrence were noted. Margins’ specimens were sent to pathology, with no residual sarcoma identified. The patient was discharged home without further complications on her post-operative day #2.

2.4. Adjuvant therapy and outcome

She received 27 sessions of radiotherapy starting two months after the second intervention. At 24 months follow up, the patient remained free of disease and independent in her activities of daily living.

3. Clinical discussion

Leiomyosarcomas are malignant neoplasms that represent around 6% of all soft tissue sarcomas [6]. Those SMS involving major vessels comprise less than 2% of all leiomyosarcomas [1,4,5]. The vascular structure most commonly affected is the IVC (35–60% of the cases). Nonetheless, some cases have been reported involving the GSV in the lower extremities [1,2]. A mean age of 55 to 61 years had been described for those patients with a GSV leiomyosarcoma [1,6,8]. However, due to the few reported cases, most of the demographic data retrieved remain without statistical significance [6]. Hence, our intention to present the case of a 67-year-old female with an SMS in the GSV in a moment that literature remains limited since the first case reported in 1868 by E. Aufrecht [9]. Up to this date, only 44 cases have been reported in the literature (Table 1).

Vascular leiomyosarcomas (VL) usually progress in an intraluminal to extraluminal fashion [6]. Their relatively slow development can be divided into three stages: the nonocclusive stage, occlusive stage, and terminal stage [6,10]. The nonocclusive stage is primarily asymptomatic, and the diagnosis is commonly made during a routine examination, usually as an incidental finding. The second stage is often found without symptoms because of sufficient collateralization, although phlebitis and unilateral edema have been reported. Unfortunately, these tumors, especially in the inguinal region, are not diagnosed until the terminal stage when distance metastases have already occurred [6].

Their histopathologic features consist of malignant spindle cells and cigar-shaped nuclei arranged in interweaving fascicles. They also contain bundles of filaments with dense bodies of pinocytic vesicles [6,11]. Moderate amounts of eosinophilic cytoplasm may also be observed [6]. Immunohistochemistry shows antibodies to vimentin, SMA, and desmin proving the muscular origin of these tumors [6,11]. Both SMA and desmin were reported positive in our patient.

Physical examination usually shows a painless enlarging mass [1,5,6]. However, these neoplasms could present as hard, lobulated, bruit-less, or even as a soft and subtle mass that could mislead to other etiologies such as a ganglion cyst or a fibrous reaction. They could be superficially mobile but still be adhered to deeper structures [5,6]. The presentation of edema on physical examination may suggest an intraluminal or extramural extension of the tumor [5]. Nonetheless, a painless lump remains the principal symptom reported by patients with lower extremity sarcoma [2].

The nonspecific findings typically observed with subcutaneous masses must direct the physician towards a comprehensive diagnostic approach. MRI should be the modality of choice, especially if there are concerns for malignancy [5,12]. However, other functional imaging modalities, such as magnetic resonance angiography, CD-US, or computed tomography (CT), could be considered. Particularly, CD-US is an accessible, non-invasive, and less expensive alternative that could be considered initially to characterize a mass before proceeding with an advanced imaging study such as an MRI. In addition, CT staging should be obtained to rule out metastases if prior studies suggested malignancy [5,12]. Our patient’s history in association with the combined radiologic approach (CD-US + MRI) provided enough information to our musculoskeletal radiologist to present VL as the most likely diagnosis based solely on imaging.

For diagnostic confirmation, direct tissue evaluation should be performed. Biopsy techniques should follow the standard sarcoma guidelines indicating excisional biopsy for tumors with less than 3 cm or an incisional biopsy for larger tumors [6]. Moreover, tumor resection with wide margins is mandatory for leiomyosarcoma management [4,13].
Even when GSV involvement is observed, establishing vascular continuity during the procedure is not mandatory. Although excision followed by autologous vein patch or polytetrafluoroethylene prosthesis has been attempted for small pedunculated endoluminal tumors [2,5,10]. Ultimately, complete tumor resection is crucial to decrease the risk of local recurrence [4,10]. For this reason, post-procedural specimens should receive a histopathological evaluation for diagnostic confirmation and margins assessment.

Currently, there is no substantial evidence to establish the superiority of adjuvant therapies such as radiation or chemotherapy in the management of VL [4,11]. However, current data lend support to radiation therapy for the local control of the disease [4]. Nonetheless, the role of radiation depends on the tumor grade, postsurgical margins, and institutional preferences [5,12]. The role of preoperative radiation
therapy of SMS remains unclear. Up to this date, chemotherapy has not shown a clear survival benefit [4,6,8,14]. Even so, in patients with metastatic disease evident at diagnosis, the recommended treatment is local excision, regional radiation, and adjuvant chemotherapy [3].

Post-surgical surveillance consists of close follow-up every three months to evaluate for signs of recurrence [4]. Routine chest radiograph and site-specific MRI should be ordered every six months for a two-year period [11,12]. Unfortunately, up to 60% of patients with VL have metastasis at the time of resection, with the lung representing the organ most commonly affected [1,11,12].

4. Conclusion

VL remains a rare neoplasm with a limited literature, clinical experience, and research regarding its treatment [11]. Our experience should remind physicians about the nonspecific presentation of these sarcomas and the importance of a comprehensive diagnostic approach, including a thorough radiologic assessment. Early diagnosis and adequate management are fundamental against these aggressive tumors due to their high incidence of metastasis when the diagnosis is delayed.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

This study is exempt from ethical approval in our institution.

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CRediT authorship contribution statement

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Norman Ramirez, MD: Supervision, reviewing, and editing.

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Declaration of competing interest

The authors have no conflict to disclose.

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