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Case Report

Great saphenous vein leiomyosarcoma: A case report

Awfa Abdulaziz Alkhaled *, Sumaiah Saleh Alkazmi, Haya Ibrahim Altheyab, Eman Abdulwares Cheikhzen, Rafat Ismaeel Abu Shakra

Department of Radiology, International Medical Center, Jeddah, Saudi Arabia

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ABSTRACT

Background: Leiomyosarcoma is a type of soft tissue sarcoma and one of the most common subtypes among all sarcomas. It is also the most common malignancy affecting the vascular system with an overall poor prognosis with 10-years survival rate of <60%. Leiomyosarcoma can occur with identifiable risk factors such as radiotherapy exposure or due to unidentifiable factors and genetic factors.

Case: We present a 49-year-old female patient with painful left inner thigh lump for six months which has been increasing in size. The swelling location was in the medial side of the left thigh, non-pulsatile and tender with no signs of hotness, discoloration or discharge. Ultrasound was performed and reported a subcutaneous hypoechoic well-defined lesion that is in close proximity to Great Saphenous Vein. The diagnosis was confirmed after surgical excision and was followed up by computed tomography for metastatic lesions.

Discussion: Cases of leiomyosarcoma is rare and usually present with painful swelling. However, non-painful and non-tender lesions can present, and clinicians should be wary of such swelling, especially when they are in close proximity to vascular area. Great saphenous vein leiomyosarcoma is difficult to diagnose solely by radiology, but ultrasound can help guide the course. Other associated factors such as deep venous thrombosis may arise.

Conclusion: In conclusion, a thorough radiological assessment is necessary in lesions close to vascular lesions to accurately diagnose it.

1. Introduction

Leiomyosarcoma (LMS) is a vascular soft tissue sarcoma (STS) and is one of the most common subtypes of all sarcomas contributing to 25% of all STS [1–3]. LMS is the most common malignancy affecting the vascular system with more tendency to arise from the venous system [4]. The malignancy have a poor prognosis with reported 10-year survival rate of 31–66% with some studies reporting it as low as 22% [4]. LMS have a predilection to arise from abdominopelvic and soft tissues while it is rare for it to appear in the extremities [3].

LMS is a disease of the elderly peaking after the seventh decade of life but can occur in the third decade in cases of uterine LMS in women [3]. . Previous exposure to radiotherapy has been hypothesized as a risk factor predisposing to the development of STS which progresses to LMS [3]. In addition, patients with genetic syndromes such as Li-Fraumeni syndrome and hereditary retinoblastoma can develop LMS among other types of sarcomas [5].

Clinically, LMS presentation is non-specific as they often present with a mass causing compression or displacement of adjacent organs [5]. Meanwhile, patients with uterine LMS can present with abnormal uterine growth, uterine bleeding or dysmenorrhea [3]. There are no specific laboratory or radiological investigations that help in diagnosing LMS. and the only way to confirm the diagnosis is by performing a biopsy with a histopathological examination of the sample. This case report has been written in accordance with the SCARE criteria from the SCARE 2020 guideline [6].

2. Case presentation

A 49-years-old female patient, known case of hypothyroidism, presented to the family medicine clinic complaining of painful left inner thigh lump for six months and has been increasing in size. Her past medical history is significant for impaired glucose tolerance, vitamin D deficiency, anemia and knee bursitis. Afterwards, the patient was transferred to the surgical unit for assessment of the lump. The patient was vitally stable and on local examination of the lump. The swelling location was in the medial side of the left thigh, non-pulsatile and tender with no signs of warmth, discoloration or discharge. Laboratory

* Corresponding author.
E-mail address: Awfaaa90@gmail.com (A.A. Alkhaled).

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investigations included complete blood count, electrolyte profile, kidney function test, liver function test, serology and bleeding profile were all within the normal limits. Ultrasound was performed and reported a subcutaneous hypoechoic well-defined lesion that is in close proximity to Great Saphenous Vein (GSV) measuring 1.2 × 0.8 × 0.8 cm. The patient was then prescribed local Lidocaine 5% Patch for local pain management and was scheduled for biopsy. Three months later, the patient did not undergo the biopsy and presented with the same complaint. The second ultrasound showed the same finding, but the lesion increased in size measuring 1.6 × 1.6 × 1.3 cm. The patient also reported increase in pain and that it limited her daily activities.

The patient was then scheduled for surgical removal of the lesion due to the progressive increase in size and pain. The surgery was performed by (Operating Surgeon) at (Name of Hospital). During surgery a xxxxxx excision, was given it was confirmed that the tumor originated from xxxxx and was invading xxxxxxx. Surgically, the lesion was excised with safe margins and was sent for histopathology. Histopathological examination in ×4 magnification showed the tumor was adherent to a large blood vessel (Fig. 1) while in ×20 magnification it showed marked nuclear atypia, intersecting fascicles of spindle cells with mitotic figures, with a mitotic index of (X/X HPF). I Immunohistochemical studies were tumor positive for desmin and smooth muscle actin (SMA) (Fig. 2). The surgery was uneventful, and patient was discharged on the X post-operative day without any complications.

Regarding immunohistopathology, the slides were stained with Ki67 and it showed a Ki67 index of 15–20% (Fig. 3). The diagnosis was then confirmed to be LMS, and the patient was scheduled for chest, abdomen and pelvic computed tomography (CT) scan to exclude any metastasis. Her CT scan reported tiny non-specific left lower lobe pulmonary nodule, tiny splenic lesions, intramural fundal fibroid with signs of intrabdominal metastasis. The patient was then scheduled for follow up every 6 months for monitoring to detect evidence of metastasis There were no reports of recurrence.

3. Discussion

Vascular LMS originates from the smooth muscle of the vessel walls [7]. The first case of venous LMS was reported by Perl in 1871. Ever since, 50% of the reported vascular LMS arise from the inferior vena cava while 15% arise from the iliac veins [8]. LMS occurring in major vessels is very rare with 2% and is likely to happen in veins five more times than in arteries [9]. In the lower extremity, the most common vessel affected is the GSV accounting for almost 30% of all venous LMS [9]. Moreover, females are more likely to be found with LMS of the GSV

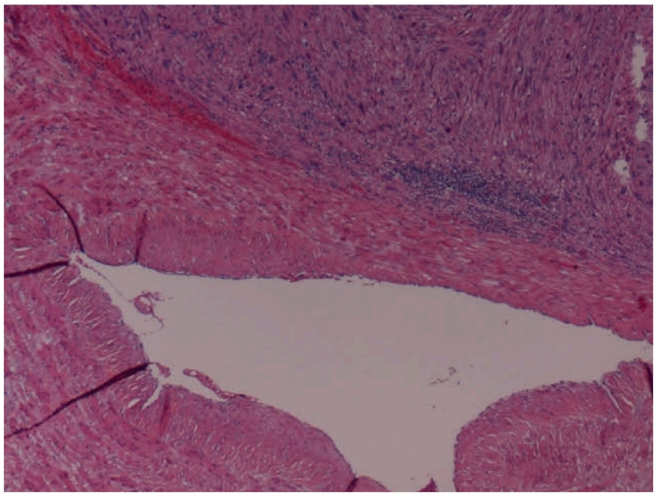

Fig. 1. The tumor (upper left aspect) is adherent to a large blood vessel (lower right aspect) ×4 magnification.

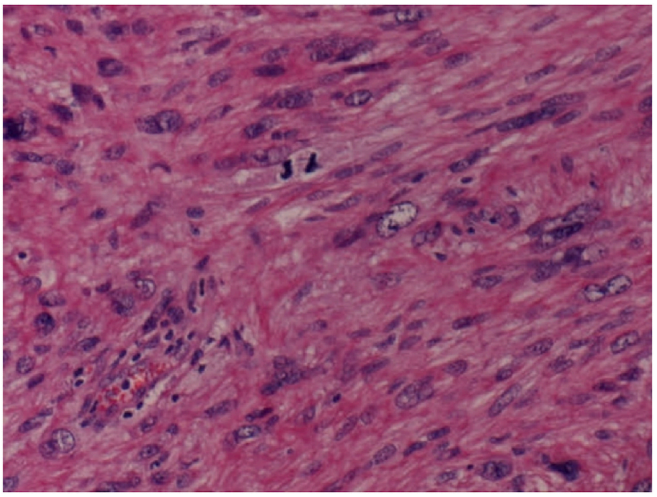

Fig. 2. The tumor is formed of spindle cells. A mitotic figure is seen in the upper aspect, 20× magnification.

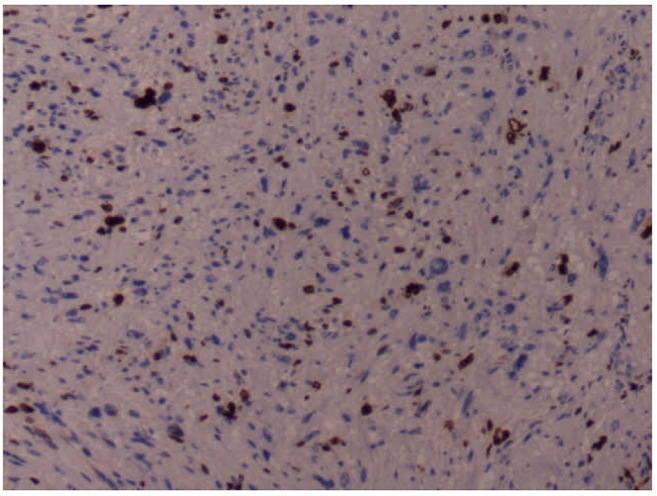

Fig. 3. Immunostain for Ki67. The ki67 index is about 15–20%, 10× magnification.

according to previously reported cases [7,9–11]. According to Cangiano et al., the mean age of diagnosing LMS of the GSV is 54 years with 3:2 ration of female to male [9]. Non-cutaneous soft tissue and cutaneous LMS occur commonly in men while vascular and retroperitoneal LMS occur more commonly in women [5].

LMS of the vessel walls can be categorized into three stages: non-occlusive, occlusive and terminal [9]. The majority of diagnosed cases of vascular LMS is incidental because it is asymptomatic in most cases, especially in inguinal regions. Therefore, LMS occurring around the inguinal region is diagnosed late in the terminal stage when the metastases usually occur [12]. However, around 10% of large vessel LMS develop early metastases and are found at the diagnosis [12]. Comparatively, our patient complained of a persistent, painful slow growing lesion for six months, which was atypical for large vessel LMS. It was small in size measuring around 1.5 cm while other cases in the literature reported a median tumor size of 4 cm [13]. The reason for having a large median size on diagnosis is that LMS are more likely to be discovered late and are usually asymptomatic. It is unusual to be painful in the early stages unless there is a compression on an adjacent nerve. According to Murakami et al., the anterior cutaneous nerve runs intimately along the GSV in the thigh region in around 42% of the population which is possibly the reason our patient developed pain early on [14]. Thus, due

to the natural slow progressing course of LMS and our patient not undergoing her biopsy as scheduled, caused a considerable delay in arriving at a diagnosis.

Radiologically, there is no specific imaging modality to confirm the diagnosis of LMS. Ultrasound can help raise suspicion of the disease and doppler can rule out other causes of lower limb swelling and deep vein thrombosis (DVT). The incidence of developing DVT with LMS can occur according to Narayanen et al. where they reported a case of 56-year-old woman who presented with a femoral triangle lesion associated with DVT of the femoral vein extended to the iliac vein [15]. On the other hand, CT and magnetic resonance imaging (MRI) are usually performed to stage the disease and rule out metastasis, especially in the liver and lung region as the tumor spread via hematogenous route [16]. Histopathologically, vascular LMS arises from the smooth muscle cells or the mesenchymal cells of the vessel walls [17]. In general, all types of LMS show typical sharply marginated fascicles of spindle cells with elongated, hyperchromatic nuclei [18]. However, non-spindle cells morphology has been known to occur but usually have a worst outcome [3]. Even though LMS can be identified solely by the light microscopy, immunohistochemistry stains can help confirm the diagnosis of undifferentiated tumors such as desmin, and hematoxylin and eosin stains. In addition, Ki67 proliferation index have a very high specificity and sensitivity value in differentiating LMS from other STS [19]. Similarly, our patient histopathology showed spindle cells and high Ki67 proliferation index.

Localized LMS are typically treated by surgical excision of the lesion with safety margins. However, metastatic cases are considered incurable, and the surgical treatment is aimed toward controlling the symptoms and decreasing the bulk which can prolong the survival rate of the disease [5].

A collaborated multidisciplinary plan of care between vascular surgery and oncology must be implemented, wherein post-operative surgical management is foreseen by vascular surgeons and referral for long term follow and monitoring for possible recurrence and metastasis is led by oncologists. Monitoring should commence 2–3 years after completion of treatment as this is the time period for highest chance of recurrence. Regular surveillance for a possible recurrence should be carried out with a follow-up every 3 months. Magnetic Resonance Imaging (MRI) is the most valuable imaging modality to monitor recurrence. A baseline MRI should be ordered 3 months after surgery and bi-annually at every 6-month interval for a period of 2 years [20]. Computed Tomography of the head, neck and abdomen can be used to rule out metastasis [21].The evidence of adjuvant therapy in the form of radiotherapy and chemotherapy is inconsequential [22]. Very few studies have been reported on the role and use of adjuvant therapy prior to surgery. Moreover, pre-operative chemotherapy has not proven to be effective [21]. Post-operatively, uterine and abdominal LMS can be treated with chemotherapy, radiotherapy or hormonal therapy, however, vascular LMS have been known to be chemo resistant while the role of hormonal therapy is feasible if the tumor is hormone-receptor positive [17]. Therefore, the role of post-operative chemotherapy is ambiguous and show no superiority as a management protocol. Surgical excision remains the treatment of choice with close monitoring for recurrence and metastasis.

4. Conclusion

The incidence of leiomyosarcoma of the great saphenous vein in the thigh region is very rare. Patient may present with early symptoms of swelling and pain. Excluding other causes of lower limb and inguinal region pain and edema. Suspicion of leiomyosarcoma should be raised by ultrasound while computed tomography is used for staging of the disease and to find any metastases. Histopathology and immunohistochemistry is highly sensitive and specific for leiomyosarcoma. Surgical excision of leiomyosarcoma remains the treatment choice with close monitoring for recurrence.

Ethical approval

An informed consent was obtained from the patient as well as the institute to publish this case.

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Author contribution

All authors contributed evenly to the conceptualization, drafting, data analysis, writing and proofreading of the research.

Trial registry number

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2. Unique Identifying number or registration ID: NA.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked):NA.

Guarantor

Awfa.

Consent

Informed consent was obtained according and in guidelines of the declaration of Helsinki.

Patient consent

Informed consent was obtained according and in guidelines of the declaration of Helsinki.

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Declaration of competing interest

The authors declare no conflict of interest.

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