CASE REPORT

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Leiomyosarcoma of the left external iliac artery: a case report and narrative review of the literature

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ABSTRACT

Leiomyosarcomas (LMS) arise from smooth muscle and represents only 6% of all sarcomas. LMS originating from major blood vessels, called vascular LMS, are detected mostly in the inferior vena cava. Arterial LMS are a rarity. We present a 43-year-old patient with a LMS arising from the left external iliac artery. The patient was referred to us with symptoms of left lower abdominal pain extending to the left limb and underwent a contrast computed tomography which suggested a suspicious mass near the left iliac vessels. She underwent laparoscopic excision of the tumour, whose histological examination revealed an LMS G2 arising from the external iliac artery. Immunohistochemically CD34, p53, Desmin, as well as smooth muscle actin, tested positive.

Keywords: Leiomyosarcoma, iliac vessels, external iliac artery, laparoscopy

Introduction

Leiomyosarcomas (LMS) are a rare type of sarcoma, originating from the smooth muscle and represent only 6% of all sarcomas. They can be classified into two types, the cutaneous type, derived from the arrector pili muscles associated with hair follicles and the subcutaneous type, derived from vascular smooth muscle.[1,2] LMS can arise in different areas of the human body such as the retroperitoneum, gastrointestinal tract, urogenital tract and soft tissue. LMS originating from major blood vessels' muscular walls, known as vascular LMS, are exceptionally rare, representing only 2% of all sarcomas.[1,3,4] The inferior vena cava (IVC) is the most commonly involved vessel.[5] Due to their rarity, there is a scarcity of evidence in the field of the diagnosis and treatment of vascular LMS. Our review aims to delve into the laparoscopic, imaging and immunohistochemical findings of this entity and to raise awareness among experts of this uncommon type of tumour thus improving its early detection and appropriate management.

Case Report

A 43-year-old patient was referred to our outpatient gynaecology department with a history of chronic left lower abdominal pain, radiating to the left lower limb. The patient had one normal delivery, a normal body mass index, a groin hernia operation, no other previous abdominal surgeries and no relevant family history. Our clinical examination did not reveal any abnormalities. Laboratory examination, including tumour markers (CEA, Ca19-9, Ca125, AFP) documented nothing of note. Transvaginal ultrasound detected a 4.5x4 cm, Doppler positive mass, of high

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malignancy suspicion in the left lower abdomen near to the left adnexa. The aforementioned finding was verified with further investigation by contrast computed tomography (CT) examination of the abdomen that demonstrated a suspicious mass (4.6x4 cm), near the left iliac vessels (Figure 1).

Based on the high suspicion of malignancy, derived from the CT scan results, an excision of the tumour via laparotomy was suggested to the patient, who did not consent and preferred the laparoscopic approach. Therefore, she was scheduled for a laparoscopic excision of the tumour. An informed, written consent was obtained. The operation was carried out by a gynaecology team with the local vascular surgery team available if needed. Intraoperatively a solid mass was detected lying on the left external iliac artery. The peritoneum was opened, and the tumour was stepwise separated from the left iliac artery (Figure 2). The tumour was then safely placed in a laparoscopic specimen retrieval bag and was extracted through mini-laparotomy in the suprapubic region. At the end of the procedure, no residual tumour was detected. The operation was performed successfully without any peri- or post-operative complications. The patient had an uneventful recovery and was discharged on the first postoperative day. Pathological and immunohistochemical examination of the mass followed, which revealed a leiomyosarcoma G2 according to Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC), arising from the external iliac artery.[6]

Histopathological Findings

a. Microscopic Findings

Histologically, there are components of a smooth muscle tumour, consisting of spindle cells in fascicular arrangement with occasional nuclear atypia and necrotic areas (Figure 3). Additionally, occasional mitotic figures are observed (up to four mitoses in 10 high-power fields). These findings align with a diagnosis of LMS with a G2 malignancy grade, according to FNCLCC, indicating an arterial-originated leiomyosarcoma.

b. Immunohistochemical Findings

Immunohistochemically, the tumour cells strongly express Desmin, smooth muscle actin (SMA), and predominantly caldesmon. CD34 highlights a dense network of compressed capillaries. There is no evidence of S100 protein. CD68 identifies numerous macrophages within the lesion. There is no convincing expression of MDM2, oestrogen- or progesterone receptor. RB1 shows heterogeneous expression, with some cells being negative, whereas p53 is detected in nearly all tumour cells. The Ki67 index is high, reaching over 20% in the examined paraffin block. A liposarcoma was excluded by using a FISH analysis with MDM2 amplification.

The case was presented to the multidisciplinary tumour board, which advised that the patient should undergo a laparotomy to exclude residual disease. The laparotomy was carried out two weeks later, and it was negative. The patient was discharged and scheduled for a 6-month

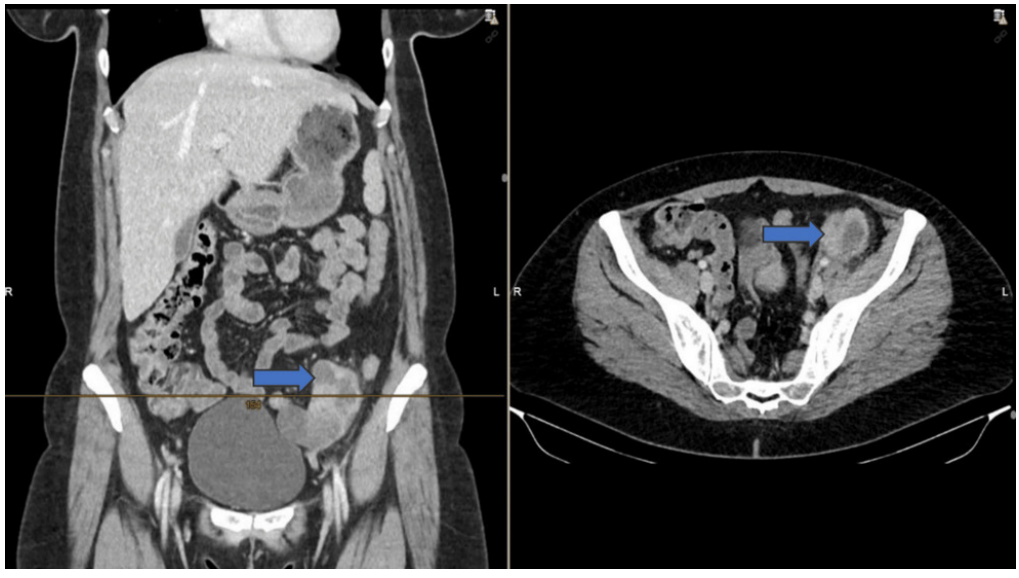

Figure 1. Abdominal CT scan. CT scan image, showing the left pelvic iliac tumour (see blue arrows). CT: Computed tomography.

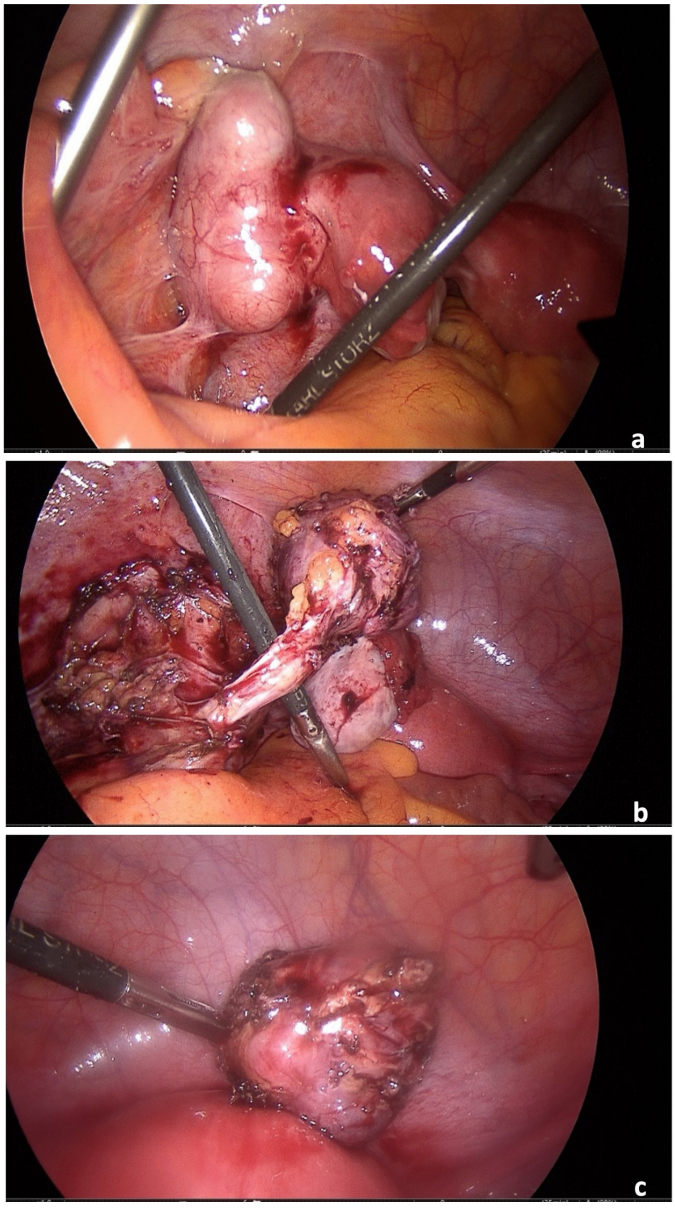

Figure 2. Laparoscopic tumour excision. a-c) Intraoperative photos of the tumour surrounding the left external iliac artery.

follow-up during the first postoperative year. No chemo- or radiotherapy was indicated.

Discussion

Vascular LMS represent a rarity amongst LMS and tend to originate within the venous vasculature.[7,8] The adult population, especially the female gender, are mostly affected by this type of tumour (commonly between 37-80 years).[9,10] In the 1970s Kevorkian and Cento[11] showed in a cohort of 86 vascular LMS cases, that the majority of the tumours (79%) are associated with venous origin, and most frequently the IVC. On the other hand, LMS arising from an artery represented only 21% of all cases, with the pulmonary artery being mostly involved. Furthermore, Leeson et al.[12] reported two rare cases of LMS, originating from the aorta and nine cases of the peripheral arteries, including the common iliac artery. In the reported cases where the lumen of the common iliac artery was implicated, the patients developed clinical signs and symptoms of Leriche syndrome.[12]

The clinical presentation of arterial LMS varies and depends on the location of the tumour. LMS involving peripheral arteries can lead to nerve compression, causing neurological symptoms.[13] Multimodality imaging plays a key role in the diagnostic algorithm of LMS, interpreting clinical symptoms and their association with their origin. Magnetic resonance imaging and CT scanning are of great importance in the evaluation of high LMS-suspicion tumours, giving the opportunity to optimally evaluate the tumour's size, its association with neighbouring tissues and structures, as well as providing evidence regarding potential metastases.[14]

Histopathological examination is of great value in the diagnosis and differentiation of LMS. Pathological diagnosis of a vascular LMS demands the use of standard haematoxylin-eosin and can be really challenging. The microscopic histological features of vascular LMS are similar to the other types of LMS, including necrosis, cellular atypia, and mitotic activity.[15] Spindle cells are typically found in these tumour types, mostly combined with subintimal fibrous changes. However, an immunohistochemical staining is essential for the diagnosis. Many biochemical and molecular markers, especially Cyclin-dependent kinases, and their role in diagnosis and prognosis of vascular LMS have been investigated.[16] In our case CD34, p53, Desmin as well as SMA were tested positive. Furthermore, Ki67 index was found to be 20%, while S100 protein was negative in all examined tissues.

The gold-standard treatment of vascular LMS is the surgical resection of the tumour. In most cases of vascular LMS published in the literature, the tumour was excised en bloc with the segment of the affected vessel with or even without reconstruction with a graft.[17] However, it can be challenging to achieve healthy resection margins, as LMS tends to have great proximity to vital anatomical structures. In cases where local tumour control is not possible, radiation could be an option. In our case, the tumour was extraluminal, arising from the surface of tunica media and also invaded tunica adventitia. So, its excision

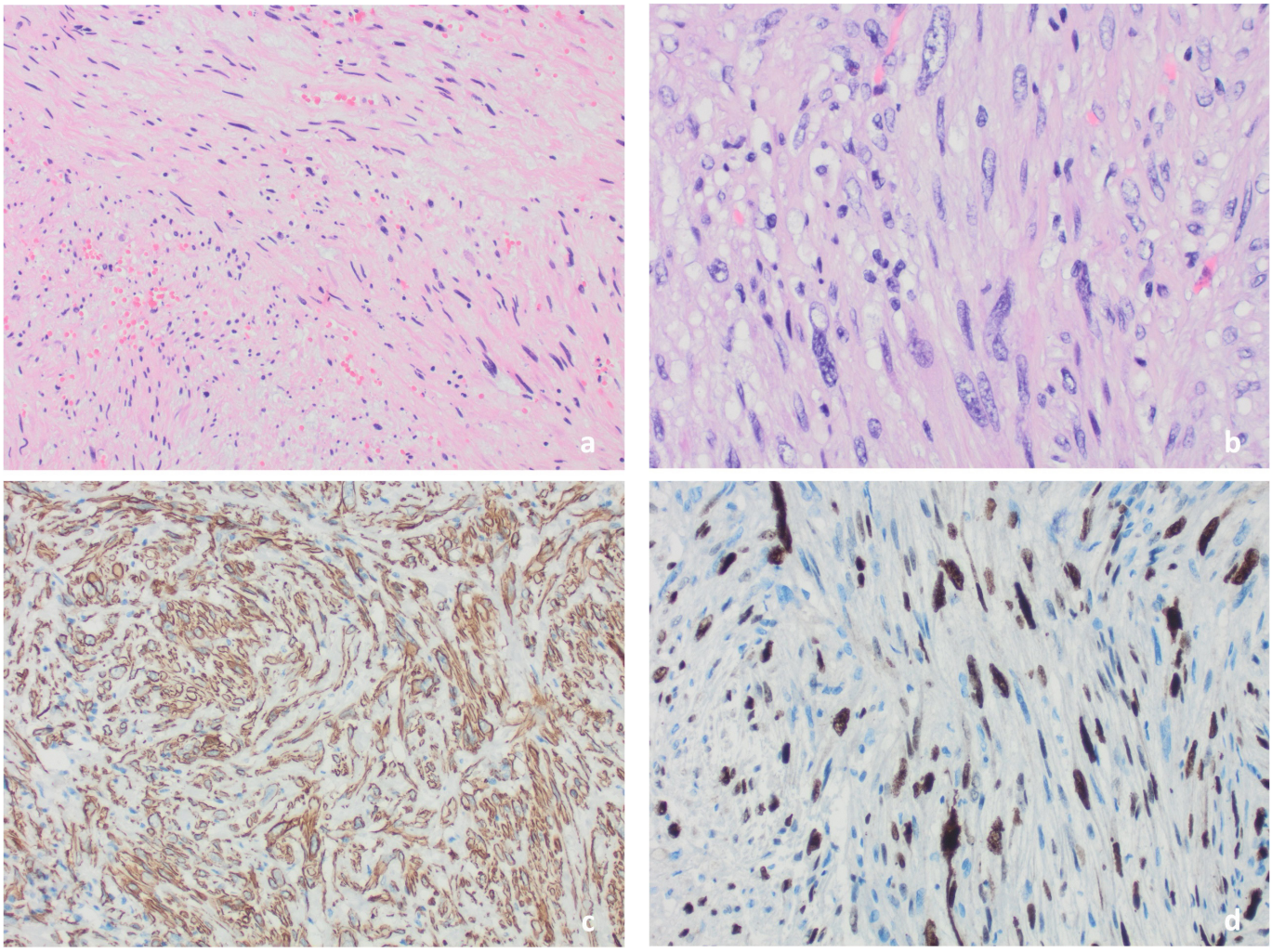

Figure 3. Histopathological findings. a, b) Hematoxylin and eosin stain (a: 200 magnification, b: 400 magnification). Nuclear pleomorphism, cytologic atypia and spindle cells arranged in haphazard fascicles with numerous mitotic figures.c) Caldesmon staining (200 magnification). Tumour cells with strong diffuse expression for caldesmon by immunohistochemistry. d) Ki67 staining (200 magnification). Expression of Ki67 in LMS (Ki67 20%).

was possible without affecting the lumen of the vessel, which remained intact, and no vessel reconstruction was needed.

Furthermore, not only surgical resection but also close follow-up of such patients is of great importance.[18] In our patient, the tumour surrounded the left iliac artery and was uncomplicatedly separated from the vessel. LMS, like other sarcomas, seem to have only a poor sensitivity to chemotherapy.[13,19]

Conclusion

Vascular and especially arterial LMS are a rarity. Early tumour diagnosis plays an important role in defining patients' prognosis. Due to the vascular origin and proximity of the tumour, general and gynaecology surgeons should be familiar with both clinical signs and symptoms and imaging features. As literature evidence is scarce, it is noteworthy to add that there is a definite need for multi-centre registries aiming to improve early diagnosis and disease treatment further.

Ethics

Informed Consent: Informed consent was obtained.

Acknowledgments: None.

Footnotes

Authorship Contributions

Surgical and Medical Practices: A.P., G.K.N., Concept: A.P., S.I.K., Design: A.P., G.K.N., Data Collection or Processing: A.P., S.I.K., Analysis or Interpretation: A.P., S.I.K., G.K.N., Literature Search: A.P., S.I.K., Writing: A.P., S.I.K., G.K.N.

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