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A Case of Leiomyosarcoma of the Femoral Artery; Rare Location and Management at an Advanced Stage

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Vascular leiomyosarcomas are extremely rare tumours, accounting for less than 1% of all malignant tumours. Venous leiomyosarcomas occur five times more frequently than arterial leiomyosarcomas. These are most often found in the large vessels and less than 50% occur in the peripheral circulation. Median survival has not been quantified. It can be good if radical surgery is performed. Treatment, whatever the stage, requires multidisciplinary management. Surgery with en bloc resection remains the treatment of choice for localised disease; in patients with unresectable locally advanced or metastatic disease, systemic treatment with essentially palliative aims may be proposed. Anthracycline-based treatment is the standard first-line therapy.

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We report a case report of a 50-year-old female patient with local, pulmonary and bone relapse of an operated left femoral artery leiomyosarcoma in whom we undertook palliative mono-chemotherapy.
Conclusion: Vascular leiomyosarcomas are extremely rare tumours, accounting for less than 1% of all malignant tumours. Median survival is dramatic for metastatic patients, with a median survival of 8 months, ranging from 5 to 20 months. Surgery remains the standard curative treatment for the localised stage; for stage 4, single chemotherapy is the treatment of choice.

Keywords: Leiomyosarcoma; femoral artery; surgery; localised disease; metastasis; chemotherapy; doxorubicin.

1. INTRODUCTION

Adult soft tissue and visceral sarcomas (excluding GIST) are rare tumours, with an estimated average incidence of 4-5/100,000/year in Europe. The most common types of soft tissue sarcoma are liposarcoma and leiomyosarcoma, each with an incidence of less than 1/100,000/year, while the majority of sarcoma histotypes have an incidence of less than 2/1,000,000/year [1].

Vascular leiomyosarcomas are extremely rare tumours, accounting for less than 1% of all malignant tumours. Venous leiomyosarcomas occur five times more frequently than arterial leiomyosarcomas. The latter are most commonly found in large vessels and less than 50% occur in the peripheral circulation [2].

To date, there are very few reports in the literature of arterial leiomyosarcoma involving the femoral artery, particularly in medical oncology.

Median survival has not been quantified. It may be good in the case of radical surgical treatment. A series of 22 patients published by Kieffer showed an average survival of 52% and 34.8% respectively at 3- and 5-years follow-up. However, the survival rate is dramatic for metastatic patients: median survival of 8 months, with survival ranging from 5 to 20 months [3].

The treatment of leiomyosarcoma at any stage requires multidisciplinary management [4]. Surgery is the standard treatment for all patients with localised adult-onset disease. It should be performed by a surgeon specifically trained in the treatment of sarcomas [5].

Radiotherapy is usually added to surgery as part of standard treatment for high-grade lesions (G2-3) [6,7]. It may also be omitted after multidisciplinary discussion, taking into account risk factors for local recurrence, including expected surgical margins, tumour size and histological type [8]. For metastatic patients, treatment depends on the resectability of the metastases, the patient's general condition and the oncologist's objective (objective response).

In this article, we report on the oncological management of a stage 4 leiomyosarcoma of the left femoral artery in a 50-year-old female patient.

2. CASE STUDY

A 50-year-old female patient presented with a hard, progressively enlarging, slightly painful mass in the left groin. She was grading 2 hypertensive and taking amlodipine 10 mg daily. She had no other medical or surgical history.

An MRI angiogram of the thigh showed an endovascular tumour process in the left femoral vein, initially suggestive of a leiomyosarcoma involving the femoral artery (Fig. 1). Angio scan of the aorta and lower limb confirmed the malignant nature of the mass and its locally advanced status, with involvement of the superficial femoral artery and superficial left femoral vein. A thoracic-abdominal-pelvic CT scan performed as part of the extension work-up was otherwise unremarkable.

Anatomopathological study of the biopsy of the mass showed a morphological appearance and immunohistochemical profile in favour of an intermediate grade 11 (3+1+1) leiomyosarcoma according to the FNLCC histological grading (Fig. 2, Fig. 3).

The patient was initially followed up in the vascular surgery department, where she underwent immediate surgery with complete resection of the tumour mass, and en bloc excision was performed. A mass measuring 3.1 x 2.7 cm was removed, and pathological examination confirmed the same histological

type. The patient was placed under surveillance with a follow-up scan every 3 months after a multidisciplinary consultation meeting.

Six months after surgery, a second post-surgical scan revealed secondary pulmonary and bone metastases. Given the appearance of these metastases, the patient was referred to the medical oncology department for further treatment.

We performed an MRI of the pelvis, which showed postoperative subcutaneous changes in the anteromedial aspect of the upper third of the thigh, with individualisation of an enhanced tissue mass after injection of contrast partially enveloping the superficial femoral artery, compatible with a tumour residue, and individualisation of a bone lesion in the anterior column of the left acetabulum, which remained suspicious (Fig. 4).

Clinically, she was in good general condition (WHO1). On physical examination, she had a surgical scar on the inside of her left thigh.

In clinical conclusion, we are faced with a 50-year-old patient, hypertensive on treatment, presenting with a metastatic relapse in the lung and bone of an operated leiomyosarcoma of the femoral artery, in whom the decision of the multidisciplinary consultation meeting concludes with a single chemotherapy with doxorubicin at a dose of 75 mg/m2 every 3 weeks.

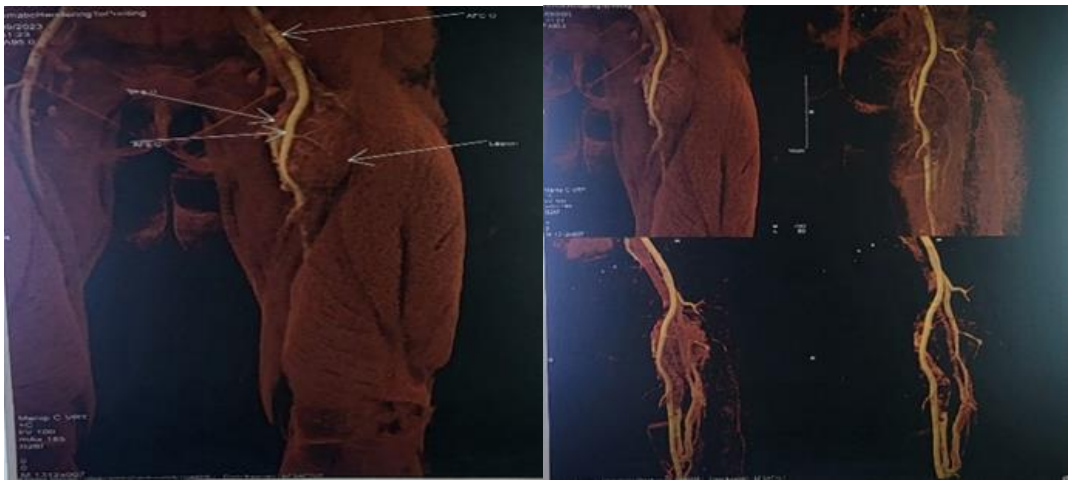

Fig. 1. MRI angiography: Endovascular tumour process of the left femoral vein, suggesting in the first instance a leiomyosarcoma encompassing the femoral artery

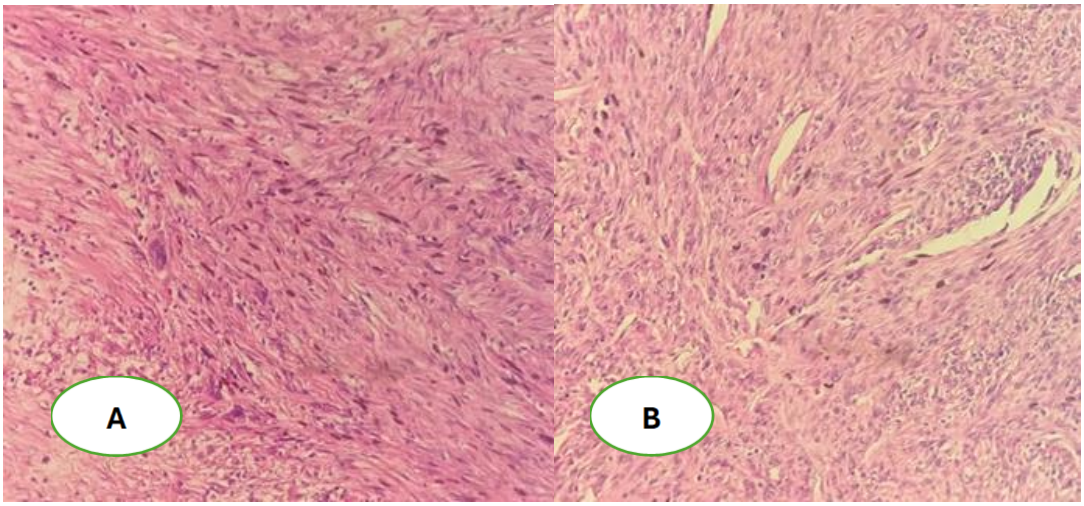

Fig. 2(A and B). Spindle cell tumour proliferation showing atypia and mitosis, HE 40X

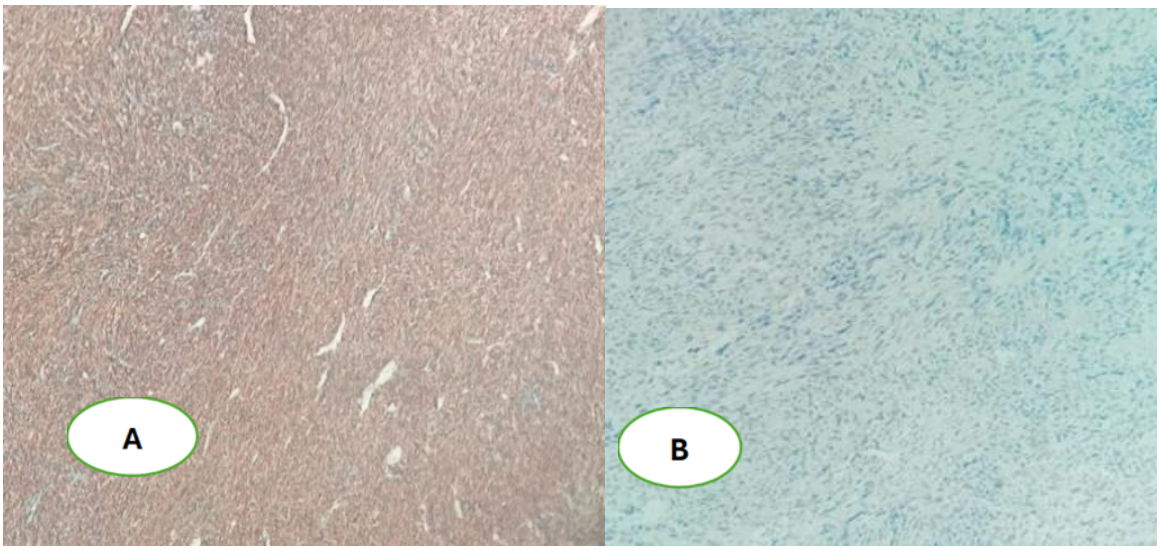

Fig. 3(A and B). Positivity of Anti-AML, H-Caldesmone antibodies (A), absence of expression of Anti- PS100, Anti-CD31 & CD34 antibodies (B)

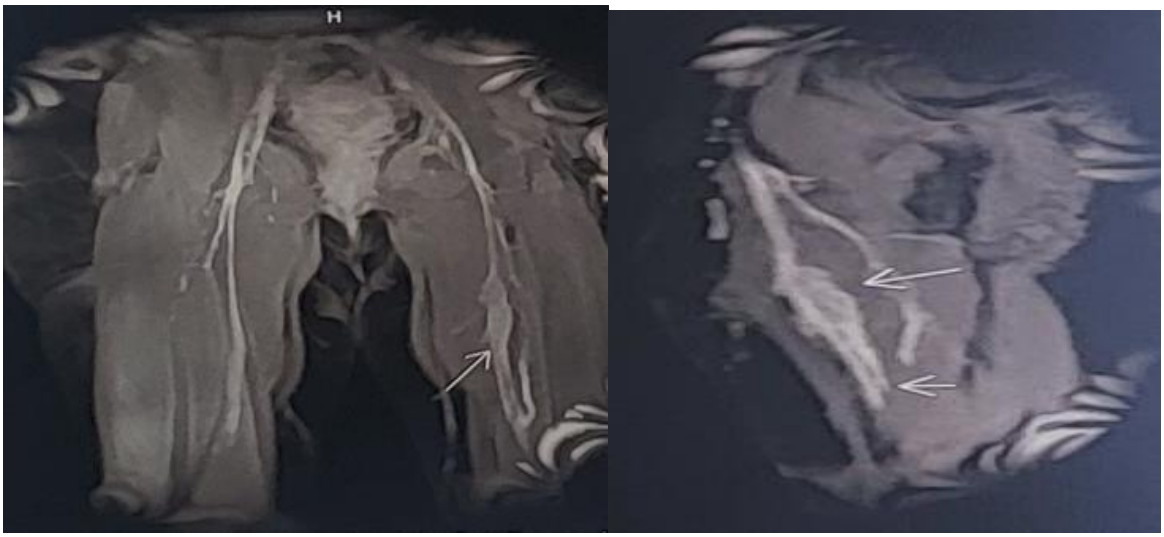

Fig. 4. MRI showing postoperative remodelling with tumour residue

3. DISCUSSION

Soft tissue sarcomas comprise approximately 80 entities defined by the World Health Organization (WHO) classification on the basis of a combination of distinctive morphological, immunohistochemical and molecular features [9].

First described in 1871 by Perl, vascular leiomyosarcomas are rare tumours. Their prevalence is difficult to quantify: it is estimated at 0.001%. The average age of onset is 59.5 ± 13.2 years. Leiomyosarcomas affect women 3 times more than men (73% versus 27%) [3].

In 2008, an estimated 300 cases were described in the medical literature [10]. Peripheral arterial locations are rare [2].

Leiomyosarcomas can give rise to metastases, but few series have been described. In Penel's series of 8 patients, 6 were metastatic. All had pulmonary metastases, two also had bone metastases, two had brain metastases and one patient had an adrenal metastasis. Median survival is poor for patients with advanced leiomyosarcoma: 8 months median survival with survival ranging from 5 to 20 months [11].

Post-surgical monitoring consists of close follow-up every three months to assess for signs of recurrence [12]. Routine chest X-ray and site-specific MRI should be requested every six months for a period of two years [13].

When managing patients with advanced or metastatic soft tissue sarcoma, decision-making is complex, depending on the various histological presentations, and should always be multidisciplinary. Resectable metachronous lung metastases (disease-free interval ≥ 1 year) without extra pulmonary disease are managed by surgery as standard treatment, if complete excision of all lesions is feasible, taking into account all prognostic factors [14].

In patients with unresectable locally advanced or metastatic disease, systemic treatment with essentially palliative aims may be offered. Anthracycline-based therapy is the standard first-line treatment. There is no formal proof that multiple chemotherapy is superior to single chemotherapy with doxorubicin alone in terms of overall survival. However, according to several randomised clinical trials, but not all, a higher response rate and longer progression-free survival can be expected in a number of sensitive histological types [15,16].

Consequently, polychemotherapy with an adequate dose of doxorubicin + ifosfamide may be the treatment of choice, particularly in histological types sensitive to ifosfamide, when a tumour response is considered potentially advantageous and the patient's performance status (PS) is good. Doxorubicin plus dacarbazine is an option for first-line polychemotherapy for leiomyosarcoma, in which the activity of ifosfamide is much less convincing, and for soft tissue sarcoma [17,18].

A phase III study compared doxorubicin monotherapy with the gemcitabine-docetaxel combination as initial treatment in patients with advanced soft tissue sarcoma of all types. The combination showed no improvement in progression-free survival or objective response rate and is not recommended as first-line treatment for patients with advanced soft tissue sarcoma including leiomyosarcoma [19]. Gemcitabine has also shown antitumour activity in leiomyosarcoma, angiosarcoma and epithelioid sarcoma administered as monotherapy [20].

In the case of our patient, in the light of all these recommendations, we opted for a protocol of single chemotherapy with doxorubicin at a dose of 75mg/m2 every 3 weeks after a cardiac ultrasound in which the LVEF was 65%. This treatment was combined with zoledronic acid 4mg every 3 weeks after dental treatment and monitoring of renal function.

Therapeutic evaluation after the 3rd cycle of treatment showed stable lesions in the pulmonary and bone metastases on CT scans, and clinical benefit with a reduction in symptoms (bone pain, cough and asthenia). However, during the course of treatment she experienced a few episodes of grade 2 neutropenia, which had no impact on the continuation of treatment.

4. CONCLUSION

Leiomyosarcomas are rare tumours with an unfavourable prognosis. Surgery with en bloc resection remains the treatment of choice for localised cases and certain metastases. However, cases of recurrence are still very common, and if they are unresectable, they should be managed with palliative chemotherapy. Preferably, single-drug chemotherapy should be administered in order to spare the patient the toxicities of systemic drugs, which can impair quality of life and are less effective.

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc) and text-to-image generators have been used during writing or editing of manuscripts.

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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